Methods: We reviewed the medical charts of the pediatric SLE patient in National Taiwan University Hospital from August 2015 to September 2019, and 50 SLE patients presented 185 episodes of suspicious activity or infection and received CRP, ESR, and Procalcitonin measurement were included. Time matched other laboratory parameters and clinical assessments were also collected. Episodes were divided into 4 groups: infected-active, infected-inactive, noninfected-active, and noninfected-inactive. Association of parameters with outcomes were predicted by generalized estimating equation. The receiver operating curve and the area under the curve were used to evaluate the diagnostic performance. We also used multinomial logistic regression model for nominal outcome, by setting noninfected-inactive group as the reference category.

Results: There were 7 males (14%) and 43 females (86%), with the mean ages 13.9 ± 4.4 years old. Most of the patients had renal (72%) or mucocutaneous (72%) involvement. The most common infection site was respiratory system (56%). Multivariate GEE analysis showed Damage index(DI), SLEDAI-2k, neutrophil-to-lymphocyte ratio (NLR), hemoglobin, platelet, RDW-to-platelet ratio (RPR), and C3 are independent parameters for predicting SLE activity flare. Combination of these seven parameters resulted in a model with calculated AUC of 0.8964 and with sensitivity of 82.2 % and specificity of 90.9%. Multivariate GEE analysis showed DI, fever, CRP, Procalcitonin, lymphocyte percentage, NLR, hemoglobin, and renal score in SLEDAI-2k are independent parameters for predicting acute infection. These eight parameters resulted in a model with calculated AUC of 0.7886 and with sensitivity of 63.5% and specificity of 89.2%. We select a total of 10 variables (DI, SLEDAI-2k, Fever, Procalcitonin, lymphocyte percentage, NLR, hemoglobin, platelet, RPR, C3) to establish multinomial logistic regression, then predict four groups with accuracy of 70.13% for infected-active, 10% for infected-inactive, 59.57% for noninfected-active, and 84.62% for noninfected-inactive.

Conclusion: The proposed predictive calculator could be a useful tool for differentiation between activity flares and acute infections in pediatric SLE. Obtaining and combination of several parameters is effective and helpful to make appropriate judgement and treatment decisions for SLE patients.

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FRI0470 CARDIORESPIRATORY FITNESS IN JUVENILE IDIOPATHIC ARTHRITIS IN 6 – 17-YEAR-OLD CHILDREN- CROSS-SECTIONAL STUDY

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Background: Children with juvenile idiopathic arthritis (JIA) have been found to have reduced cardiorespiratory fitness and lower physical activity. Poor cardiorespiratory fitness is associated with a risk of cardiometabolic diseases.

Objectives: The aim of this study was to study the levels of cardiorespiratory fitness, respiratory function and hemodynamic responses during and after maximal cycle ergometer exercise test in children with JIA aged 6-17 years and compare the results with healthy controls.

Methods: Study group in this analysis consisted of 43 patients with JIA who were treated in Department of Pediatrics in Kuopio University Hospital, Finland and 40 healthy age- and sex matched controls. Maximal exercise tests were carried out with an electromagnetic cycle ergometer using a pediatric saddle module. Maximal workload per kilogram ($W_{\max}/kg$) was used as a measure of cardiorespiratory fitness and was presented relative to bodyweight. In addition the peak values of VO_2 per kilogram ($VO_{2\max}/kg$) were used as a measure of highest amount of oxygen that an individual can consume during exercise. Values of $VO_{2\max}$ were collected from respiratory gases measured directly from breath by breath method and was presented relative to body weight.

Physical activity and sedentary behavior (minutes per day) was assessed by the PANIC (Physical activity and nutrition in children -study) Physical Activity Questionnaire which the participants filled.

