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 1.29 ± 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 [1.2; 3, 7] mg/dl) and an increase in ApoB (59.8 [51.9; 67.8] mg/dl) in children with SLE were found compared with the control group (127.2 [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 > 1 was established 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 in blood serum and CRP ($r_s = 0.87$, $p < 0.001$). 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 ($479.57 \pm 8.01 \times 10^9/l$) in peripheral blood compared with the control group (3.24 ± 0.07 g / l and $294.23 \pm 5.39 \times 10^9/l$, respectively). These indicators correlated with serum CRP concentration ($r_s = 0.62$; $p < 0.01$) and ESR level ($r_s = 0.73$; $p < 0.01$). A relationship was established between elevated serum levels of fibrinogen and disease activity indicators ($r_s = 0.74$; $p < 0.01$).

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 events risk factors (pulmonary thromboembolism, myocardial infarction and brain).

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AB1009

THE EFFICACY OF TOCILIZUMAB ON THE TREATMENT OF TAKAYASU ARTERITIS IN CHINESE CHILDREN

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Background: Takayasu arteritis (TA) is the most prevalent large-vessel vasculitis in children. Patients with TA have a high morbidity and mortality. It remains a therapeutic challenge because corticosteroids monotherapy can rarely cure TAK and the relapse rate is high during GC tapering.

Objectives: The aim of this study is to investigate the efficacy and safety of tocilizumab (TCZ) in Chinese children with Takayasu arteritis (TAK).

Methods: We retrospectively studied 6 TAK children treated with TCZ in our hospital from July 2017 to October 2018. The demographic and clinical data, laboratory examination results and vascular imaging data were collected.

Results: Six pediatric patients with critical or refractory TAK treated with TCZ were analyzed, including 3 males and 3 females. The diagnosis age was ranging in age from 2 to 13 years (median age: 7 years). Three patients were initially treated with TCZ and Mycophenolate Mofetil (MMF) as the first-line regimen without corticosteroid or with a quite rapid GC taper duration, two of which had life-threatening coronary arteries involved and heart failure. The other three patients were switched to TCZ from conventional disease modifying anti-rheumatic drugs (DMARDs) or other biologics due to being refractory to them and recurrent relapses. Four patients were given TCZ at 4 weeks regular intervals for 10 to 22 months, while two patients withdrew TCZ because of disease deterioration and unbearable abdominal or chest pain after the second dose. After 6 months follow-up, four patients experienced significant clinical and biological improvement with angiographically progression in one patient. A corticosteroid-sparing effect is obvious. Drug-related side effects occur in 1 patients manifesting as a mild elevated liver function. Neither neutropenia nor infection was observed.

Conclusion: Our study shows a clinical, biological, and radiological response in patients with refractory TAK treated with TCZ.

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AB1010

CLINICAL SPECTRUM AND IMMUNE ANALYSIS OF PATIENTS WITH CRYOPYRIN-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 signs suggestive of CAPS(1) between 2016-2019 were sent for NLRP3 gene analysis. Clinical presentations, laboratory data, treatment regimens, as well as inflammation 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 refractory Kawasaki 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-18, 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-18 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 are important.

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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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Background: Anti-nuclear antibodies (ANA) are a group of the antibodies that develop against intracellular components of the cells. It is usually useful for diagnosing some of the connective tissue diseases like systemic lupus erythematosus, mixed connective tissue disease. But it is reported that its positivity rate is about %20 in healthy individuals. Therefore, it can be confusing to check ANA test, if there is not really high suspicion for connective tissue diseases or juvenile idiopathic arthritis.

Objectives: We aimed to evaluate results of long-term follow-up of the patients with ANA positivity who had initially no identifiable rheumatic diseases.

Methods: Six hundred and ninety-four patients with ANA positivity who did not diagnosed as any of the rheumatic diseases at the first examination were found in database. Two hundred and eighty-two patients of them were called so far and questioned about their demographic features and symptoms that are related with rheumatic diseases.

Results: Mean age of the patients at the time of study and at the time of testing were 13.4 ± 4.5 and 9.1 ± 4.0 years. The female: male ratio was 1.05. Mean follow-up duration was 4.3 ± 2.8 years. Most common reasons for the request for ANA test were arthralgia and skin eruptions. ANA testing was most commonly requested by a general pediatricists. Demographic features of the patients were summarized in Table 1.

