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HEALTH-RELATED QUALITY OF LIFE IN CHILDREN WITH PRIMARY IMMUNODEFICIENCY

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Background:

Improved diagnostic and treatment of PID led to an increasing the number of patients who survive and reach adult age. This raises the problem of minimizing the limitations caused by the disease. **Aim**: to study health-related quality of life (HRQL) in children with PID.

Methods:

Evaluation of the parameters of HRQL was performed in 67 children with different PID compared with 76 healthy children aged 8-18 years using the pediatric questionnaire PedsQL^{tm}4.0 Generic Core Scale.

Results:

Total score of HRQL in PID patients differs significantly from healthy controls (26.25±1.75 vs 15.61±1.01, respectively, $p<0.05$), as well as all categories of HRQL. In all groups of PID the quality of life is mostly affected by the domain of physical activity. In CID with syndromal features and autoinflammatory diseases a significant contribution to the impaired quality of life is made by emotional component. All components of HRQL are significantly better in patients receiving regular treatment and having good treatment compliance. Medium strength feedback between the total quality of life and duration of continuous replacement therapy was revealed in patients with antibody deficiencies ($r=-0.45$, $p<0.05$), while there was no detected relationship between quality of life and the dose of IVIG as well as the intervals between administrations.

Conclusion:

PID adversely affects the quality of life of children, limiting their physical capabilities and social activity. The greatest impact on quality of life is caused by treatment regularity and its continuity. Forming the treatment compliance is the reserve to improve quality of life.