

„Badanie przebiegu klinicznego zapalenia naczyń związanego z IgA (d. plamicy Schönleina-Henocha) u dzieci i młodzieży w regionie łódzkim w latach 2015-2023.”

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Abstract

Introduction:

Immunoglobulin A vasculitis (IgAV, formerly: *HSP*, *Henoch-Schönlein purpura*) is the most common systemic vasculitis in children. The disease typically follows a mild course, although a subset of patients may experience severe or atypical manifestations. Skin involvement (purpura, petechiae) remains the dominant clinical feature, whereas gastrointestinal and renal involvement are key determinants of long-term prognosis. Data on predictors of severe disease and on differences in clinical presentation between the pre- and post-COVID-19 pandemic periods remain limited.

Objectives:

The aim of the study was to perform a retrospective epidemiological assessment of the clinical course of IgAV in children in the Lodz region. Additional objectives included the identification of triggering factors, evaluation of predictors of severe disease and the risk of IgAV-associated nephropathy, as well as a comparison of patient characteristics before and after the COVID-19 pandemic.

Material and methods:

The analysis included archived data of 160 hospitalized children diagnosed with IgAV between 2015 and 2023 across three pediatric centres in the Lodz region. Demographic data, clinical manifestations, organ involvement, laboratory parameters, imaging results, length of hospital stay, and the treatment course were assessed. Triggering factors were identified, and the frequency of IgAV-related nephropathy was evaluated. Patients were divided into pre-pandemic (2015-2020) and post-pandemic (2021-2024) groups. For the purpose of this study, a clinical severity scale for IgAV was constructed based on current EULAR/PRINTO/PRES^{9,46} and IPNA⁹³ guidelines, with the final version validated through consultations with pediatric, rheumatology and pediatric nephrology specialists involved in clinical care across the study centres.

Results:

Most patients presented with a classic IgAV phenotype dominated by cutaneous manifestations and 85% of cases had a mild or moderate disease course. A subset of children developed atypical or severe forms of the disease. Sex did not influence clinical severity. Viral upper respiratory tract infections were the most common triggering factor, whereas drug- or vaccine-induced IgAV accounted for the smallest proportion of cases. The strongest predictors of severe disease included: the number of organ systems involved, markers of systemic inflammation (elevated CRP and D-dimer), abnormalities in urinalysis, pathological findings on abdominal ultrasound, and positive fecal occult blood test. Serum IgA concentration was not confirmed as a prognostic marker.

A widespread tendency to use hemostatic agents (etamsylate) in IgAV was observed despite insufficient evidence supporting their effectiveness. Their use did not shorten hospitalization. In accordance with current IgAV management guidelines, glucocorticoids were administered to patients with the most severe disease course, who accounted for 25,7% of the study population.

IgAV-associated nephropathy occurred in 8% of patients. Older age and urinary tract infection identified as a triggering factor were associated with an increased risk of nephropathy. In this study, neither inflammatory marker levels, coagulation parameters, the interval from triggering infection, nor the duration of hospitalization significantly affected the risk of developing IgAV nephropathy.

Comparison of the pre- and post-pandemic periods revealed no differences in patient age or sex profile. However, after the COVID-19 pandemic a significantly shorter hospital stay, a lower proportion of severe disease and a reduced frequency of renal complications were observed. The post-pandemic period was also associated with significantly lower CRP levels, as well as decreased use of antibiotics and etamsylate, with no change in the frequency of glucocorticoid use. These findings are most likely attributable to changes in healthcare organization, improved diagnostic pathways and modifications in therapeutic approaches rather than to a true change in disease severity.

Conclusions:

Most children in the Łódź region were presented with a typical skin-dominant IgAV phenotype, although severe or atypical cases were also observed. Sex did not influence disease severity. Viral upper respiratory tract infections were the predominant triggering factor. The strongest

predictors of severe disease included elevated CRP and D-dimer levels, abnormalities in urinalysis, pathological abdominal ultrasound findings, and positive fecal occult blood testing. Serum IgA concentration had no prognostic significance. The COVID-19 pandemic did not notably affect the demographic profile or overall disease characteristics but was associated with a milder clinical course, shorter hospitalization time and changes in therapeutic practices. Symptomatic treatment with limited evidence of efficacy, such as etamsylate, were commonly used and did not reduced hospitalization duration or disease severity. IgAV- associated nephropathy remains a relatively rare but clinically important complication requiring further follow-up, whereas older age and urinary tract infections appear to be potential risk factors for its development.