**Results:** In children with SLE a significant increase in the content of total cholesterol (5.56±0.36 mmol/l) and a decrease in HDL-cholesterol levels (0.94±0.18 mmol/l) were found in comparison with the control group (3.71±0.69 mmol/l and 1.92±0.33 mmol/l, respectively). A significant (p <0.05) decrease in the concentration of ApoA (85.1 [59.8; 94.9] mg/dl), ApoE (2.1 [12; 3.7] mg/dl) and an increase in ApoB (59.8 [519; 678] mg/dl) in children with SLE were found compared with the control group (1272 [122.1; 132.3] mg/dl, 3.2 [2.3; 5.9] mg/dl and 32.1 [19.9; 50.8] mg/dl, respectively). ApoB / ApoA was increased in 7 (28%) children with SLE. The study found a significant (p <0.05) increase in the level of intermediate (DK233, DK278) and final (MDA) LPO products in the blood serum of children with SLE in comparison with the control group, which indicates the activation of LPO processes in these patients. During the correlation analysis, a positive correlation was established between the levels of DK233, DK278 and blood serum and CRP (r = 0.87, p <0.001). When studying the main indicators of the blood lipid spectrum in children with SLE, a significant increase in the serum concentration of total lipids (p <0.01) and triglycerides (p <0.001) was revealed when compared with the control group. When determining indicators of coagulation hemostasis, in children with SLE, a predominance of hypercoagulation was detected, accompanied by a significant increase in serum fibrinogen level (5.08 ± 0.14 g/l) and an increase in platelet level (479.57 ± 8.01*10^9/l) in peripheral blood compared with the control group (3.24 ± 0.7 g/l and 294.23 ± 5.39*10^9/l, respectively). These indicators correlated with serum CRP concentration (rS = 0.62, p <0.01) and ESR level (rS = 0.73; p <0.01). A relationship was established between elevated serum levels of fibrinogen and disease activity indicators (r = 0.74; p <0.01).

**Conclusion:** The atherogenic orientation of the blood lipid spectrum, characterized by hyperlipidemia, hypertriglyceridemia, hypercholesterolemia and dyslipoproteinemia in the form of a decrease in HDL-cholesterol and an increase in LDL-cholesterol, as well as an increase in ApoB/ApoA ratio> 1 and a decrease in ApoE, the activation of LPO processes and a significant decrease in ACW and ACL in the serum of children with SLE are cardiovascular risk factors (pulmonary thromboembolism, myocardial infarction and brain).

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**AB1010**

**CLINICAL SPECTRUM AND IMMUNE ANALYSIS OF PATIENTS WITH CRYOPRYVIN-ASSOCIATED AUTOINFLAMMATORY SYNDROME IN TAIWAN**

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**Background:** Cryopyrin-associated periodic syndromes (CAPS) are emerging autoinflammatory diseases with available treatment. No reports have yet been reported from Taiwan.

**Objectives:** We reviewed cases suspected with CAPS to identify its existence in Taiwan.

**Methods:** Genomic DNA from one hundred and ten cases with symptom suggestive of CAPS1 (1) between 2016-2019 were sent for NLRP3 gene analysis. Clinical presentations, laboratory data, treatment regimens, as well as infammasome activities were analyzed among those treated in a tertiary medical center in northern Taiwan.

**Results:** Among the 110 cases sequenced, 16 of them were found to carry missense mutations within the NLRP3 gene. Fourteen cases harbored known pathogenic genetic variants (c.1316C>T; c.1574A>T; and c.907G>C) and two carried novel NLRP3 missense mutations (c.210G>A, c.1371G>T)(2) with unknown pathophysiological roles. Through chart review, chronic urticarial, systemic juvenile idiopathic arthritis, Behcet’s disease, and reflexory Kawasak disease were most likely diagnosed before genetic analysis were arranged. As compared to chronic infantile neurological, cutaneous and articular syndrome (CINCA) and Muckle-Wells syndrome (MWS), familial cold autoinflammatory syndrome (FCAS) was the most frequently observed clinical presentation. Plasma serum amyloid A (SAA) and IL-1b were both significantly elevated among the cases diagnosed with CAPS as compared to the controls (p<0.05). IL-1b, on the other hand, showed no significant differences between the groups. While the presence of LPS without ATP significantly increased the level of IL-1b in the PBMC stimulation test, IL-1b were significantly elevated in the confirmed CAPS with or without ATP upon LPS stimulation (all p<0.05). Caspase 1 activity were also tested positive among the cases with CAPS. Furthermore, we compared the immune profiles between those CAPS cases harboring pathogenic mutations with the 2 harboring unreported NLRP3 missense mutations and discovered that the PBMC stimulation test in cases with c.210G>A and c.1371G>T mutation did not differ from the healthy controls.

**Conclusion:** The number of NLRP3 gene alterations among patients suspected with CAPS in Taiwan is not low. In order to identify potential patients for proper medical intervention in the future, physician awareness, genetic testing as well as functional analysis is important.

**References:**

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**AB1011**

**LONG TERM FOLLOW-UP OF THE PATIENTS WITH ANTI NUCLEAR ANTIBODY POSITIVITY WHO HAD INITIALLY NO IDENTIFIABLE RHEUMATIC DISEASES**

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**Objectives:** To evaluate the long-term outcome of the patients with ANA positivity who initially did not have an identifiable rheumatic disease.

**Methods:** A total of 37 children were included in this study. All were positive for anti-nuclear antibody screening, ANA by indirect immunofluorescence (IIF) test. 19 patients had ANA positivity only and 18 patients had ANA positivity with other autoantibodies. Eight patients (21.6%) were positive for anti-dsDNA, 2 patients (5.4%) for anti-Sm, 1 patient (2.7%) for anti-SSA, 9 patients (24.3%) for anti-SSB, 1 patient (2.7%) for anti-SSA/SSB, and 2 patients (5.4%) for anti-CENP.

**Results:** The median duration of follow-up was 8 years (0.5-13 years). At the end of follow-up, 20 patients (54%) were still ANA positive. No evidence of rheumatic disease suggesting a new diagnosis was found in 6 patients (16%). Two patients (5.4%) were found to have new diagnoses of juvenile rheumatoid arthritis (JRA) and systemic lupus erythematosus (SLE) respectively. Three patients (8.1%) were excluded from the study. The median age at the end of follow-up was 15 years (range: 12-17 years). The median age at diagnosis was 3 years (range: 1-11 years).

**Conclusion:** The majority of patients with ANA positivity who initially did not have identifiable rheumatic disease were still ANA positive after a median follow-up of 8 years. Only 21% of the patients had new rheumatic diagnoses during follow-up. Therefore, the ANA positivity in children without any identifiable rheumatic disease had a benign course.