ferritin and d-dimer levels, with high C3, IL-6 and TNF-alpha levels. Microbial cultures (blood cultures, nasopharyngeal and throat swab) were sterile, and serology for Cytomegalovirus, Epstein-Barr virus, Human parvovirus B19 and Bartonella henselae were negative, while serology for Mycoplasma pneumoniae (IgM and IgG) was positive. Azithromycin was prescribed, but fever and elevated inflammatory blood markers persisted and other procedures were performed to rule out rheumatic disease. Radiogram and bone scintigraphy were unremarkable, but an ultrasound revealed a retroperitoneal mass. MRI was performed, and retroperitoneal mass measuring 3x2.5 cm, arising from the pancreas and suspected to be a neuroendocrine tumor or enlarged lymph node, along with hepatosplenomegaly was described.

FDG-positron emission tomography (PET)/CT showed a mass near the pancreas with moderately increased FDG uptake. Complete surgical resection of the mass was performed, and the histopathological evaluation revealed lymphoid hyperplasia consistent with the hyaline vascular type of Castleman disease.

An immunohistochemical study excluded HHV-8 positivity. After the surgical procedure, the patient recovered completely. On follow up visits he was without fever or other symptoms suggestive of the disease, with normal laboratory findings.

CD with systemic manifestations is rare in children emphasizing the need for thorough evaluation of unexplained prolonged symptoms.

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SEVERE SKIN MANIFESTATIONS IN PATIENTS WITH HENOC SCHOLENLEIN PURPURA (HSP) IN FIVE TERTIARY CENTRES IN CROATIA BETWEEN 2009 AND 2019

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To evaluate characteristics of patients with severe skin manifestations of Henoch Schoenlein purpura (HSP) in five tertiary centres in Croatia in time period from 2009 until 2019.

Retrospective statistical chart analysis of clinical symptoms and laboratory parameters of patients with HSP was done. Severe skin manifestations were defined as haemorrhagic bullous, ulcerated or necrotic lesions.

During observed time period 583 patients (303 boys and 280 girls) were admitted to hospital due to HSP. Prevalence of severe skin manifestations was 2.41% (14 patients, CI 1.32 – 4.003%). Severe skin manifestations occurred more often in boys than in girls with a difference on a margin of statistical significance (11 boys, 3 girls, $p=0.057$). Patients with severe skin manifestations were more prone to relapses than those with milder skin forms (severe 28.57%, milder 18.66%), with most notable difference in patients with 2 relapses of disease (severe 14.29%, milder 3.17%, $p=0.029$).

We were able to isolate infectious trigger of disease more often in patients with milder form of disease than in those with more severe (milder 38.42%, severe 7.69%, $p=0.036$).

Patients with severe skin manifestations had more often gastrointestinal system affected than those with milder skin forms

(severe 70%, milder 36.03%, $p=0.043$). There was no statistically significant difference between those two groups in other organs involvement (kidneys, joints, central nervous system, lung).

Regarding distribution of skin rash, patients with severe skin manifestations more often had a rash on a trunk, with a difference on a margin of statistical significance (severe 85.71%, milder 57.19%, $p=0.051$).

There was no statistical difference between two groups in most of other clinical and laboratory parameters. Only blood leukocyte levels were near to statistical significance with higher levels in patients with severe skin manifestations ($12.1 \times 10^9/L$ (SD 10-17.62): $10.71 \times 10^9/L$ (SD 8.45-13.42), $p=0.059$).

Patients with severe skin manifestations of HSP were more prone to relapses, and had gastrointestinal symptoms more often affected than those with milder forms, but less often had infectious trigger isolated. There was no significant statistical difference in other clinical symptoms and laboratory parameters. We couldn't set predictive model for disease outcome based on these data. Small number of patients with severe manifestations and retrospective data analysis could be the reason. Perhaps better structured prospective trial would be more appropriate in that sense.

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DISLIPIDEMIA AS AN ATHEROGENIC FACTOR IN PATIENTS WITH DIFFERENT FORMS OF JUVENILE ARTHRITIS

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Close connection between autoimmune inflammation and proatherogenic lipid changes in rheumatoid arthritis has been well established, while for juvenile idiopathic arthritis (JIA) is under discussion. The aim of our work was to study the incidence and intensity of lipid disturbances in patients with different forms of JIA.

90 children with JIA 6-18 years were examined using clinical, biochemical methods, ultrasonic duplex scanning of vessels, thin-layer chromatography, bioimpedance measurement. 49 children without chronic diseases made up the control group.

Dyslipidaemia, was revealed in 48.9% of patients, mainly with systemic and polyarthritis. Most often an increase in atherogenic coefficient (AC) was noted. Average AC value in JIA patients was higher than in control group (2.9 ± 0.2 versus 2.0 ± 0.1 , $p < 0.05$) and correlated with the disease activity index according to JADAS71 ($r = 0.78$), a doctor's global assessment of the disease severity according to VAS ($r = 0.61$), the degree of joints dysfunction ($r = 0.55$), C-reactive protein ($r = 0.53$) and ESR ($r = 0.68$) level. An increase in the concentration of total cholesterol was observed in 28 (31.1%) children with JIA, commonly with a long-lasting disease, and in case of CS intake. In some JIA patients atherogenic changes were detected due to apolipoprotein A1 (ApoA1) deficiency (26.7%). Patients with JIA (maximum with

polyarthritis) showed a decrease in the content of the main fractions of polar phospholipids and an increase in the phospholipid lysoforms and free fatty acids by 8.9% 21.4% and 9% compared with the control group. This leads to an increase erythrocytes membranes microviscosity. Dyslipidaemia was combined with body composition changes. In this patients IMCT was higher than in control group (0.73 ± 0.03 versus 0.45 ± 0.02 , $p < 0.05$).