Results: Statistical analyses were performed for 43 children with JIA and 40 controls. Mean age in JIA group was 12.09 years (95%CI 11.04-13.14), and 11.72 years (95%CI 10.52-12.93) in controls ($p=0.572$). Mean body mass index for age (BMI) was $22.58 kg/m^2$ (95%CI 21.54-23.62) in JIA and $18.95 kg/m^2$ (95%CI 17.73-20.16) in controls ($p<0.05$). In JIA group BMI was 19.18 % higher compared to controls. Mean physical activity in JIA group was 94.11 minutes per day (95% CI 81.09-107.13), and 122.54 minutes per day (95% CI 102.84-142.24) in controls, thus JIA group was 23.20 % less physically active than controls ($p=0.015$).

Mean $W_{\max}/kg$ was $2.65 W/kg$ (95% CI 2.49-2.82) in JIA and $3.01 W/kg$ (95%CI 2.86-3.15) in controls thus $W_{\max}/kg$ in JIA was $0.36 W/kg$ (11.8 %) lower than in controls, ($p = 0.002$). $VO_{2\max}/kg$ was 37.00 (95%CI 33.96-40.84) ml/kg/min in JIA and 43.30 (95%CI 40.79-45.82) ml/kg/min in controls thus in JIA group mean $VO_{2\max}/kg$ was $6.3 ml/kg/min$ (14.4 %) lower than in controls ($p=0.001$).

Conclusion: Children with JIA were found to have significantly lower cardiorespiratory fitness. In addition, BMI in JIA patients was higher compared to healthy age- and sex-matched controls. Impaired cardiorespiratory fitness and higher BMI may predispose children with JIA to cardiometabolic comorbidities later in life. In addition to disease-control, more attention should be paid to maintaining good cardiorespiratory fitness and normal BMI in these patients already before adulthood.

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FRI0471 ADVERSE FACTORS OF COMORBID DISEASES DEVELOPMENT AT DIFFERENT VARIANTS OF JUVENILE IDIOPATHIC ARTHRITIS (JIA)

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Background: It is known that rheumatic diseases are the pathogenetic basis for the formation of many comorbid conditions, the most important of that are cardiovascular pathology, atherosclerosis, osteoporosis, chronic kidney disease and amyloidosis, chronic obstructive pulmonary disease. The start of the disease at an early age, the long-term duration of JIA, the use of basic immunosuppressive therapy lead to the possibility of the onset of the first signs of comorbid conditions in childhood.

Objectives: To study risk factors for the formation of damage of internal organs and systems in children with non-systemic JIA.

Methods: The case histories of 121 patients aged 7-18 years (mean age 11.0 ± 0.3 years) with polyarticular (67.7%), oligoarticular (14.88%) and uveitis-associated (17.35%) JIA were studied, mainly of females (73.5%). The age of the start of the disease was 5.9 ± 0.4 years, the duration of JIA at the time of analysis reached 67.1 ± 4.3 months. All children received basic methotrexate therapy (plus folic acid), short courses of NSAIDs. There are studied changes in the cardiovascular system (ECG, ultrasound, 6-minute walk test),

lungs (spirometry), kidneys (concentration function, glomerular filtration rate, daily microalbuminuria), lipid spectrum and blood coagulation. Deviations from standard age-gender normal values were taken into account. It was used statistical methods for identifying relationships (stepwise multiple regression). As the dependent value the number of detected changes in the state of internal organs and systems is taken. The independent variables included standard clinical and laboratory parameters, the age of the onset of JIA, the total duration of the disease, the characteristic of the articular syndrome, activity indicators (C-reactive protein, RF and ANA titers, JADAS27), as well as the term for the appointment of basic therapy relative to the debut of the disease, the nature of the dosage methotrexate (mg / m² / week), the duration of its use at the time of examination.

Results: Based on the analysis and the obtained regression models for the formation of extra-articular lesions in children with various JIA options, it was found that the leading factors in the formation of comorbidity are the duration of the disease ($p = 0.01$), VAS results (including the child, parents and the physician) ($p = 0.01$), the dose of methotrexate ($p = 0.01$).

In the polyarticular variant, the body mass index ($p = 0.02$), erythrocyte content ($p = 0.007$) were significant for the formation of comorbid states and should be distinguished.

The overall duration of the disease ($p = 0.02$), the low age of initiation of therapy ($p = 0.05$), VAS of child ($p = 0.003$), CRP ($p = 0.007$), dose of methotrexate ($p = 0.001$) had the greatest significance level among the clinical and laboratory indicators included in the regression model of the formation of comorbidity with the oligoarticular variant.