Table 1. Demographic features of the patients.

	n (%)
Age (years)	13.4 ± 4.5
Female	145 (51.4)
Age at the time of testing (years)	9.1 ± 4.0
Follow-up Duration (years)	4.3 ± 2.8
Reason for testing	
Arthralgia	99 (44.1)
Skin Eruption	54 (24.1)
Check-Up	20 (8.9)
Arthritis	13 (5.8)
Gait abnormalities	7 (3.1)
Hair Loss	6 (2.6)
Fever	5 (2.2)
Uveitis	2 (0.8)
Recurrent abdominal pain	2 (0.8)
Who suggested testing?	
Pediatrician	196 (87.5)
Parents	13 (5.8)
Dermatologist	7 (3.1)
Ophthalmologist	3 (1.3)
Rheumatologist	2 (0.8)
Other	3 (1.3)
Positivity of acute phase reactants	15 (5.3)
History of infection before testing	56 (24.3)
History of drug-using before testing	39 (17)

Most of the diseases were diagnosed in patients with ANA positivity were not related with autoimmune mechanisms that associated with ANA positivity therefore, these diseases are thought to be coincidence. Only in 1 patients, systemic lupus erythematosus that has certain association with ANA positivity were diagnosed. All diseases that are diagnosed were shown in Table 2.

Table 2. All diseases that are diagnosed in patients during the follow-up period.

	n (%)
Hypermobility Syndrome	29 (10.2)
Urticaria	7 (2.4)
Hypothyroidism	6 (2.1)
Transient synovitis	4 (1.4)
Chronic ITP*	4 (1.4)
Scoliosis	4 (1.4)
Familial Mediterranean Fever	3 (1)
Cryopyrin associated periodic Syndrome	2 (0.7)
PFAPA syndrome**	2 (0.7)
Celiac Disease	2 (0.7)
Acute Rheumatic Fever	2 (0.7)
Fibromyalgia	1 (0.3)
Bone Tumor	1 (0.3)
Juvenile Idiopathic Arthritis	1 (0.3)
Henoch-Shönlein Purpura	1 (0.3)
Myasthenia Graves	1 (0.3)
Sever Disease	1 (0.3)
Vitiligo	1 (0.3)
Systemic Lupus Erythematosus	1 (0.3)

*Idiopathic Thrombocytopenic Purpura, **Periodic fever, aphthous stomatitis, pharyngitis and cervical adenitis syndrome.

Conclusion: We are reporting that in only 0.3% of patients with ANA positivity who don't have any diseases diagnosed initially, were diagnosed as rheumatologic diseases during to the follow-up period. Since positivity of ANA is also common in the healthy population, requesting this test in only patients with high suspicion for connective tissue disease will reduce confusion in terms of diagnosis.

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Other orphan diseases

AB1012

INVESTIGATION OF RELATIONSHIP BETWEEN SEXUAL FUNCTION AND OPTICAL COHERENCE TOMOGRAPHY ANGIOGRAPHY IN PATIENTS WITH BEHÇET'S: PRELIMINARY STUDY

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Background: Patients with Behçet's have sexual dysfunction in recently literature. However, depression and anxiety accompanying these studies were not excluded. In addition, increased sexual dysfunction in patients with Behçet's has been reported in these studies in relation to depression and anxiety. Also it has been stated that endothelial dysfunction decreases the vasodilator neurotransmitter level. It was emphasized that this may lead to erectile dysfunction in disease of Behçet's.

Objectives: The aim of this study is to evaluate the relationship between erectile dysfunction (ED) and radial peripapillary capillary (RPC) density determined by Optical Coherence Tomography Angiography (OCTA).

Methods: Patients under 45 years old who had no known additional disease and had no eye and neurological involvement were included in the study. The patients were evaluated with Beck Depression and Anxiety Scale. Accordingly, patients without anxiety and depression were included in the study. An International Index of Erectile Function (IIEF) questionnaire was applied to a total of 18 patients who met the inclusion criteria. Detailed ophthalmological examination and OCTA imaging were performed by the ophthalmologist. Patients whose eye bottom could not be evaluated clearly (Corneal pathology, cataracts, etc.), image quality was 0.5 and below, and additional eye disease (glaucoma, optic neuropathy, maculopathy) were excluded from the study.

Results: The mean age of the patients was 37 ± 6.5 years, and the duration of the disease was 82.37 ± 47.78 months. There was a strong relationship between RPC Small Vessel Whole Density (RPC-SVWD) and overall satisfaction and sexual satisfaction ($p: 0.036$ and 0.045). There was a strong positive relationship between RPC Small Vessel Peripapillary Density (RPC-SVPD) and erectile function, sexual desire, sexual satisfaction and general satisfaction ($p: 0.036, 0.032, 0.005, 0.004$). There was a strong positive relationship between RPC All Vessel Peripapillary Density (RPC-AVPD) and erectile function and sexual satisfaction ($p: 0.048, 0.015$).

Conclusion: In our study, a significant relationship was found between sexual function and RPC density. In the literature, it has been found that nitric oxide synthesis decreases due to endothelial dysfunction in patients with Behçet's. Although there are preliminary study data, we found a significant relationship between the vascular layer of both the penis and the eye in our study. The data we obtained in this study, in which all factors that may cause erectile dysfunction are excluded, show that Behçet's, which is theoretically known to affect all vascular structures, can affect sexual functions by its nature. In our study, although there is a relationship between ED and OCTA data in Behçet's disease, the large number of cases of our ongoing study will provide clearer information.

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