

LATE IMMUNE STATUS COMPLICATED WITH ALPORT SYNDROME IN CHILDREN

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Relevance. Today, the frequency of Alport syndrome among the world's population is 1:5000. Among them, chronic renal failure (CRF) accounts for 1% of all kidney pathologies in the European population, and 2.3% of patients undergo kidney transplantation. Alport syndrome occurs in all continents and nations of the world. It is noted that the frequency of the disease is higher than the information given in the literature.

The purpose of the study is to study the state of immune parameters in children with Alport syndrome complications.

Material and methods. 102 children aged 3 to 18 years under our control in the nephrology department of the Andijan Children's Clinic in 2012-2023 underwent a clinical and immunological examination with glomerular kidney pathologies and Alport syndrome (AS) and nephrotic syndrome (NS) the contingent of patients with Among them: AS-14, NS-35, NS+AS-16.

Results. Distribution of children by age and gender showed that between 3 and 18 years of age, NS and AS were more common in boys than in girls (3:1). When maternal morbidity was studied, compared to the control group, it was found that a large percentage of complicated childbirth, pregnancy toxicosis, kidney pathology, cardiovascular, endocrine pathologies, as well as consanguineous marriages were made ($P < 0.001$). Complication of AS with NS was noted more in children with acute and chronic glomerulonephritis, pyelonephritis and metabolic nephropathy ($P < 0.001-0.01$). The clinical signs of AS include stigma of dysembryogenesis (hypertelorism of nipples, anomalies in the location of ears, bulging brow ridge,

epicant, high, Gothic palate, sandal-shaped space between 1-2 fingers, syndactyly), arterial hypotension, urinary syndrome. , signs of intoxication, visual and hearing impairment ($P < 0.001-0.01$). According to the characteristics of urinary syndrome, proteinuria, hematuria and crystalluria accounted for a significant percentage ($P < 0.001-0.01$) in all groups. According to the results of the analysis of immune parameters, in children with NS, NS+AS, compared to the control group, kidney antigen-binding lymphocytes (ABL), lung ABL ($P < 0.001-0.01$) and interleukin- 2 (IL-2) level was found to be significantly increased ($P < 0.001$). Compared with the control group, the results of the study of the indicators of complement components C3, C4 showed a significant decrease, which was found stronger in group II (AS + NS) (1.5 times) than in groups I and III ($P < 0.001- 0, 01$).

Conclusions:

1. The increased frequency of complications of Alport's syndrome with nephrotic syndrome is fundamentally related to the pathology of the mother during pregnancy, consanguineous marriage, glomerulonephritis, pyelonephritis and metabolic nephropathies.

2. When Alport syndrome is complicated by nephrotic syndrome, immunopathological changes are characterized by an increase in the production of kidney ABL, lung ABL and IL-2, as well as a decrease in the level of complement components C3, C4, and immunogenetic diagnosis is a criterion for such patients is counted.