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A CASE PRESENTATION OF A CHILD WITH NEPHROTIC SYNDROME AND COMMON VARIABLE IMMUNODEFICIENCY

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We report a patient with genetically confirmed common variable immunodeficiency (CVID) and nephrotic syndrome (NS), one of the most common glomerulopathies in children. Idiopathic NS affects 1 to 3 per 100 000 children, < 16 years of age. Whereas most of them respond well to corticosteroid treatment, but as many as 20% experience a complicated course with steroid resistance.

Aim: Was to review the case of a child with NS and CVID followed in paediatric department of Vinnytsya Regional Children's Hospital.

Methods: We have a follow up of the patient with NS and CVID, determined clinically, laboratory, genetically in a Vinnytsya Regional Children's Hospital and Kyiv Children's Hospital №1.

Results: A 9-year-old boy presented with severe generalized oedema, weight gain. The results of laboratory tests revealed normal creatinine, hypoproteinemia, hypoalbuminaemia, and nephrotic range proteinuria (10 g/day). A diagnosis of idiopathic NS was made, and steroid treatment (prednisolone 60 mg/day) was started. After 4 weeks of therapy proteinuria decreased but it did not become negative. Resistant oedema also was present for a long time. H`e was treated with 3 high-pulse doses of methylprednisolone followed by steroid therapy. During conformation period he didn't show steroid sensitivity and steroid-resistant NS was confirmed. We couldn't perform nephrobiopsy or genetic testing, so therapy with cyclophosphamide was started. He achieved partial remission and even 1 year after finishing immunosuppressive therapy he presents with mild isolated proteinuria, and using of enalapril continues. In his past medical history there were recurrent infections, pneumonias, fungal infection of skin, episode of herpes zoster and signs of physical retardation. During treatment of NS, we also met frequent sinopulmonary infections and the consultation with immunologist was performed. Immunological tests showed reduction in serum concentrations of immunoglobulin (Ig)G, in combination with low levels of IgA and IgM, poor response to immunizations (Tetanus anatoxin) and heterozygouse mutation TNFRSF13B gene. With the history of recurrent infections and low Ig levels, he was diagnosed as having CVID. Replacement therapy with IVIG (600 mg/kg every 3 weeks) was initiated.

Conclusions: CVID can be observed in NS in children. NS in children with CVID appears to be complicated and difficult to treat with corticosteroids alone. Further research is needed to understand whether CVID has a prognostic value in children with NS.