Allergology, Centre of Reference for Paediatric and Adolescent Rheumatology, Republic of Croatia, Zagreb, Croatia; 2University Hospital Centre Zagreb, Tissue Typing Centre, Clinical Department for Transfusion Medicine and Transplantation Biology, Zagreb, Croatia; 3University of Zagreb, Department of Pediatric Immunology, Rheumatology and Pulmonology, ZAGREB, Croatia; 4Children's Hospital Zagreb, Josip Juraj Strossmayer University of Osijek, Medical Faculty Osijek, Department of Pediatrics, Osijek, Croatia; 5Clinical Hospital Holy Spirit, University of Zagreb School of Medicine, Zagreb, Department of Internal Medicine, Division of Clinical Immunology. Rheumatology and Pulmonology, ZAGREB, Croatia; 6Children's Hospital Zagreb, Josip Juraj Strossmayer University of Osijek, Medical Faculty Osijek, Department of Paediatrics, Zagreb, Croatia.

Background: IgA vasculitis (IgAV) is a small vessel vasculitis occurring predominantly in childhood. Studies concerning the genetic background of IgAV have confirmed that susceptibility to the disease may be influenced by Human Leukocyte Antigens (HLA), with HLA-DRB1 gene showing a strong association with the disease. Objectives: We aimed to investigate HLA-DRB1 polymorphism among Croation patients with IgAV and to determine if there are associations with disease susceptibility and clinical heterogeneity. Methods: 123 IgAV patients, fulfilling the diagnostic EULAR/PRINTO/PRES criteria from three pediatric rheumatology centers and 202 unrelated healthy individuals were enrolled. Genomic DNA was extracted from whole peripheral blood. The HLA-DRB1 alleles were analysed using the Next Generation Sequencing (NGS) method. Results: Among 123 patients with IgAV, 68 were girls and 55 were boys with median age 6.3 (4.5-16.8) years at the time of diagnosis. All patients had purpuric rash, 75.7% had arthralgia or arthritis, 32.5% had affected gastrointestinal (GI) system, while 25.2% patients developed IgA vasculitis nephritis (IgAVN). The HLA-DRB1*12:01 allele was associated with an increased risk for IgAV (OR 4.45, 95% CI=1.17-16.95, P=0.03), while HLA-DRB1*11:01 allele was associated with an increased risk for Gl involvement in patients who developed IgAV (OR 3.29, 95% CI=1.04-10.01, P=0.03). A marginally significant (P=0.068) higher frequency of the HLA DRB1*10:01 allele in patients with GI symptoms was observed. No significant differences were found in the distribution of HLA DRB1 alleles between patients with IgAVN and those who did not develop nephritis. Conclusion: Our results demonstrated that HLA-DRB1*12:01 allele was associated with susceptibility to IgAV in the Croatian children, while HLA-DRB1*11:01 allele showed association with Gl manifestations of the disease.


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**Keywords** Autoantibodies, Adaptive immunity, Vasculitis

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