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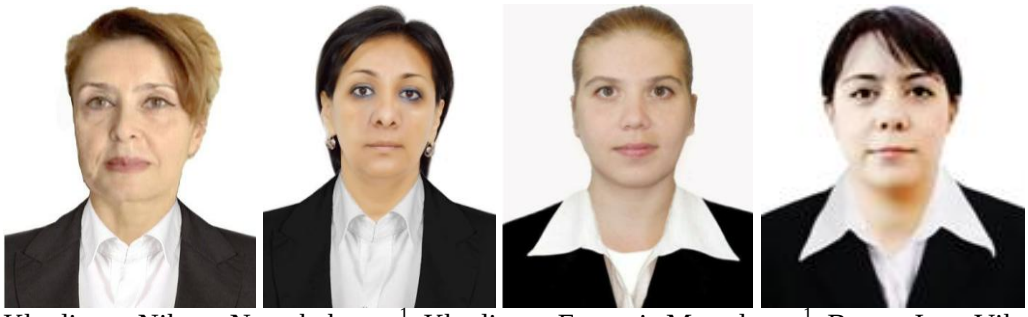
# CLINICAL AND LABORATORY FEATURES OF PATIENTS WITH THALASSEMIA



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## ТАЛАССЕМИЯ БИЛАН ОҒРИГАН БЕМОРЛАРДА КЛИНИК ВА ЛАБОРАТОР ХУСУСИЯТЛАРИНИНГ ЎЗАРО БОҒЛИҚЛИГИ

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## ВЗАИМОСВЯЗЬ КЛИНИЧЕСКИХ И ЛАБОРАТОРНЫХ ХАРАКТЕРИСТИК У ПАЦИЕНТОВ С ТАЛАССЕМИЯМИ

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***Резюме.*** *Ушбу тадқиқот ишида талассемия билан оғриган 127 нафар беморнинг клиник ва иммунологик кўрсаткичлари таҳлили ўтказилди. Тадқиқотлар натижасида талассемиянинг патогенетик таркибий қисмларининг ўзи (гемик гипоксия, гиперсидеремия) ҳужайра ва гуморал иммунитетнинг пасайишига олиб келиши ва касалланиш даражасининг ошишига ёрдам бериши аниқланди.*

***Калит сўзлар****: гемолитик анемия, талассемия, ҳужайрали ва гуморал иммунитет.*

***Abstract.*** *In this work, the analysis of clinical and immunological parameters of 127 patients with thalassemia was carried out. The study revealed that the very pathogenetic components of thalassemia (hemic hypoxia, hypersideremia) lead to a decrease in cellular and humoral immunity and contribute to an increased incidence of the disease.*

***Keywords****: hemolytic anemia, thalassemia, cellular and humoral immunity.*

**Introduction.** Hereditary hemolytic anemia (HHA), according to the children's hematology department of the NMC "Shifobakhsh", ranks second in the incidence of anemia in the Republic of Tajikistan. According to the WHO, the proportion of hemolytic anemias (HA) among other blood diseases is 5.3%, and among anemic conditions - 11.5% [4, 5].

Among all hemolytic anemias, the most common is hemoglobinopathy (thalassemia), which is accompanied by frequent intercurrent diseases leading to early disability in this group of patients [1, 2, 3].

**Purpose.** To identify the features of clinical and immunological parameters in children with thalassemia.

**Materials and methods**. In the children's hematology department of the NMC, 196 patients with HHA got treatment from the beginning of 2022 (127 patients suffering from thalassemia aged 3 months to 16 years). During the treatment all patients underwent a complex of lab tests (complete blood count, bilirubin, serum iron, ferritin, hemoglobin forrez, and myelogram). ELISA was performed to determine cellular and humoral immunity.

**Research results.** The vast majority of patients - 99 (78.2%) suffered from β-thalassemia (hemoglobinopathy), 25 (19.4%) - fermentopathy and 3 (2.4%) patients - microspherocytic anemia.

Thalassemia major was characterized by the following symptoms: regressing anemia with normoblastosis, earthy-icteric coloration of the skin of varying severity, hepatosplenomegaly, hyperbilirubinemia and urobilinuria, osteoporosis, causing peculiar changes in the bones of the face and cranial vault and a sharp lag in sexual and physical development from their peers.