A features of the prognostic model for the formation of organ lesions and metabolic disorders in uveitis-associated arthritis were GCS therapy presence ($p < 0.001$), patient age ($p < 0.001$), JADAS27 ($p < 0.001$) and methotrexate dose ($p < 0.001$).

Conclusion: Thus, in children with JIA, the formation of comorbid pathology is associated with a lower age of JIA debut in oligoarthritis and uveitis-associated arthritis, low body weight in poly- and oligoarticular JIA variants, an increase in the duration of the disease in the polyarticular variant, and the level of activity of the process, respectively, JADAS 27 ($p < 0.01$) and VAS. Prognostically unfavorable for the formation of pathological changes in the internal organs and homeostasis are anemia, high white blood cell count and ESR level.

Disclosure of Interests: None declared

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FRI0472 DIAGNOSIS AND MANAGEMENT OF ADENOSINE DEAMINASE 2 DEFICIENCY CHILDREN: THE EXPERIENCE FROM CHINA

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Background: Adenosine deaminase 2 deficiency (DADA2) is a rare antoinflammatory disease caused by mutations in ADA2 gene, few Chinese cases have been reported.

Objectives: To describe and compare the clinical features, genotypes, and treatments of Chinese DADA2 patients and foreign cases.

Methods: Primary immunodeficiency disease Panel or Whole Exome Sequencing was performed to suspected subjects, and assays for adenosine deaminase 2 (ADA2) enzyme activity were also carried out to them and their parents. Case reports of Chinese and foreign patients with DADA2 were searched from PubMed and Chinese domestic databases.

Results: Seven unrelated DADA2 children from China were included in our study, 5 were identified at Peking union medical college hospital and 2 had been reported previously (1 on PubMed and 1 in Chinese literatures). 14 mutations in ADA2 were identified, and 9 of which have not been found in other countries. Four children receiving enzymatic analysis had lower ADA2 enzyme activity compared to their parents. Phenotypic manifestations included fever, skin symptoms, vasculitis, neurologic involvement, et al. The treatments varying from steroids, immunosuppressants, and tocilizumab, anti-TNF therapy and hematopoietic stem cell transplantation (HSCT) were effective depending on different phenotype and severity.

Conclusion: This study includes the biggest number of Chinese DADA2 patients at present. We recommend combination of enzymatic analysis with gene screening to confirm the diagnosis. Genotypes of patients from China were some different, the clinical manifestations were similar. We suggest anti-TNF therapy may not be necessary for mild case and HSCT should be considered even without hematological phenotype.

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Other orphan diseases

FRI0473 ASSESSMENT OF FEMORAL VEIN WALL THICKNESS WITH DOPPLER US AS A DIAGNOSTIC TOOL FOR BEHCET'S DISEASE

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Background: Diagnosing Behcet's disease (BD) is a challenge, especially in countries with a low prevalence. International Study Group Criteria, accepted to as diagnostic, has low sensitivity, especially in early cases when major organ involvement such as uveitis or deep vein thrombosis (DVT) presents alone. We recently published a controlled study of assessing venous wall thickness (VWT) as a surrogate marker of venous disease in BD with ultrasound (US) and observed a very sensitive and specific VWT in male BD patients. The common femoral vein (CFV) thickness measurement, as the primary site of US with the cut-off values > 0.48 - 0.49 mm, had a high area under the receiver operating characteristic curve (> 0.8) with sensitivity and specificity of around 80% (1).

Objectives: In this study, we aimed to investigate the diagnostic performance of CVF thickness measurement in BD including females comparing with multiple control disease groups.

Methods: One hundred-ten patients with BD, 47 healthy controls (HC), 21 patients with systemic vasculitides, 28 patients with venous insufficiency, 29 patients with antiphospholipid syndrome (APS) having DVT history, were included the study. Bilateral CFV thickness was measured with US by an experienced radiologist blinded to cases (Figure 1).

Figure 1. Measurement of common femoral vein thickness