of IgAV patients from two Croatian University Centers for pediatric rheumatology.

81 pediatric IgAV patients were included, of whom 45 were boys and 36 girls, as well as 150 age- and sex-matched healthy controls without any history of autoimmune disease. The median (range) age of IgAV patients was 6.25 (4.60-8.20) years, and among them 71.6% had joint involvement, 29.62% had gastrointestinal manifestations, while 27.16% developed nephritis. The purpuric rash which extended from lower extremities to the trunk, upper extremities and face (generalized rash) was present in 43.20% of patients and 27.16% had at least one relapse. Among the analyzed polymorphisms, only in the rs1412125 there was a deviation from the Hardy Weinberger equilibrium. There was no statistically significant association of the analyzed polymorphisms with the IgAV susceptibility, compared to healthy controls. Polymorphism rs2070600 was significantly related with the development of nephritis in IgAV, while rs1412125 was associated with gastrointestinal involvement. The IgAV patients carrying the T allele (rs2070600) of the AGER had significantly increased risk of nephritis development compared with the IgAV patients with homozygous CC genotype in dominant (OR 4.05, CI 1.09-15.03, p = 0.037) and additive genetic models (OR 3.95, CI 1.16-13.47, p = 0.049). The minor C allele (rs1412125) of the HMGB1 was found to significantly increase the risk of gastrointestinal involvement in overdominant model with an allelic odd ratio of 2.78 (CI 1.04-7.43, p = 0.04).

Although neither of analyzed HMGB1 and RAGE polymorphisms was not associated with IgAV susceptibility, our results indicated that these polymorphisms may be involved in the pathogenesis of IgAV with possible effect on different disease phenotypes.

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447 SYSTEMIC JUVENILE IDIOPATHIC ARTHRITIS WITHOUT ARTHRITIS AS THE CAUSE OF INTERMITTENT PERSISTENT FEVER IN A 7-YEAR OLD GIRL

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Background Intermittent fever along with increased inflammatory markers (CRP and ESR) most commonly is the sign of an infectious disease. Nevertheless, without an adequate response to antibiotic treatment, it should raise concern of other conditions, including several rheumatic, like Kawasaki disease, periodic fever syndromes and systemic form of juvenile idiopathic arthritis (JIA), even when no clear arthritis is present.

Case Presentation We present a case of a 7-year-old girl with a 14-month history of intermittent fever with increased inflammatory markers, along with unspecific symptoms such as hepatomegaly, polymorphous rash and migratory arthralgia. An extensive diagnostic workup excluded infectious aetiology, genetic testing did not detect pathogenic mutations. Despite the treatment with intravenous imunoglobulines and low dose glucocorticoids (GCs), the fever did not subside. Finally, extensive laboratory workup revealed increased proinflammatory cytokines IL-6 and TNF-alpha along with chronic anaemia and thrombocytosis. The systemic subtype of JIA was considered, and treatment with pulse (30mg/kg), continued with high (2mg/kg) doses of GCs was initiated with an instant resolution of symptoms. Nevertheless, after the weaning of GCs, the new exacerbation was observed and therefore tocilizumab, humanized monoclonal antibody against IL-6 receptor, was initiated.

Conclusion Systemic form of juvenile idiopathic arthritis is a heterogeneous disease dominated musculoskeletal and systemic symptoms. While the former ones are a result of similar pathophysiologiocal mechanisms as other forms of JIA, activation of the nonspecific immune response is responsible for the sytemic ones, similar to other autoinflammatory diseases. Therefore, it is possible that in some cases of systemic form of JIA, systemic inflammation is present without the musculoskeletal symptoms. With the existing classification criteria of the International League against rheumatism (ILAR) it can be difficult to diagnose and initiate appropriate therapy. Consequently it can mislead to recognize a serious complication – macrophage activation the syndrome. The recognition of the unique nature of systemic JIA in comparison to other types of JIA as well as an increased understanding of its pathogenesis, provides a better outcome and prognosis for children who often go undiagnosed with a debilitating chronic condition.

448 TREATMENT OF JUVENILE SPONDYLOARTHRITIS

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Juvenile spondyloarthritis (jSpA) represents a spectrum of inflammatory arthritis with strong HLA-B27 association and involvement of enthesis and/or axial skeleton that appears in children and young adults. ILAR classification criteria for enthesitis related arthritis subtype of juvenile idiopathic arthritis, which is undifferentiated form of jSpA, includes arthritis, enthesitis, presence of sacroiliac joint tenderness, inflammatory lumbosacral pain, HLA-B27 positivity, positive family history and acute anterior uveitis.

We present a case of 16 years old girl diagnosed with juvenile spondyloarthritis that first presented with recurrent monthly swelling of index finger at the age of 14. Symptoms progressed to low back pain in the morning which partly declined with activity. Additionally, her right knee was swollen and painful. She was examined on multiple occasions by pediatric orthopedic surgeon and in pediatric emergency department before seeing a pediatric rheumatologist. The first examination revealed sacroiliac joint tenderness with positive FABER test and abnormal modified Schober test. Family history was negative for rheumatic diseases and there were no signs of uveitis nor enthesitis, with ANA, RF and extensive laboratory workup being either negative or within reference range. However, HLA-B27 turned positive and MRI showed right sacroiliitis. NSAID was prescribed but symptoms nevertheless persisted. Thus, after the exclusion of TBC with quantiferon test, intraarticular corticosteroid injection was applied to the right knee and oral corticosteroid was introduced as