In high disease activity both systemic onset and polyarticular JIA proatherogenic lipid disturbances and vascular disorders took place.

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HENOCH-SCHONLEIN PURPURA IN PEDIATRICS: FIVE YEARS OF EXPERIENCE AT A RESEARCH AND TRAINING HOSPITAL IN ISTANBUL

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Our aim was to describe the epidemiological, clinical, laboratory characteristics of patients with HSP and to compare them existing studies.

The medical records of children with HSP, who met the Ankara criteria for HSP diagnosis, were retrospectively reviewed at the Haydarpasa Research and Training Hospital between the years 2010-2016. The epidemiological, etiological and clinical features, laboratory results were assessed.

In our study, the age of the patients ranged from 3 to 12 years (mean age 7 years). 93.3% of the patients were under the age of 10. (53.3%) were male and 21 (46.6%) were female. The male/female ratio was 1.14/1.

In our study, 24 cases (53.3%) were most seen in the winter season, followed by the autumn season with 12 cases (26.67%), there was significant clustering in the winter-autumn seasons.

In our study, 21 cases (46.67%) had symptoms of upper respiratory infection in the last few weeks, 9 (20%) had analgesic- antipyretic and/or antibiotics use. 6 cases (13.3%) described as acute gastroenteritis and/or diarrhea, and 9 patients (20%) had a high fever. In 10 (22.2%) patients, no etiological factors could be identified.

Clinical features examined; 44 (97.7%) of patients applied to the health institution with purpura, 22 (48.8%) joint pain and/or swelling, 10 (22.2%) abdominal pain and 5 (11.1%) with fever. Only 1 patient (2.2%) complained at the time of application that they could not only walk without rash.

Anemia was detected in 6 (13.3%) patients and leukocytosis in 30 (66.6%) patients. Trombocytosis was found in 24 (53.3%) patients. Erythrocyte sedimentation rate was high in 66% of patients, while CRP was positive in 73.3% of patients. Microscopic hematuria was found in 14 (31.1%) patients, proteinuria was found in 6 (13.3%) patients. Total IgA value was high in 5 patients (11%), C3 and C4 were found to be normal in all. PT/INR values were normal in all patients.

Our results are compatible with the literature, there are no significant differences in the epidemiological and clinical profile than reported elsewhere except IgA values in the studies conducted in the literature are significantly higher than in our study.

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RELEVANCE OF ADALIMUMAB SERUM CONCENTRATION AND ANTI-ADALIMUMAB ANTIBODIES IN SECONDARY LOSS OF RESPONSE IN PATIENTS WITH JUVENILE IDIOPATHIC ARTHRITIS

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Adalimumab (ADA) is fully humanized, monoclonal antibody against tumour necrosis factor (TNF- α), and is efficacious biological treatment option for patients with juvenile idiopathic arthritis (JIA). However, some patients who respond to the ADA therapy subsequently lose response to this treatment and present with relapse of JIA.

In a period of five years (2015-2020), a cohort of 19 patients with JIA, treated with ADA in Children's Hospital Zagreb, was followed. Disease activity was evaluated based on counts of joints with active disease, erythrocyte sedimentation rate (ESR) and physician global assessment of disease activity. Blood samples were collected from patients who presented with JIA relapse and suspected loss of response (LOR), as well as randomly from patients who had achieved appropriate disease control with ADA therapy as a control group. Levels of adalimumab and anti-adalimumab antibodies (AAA) were measured using enzyme-linked immunosorbent assays (ELISA) (Immundiagnostik AG, Bensheim, Germany). Use of concomitant methotrexate (MTX) therapy was noted, as well as time of AAA development from the beginning of ADA therapy.

Out of 19 patients on ADA, six presented with LOR. Out of six, positive AAA and subtherapeutic ADA concentrations were found in four patients, while in two patients ADA concentrations were subtherapeutic but AAA negative. Two of the six relapsed patients did not receive MTX at the time of serum sampling due to adverse reactions to this drug. The median time to development of relapse of JIA and proven AAA was eleven months (2–20 months) after initiation of ADA therapy. Adalimumab has so far been replaced by tocilizumab in four patients with LOR. Of the thirteen patients in the control group, two had positive AAA and subtherapy ADA concentrations, and one had only subtherapy ADA concentrations. Four of the thirteen patients in the control group did not receive MTX at the time of sampling, all four had negative AAA, and one also had a subtherapeutic concentration of ADA.

In our patients with relapsed JIA, positive AAA and subtherapeutic ADA concentrations were associated with loss of therapeutic response. However, the occurrence of AAA and subtherapeutic concentrations of ADA in the control group also indicate the need for further research in a larger number of patients, as well as the need for serial measurement and testing the ability of AAA to neutralize ADA.

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NEW CANDIDATE BIOMARKER OF NEPHRITIS RISK ASSESSMENT IN CHILDREN WITH HENOCH-SCHÖNLEIN PURPURA

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