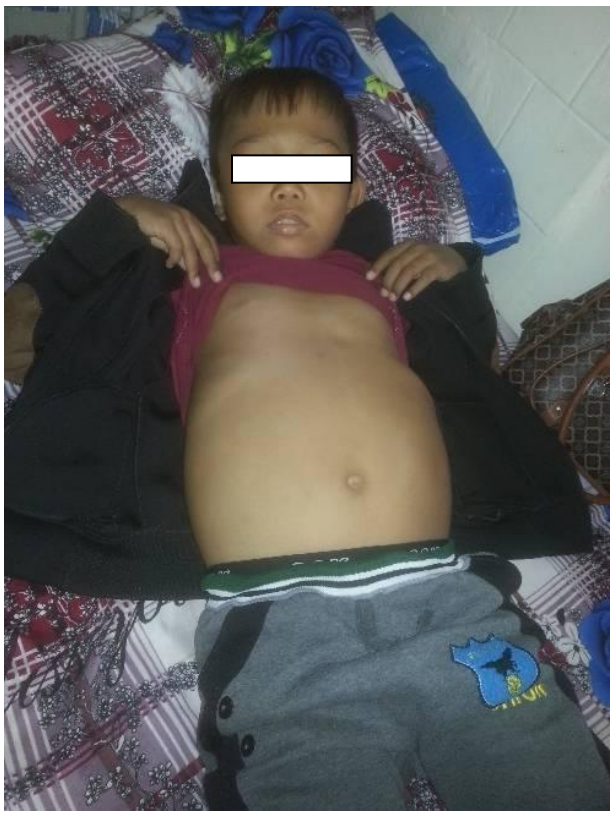
Depending on the severity of the condition, the patients were divided into the three groups: The first group consisted of 8 patients with mild anemia. Pale skin with a waxy tint, lethargy, moderate hepatosplenomegaly (enlargement of the spleen up to 1.5-2 cm from under the edge of the costal arch), changes in the skeletal system in the form of a tendency to form a towering skull were clinically noted.

The second group included 29 children with moderate anemia. Pale icteric skin with an earthy tint, skull deformity, an increase in the upper jaw and a Gothic palate, retraction of the bridge of the nose, hepatosplenomegaly (enlargement of the spleen up to 3-5 cm) were clinically noted.

The third group consisted of 62 patients with severe anemia. Objectively, the children had icteric skin with an earthy tint and icterus of the mucous membranes. The deformity of the skull, retraction of the bridge of the nose, the distance of the orbits and the Mongoloid incision of the eyes, the protrusion of the cheekbones, incisors and canines with malocclusion, anomalies of the teeth are pronounced. The size of the liver and spleen reached the level of the small pelvis.

We examined 99 patients with thalassemia to study the state of specific immunity. In patients with thalassemia of the first group, the indices of T-cells and their subpopulations did not differ from the norm, but tended to decrease.

In patients with moderate anemia, compared with the group of healthy children, there was a marked decrease in T-lymphocytes with various immunological markers (CD4, CD8, CD16) ($p<0.001$). Comparison of the average values of T-lymphocyte subpopulations with CD4, CD8 and CD16 markers in this group of patients with those of patients with mild anemia also revealed significant differences ($p<0.001$).



**Pic. 1.** A 15-year-old boy with thalassemia

**Pic. 2.** A 8-years-old girl with thalassemia

In the group of patients with severe anemia, compared with the group of healthy children, there was a significant decrease in T-cells and their subpopulations ($p<0.001$). When comparing the average values of T-lymphocytes with CD4, CD8 and CD16 receptors in patients of this group with the corresponding indicators of children with mild and moderate anemia, a highly significant difference was revealed.

Thus, the study of cellular immunity revealed a change in its parameters in all groups of patients with thalassemia in the form of a decrease in the content of both absolute and relative indicators of T-lymphocytes and their subpopulations in peripheral blood, which was also a statement that the genetically determined anemic process contributed to negative change in the population of immunocompetent cells in the hematopoietic system in this category of patients.

The average content of IgA, M, G in patients with mild anemia in the blood serum practically did not differ from those in the control group.

At the same time, in patients with moderate anemia, a significant decrease in the average levels of IgA, M in blood serum was found compared with the same indicators in the group of healthy children, while the average IgG level did not differ from the norm, but tended to decrease.

In the group of patients with severe anemia, there was a significant decrease in the average values of Ig A, M, G compared to those in the control group.

Comparison of average values of IgA, M, G in the blood of patients with severe thalassemia with corresponding indices of children with light and moderate-severe thalassemia revealed a significant decrease in their concentration ($p<0,001$).

Thus, our results indicated that children with moderate and severe anemia had a reduced number of B-lymphocyte cells (CD20 - 14.0+0.69; 11.8+1.31, respectively), which was certainly the cause of insufficient humoral response.

**Conclusion.** The study of cellular and humoral immunity revealed a change in its parameters in all groups of patients with thalassemia in the form of a decrease in both absolute and relative parameters of T-lymphocytes and their subpopulations in peripheral blood. In addition, children with moderate and severe anemia had a reduced number of B-lymphocyte cells (CD20 - 14.0+0.69; 11.8+1.31, respectively), which was certainly the cause of the insufficient humoral response and a statement that a genetically determined anemic process contributed to the adverse changes in the population of immunocompetent cells in the hemopoietic system in this category of patients.

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## *ВЗАИМОСВЯЗЬ КЛИНИЧЕСКИХ И ЛАБОРАТОРНЫХ ХАРАКТЕРИСТИК У ПАЦИЕНТОВ С ТАЛАССЕМИЯМИ*


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***Резюме.*** *В данной работе был проведен анализ клинико-иммунологических показателей 127 пациентов с талассемиями. В результате исследования было выявлено, что сами патогенетические компоненты талассемии (гемическая гипоксия, гиперсидеремия) приводят к снижению клеточного и гуморального иммунитета и способствуют увеличению частоты заболеваемости.*



***Ключевые слова:*** *гемолитическая анемия, талассемия, клеточный, гуморальный иммунитет.*