CLINICAL AND IMMUNOLOGICAL FEATURES OF THE COMORBID COURSE OF NEPHROTIC SYNDROME AND ALPORT SYNDROME IN CHILDREN

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Introduction. To date, Alport syndrome (SA) is diagnosed on all continents of the world. The frequency of the disease is much higher than that described in the literature, and a key role in the origin of the pathology is played by gene mutation due to different penetrance and expressivity. In Uzbekistan, the frequency of SA is about 6-8% of the total number of patients with kidney pathology, and one of the reasons is the impact of adverse environmental factors and infection during the first trimester of pregnancy in the mother, as well as consanguineous family marriage. At the same time, the progression of steroid-dependent and steroid-resistant forms of nephrotic syndrome (NS) in children leads to the development of minimal change disease in 85% of cases and up to 5-7% of focal segmental glomerulosclerosis.

The aim of the study was to study the clinical and immunological features of the comorbid course of nephrotic syndrome and Alport syndrome in children. **Material and methods.** A clinical and immunological examination of 102 children aged from 3 to 18 years old, who were under our supervision in the nephrology department of the ODMPMC of the city of Andijan, was carried out for 2012-2022. with glomerular pathologies of the kidneys and identified a contingent of patients with nephrotic syndrome (NS) and SA. Of these: NS-40, NS + SA-16, SA-14.orbid course of nephrotic syndrome and Alport syndrome in children.

Results. The distribution of children by age and sex showed that NS and SA are more common in boys than in girls (3:1) aged 3 to 18 years. In children with NS+SA, a large percentage is acute and chronic pyelonephritis, glomerulonephritis and metabolic nephropathy ($P<0.001$). When studying the incidence in the mother, it was found that a large percentage is kidney pathology, pregnancy toxicosis, complicated childbirth, cardiovascular and endocrine pathology when compared with the control group ($P<0.001$-0.01). Of the clinical symptoms of AS, there were

high signs of intoxication, arterial hypotension, urinary syndrome, stigmas of renal dysembryogenesis, visual and hearing impairment (P<0.001). Among the phenotypic stigmas of dysembryogenesis, a large percentage of a predominantly flattened occiput, pronounced superciliary ridges, hypertelorism, epicanthus, high gothic palate, anomalies in the position of the ears, a sandal gap between 1-2 fingers and toes, nipple hypertelorism, chest deformity, and clinodactyly were noted. The frequency of detection of dysembryogenesis stigma in HC+SA was a higher percentage with hearing loss than in children without hearing loss (P<0.01). According to the characteristics of the urinary syndrome in all groups, a large percentage was proteinuria, hematuria and crystalluria (P<0.001).

According to the results of the analysis of immunity parameters, it was found that in children with NS, NS + SA, compared with the control group, a significant increase in ASL of the kidneys and ASL of the lungs was recorded (P<0.001-0.01). Production of IL-2 in all groups was significantly reduced compared to the control group (P<0.001-0.01). In children of group II (HC+CA), the level of IL-2 compared with group I (HC) and group III (CA) was significantly low (P<0.001). Comparative evaluation of the results of the study of the indices of C3, C4 components with the control group showed a significant decrease (P<0.001-0.01), which in the II group were also pronounced (1.5 times) than in the I and III groups of children (P<0.001-0.01).

Conclusions. 1. Alport syndrome is more common in boys than in girls, the ratio of which is 3:1 and at the age of 11-18 years. 2. The frequency of detection of nephrotic syndrome with Alport's syndrome varies by year and depends on the age and pathology of the mother during pregnancy, consanguineous marriage, as well as on concomitant diseases of the child, such as acute and chronic glomerulonephritis, pyelonephritis, metabolic nephropathy. 3. In Alport syndrome, such stigmas of dysembryogenesis as a flattened occiput, pronounced superciliary arches, hypertelorism, epicanthus, high gothic palate, anomalies in the location of the ears, a sandal-like gap between 1-2 fingers of the hands and feet, hypertelorism of the eyes and nipples, deformity of the chest and clinodactyly. 4. In the comorbid course of Alport's syndrome with nephrotic syndrome, there is a significant increase in ASL of the kidneys and ASL of the lungs, a decrease in the production of IL-2 and indicators of C3, C4 complement components that are pronounced (1.5 times) than in children with Alport's syndrome without nephrotic syndrome, which requires the use of adequate methods of immunocorrection in the treatment of such patients.