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FEATURES OF GLUTEN ENTEROPATHY IN CHILDREN

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Abstract: Guy-Herter-Geibner disease (gluten enteropathy) - described by British physician S. J. Gee, 1839-1911, American physician and pharmacologist Ch. A. Herter, 1865-1910, German pediatrician O. J. L. Heubner, 1843-1926; synonyms – celiac disease, gluten-sensitive enteropathy, idiopathic steatorrhea, non–tropical sprue) - hereditary disease with the classic syndrome of impaired intestinal absorption is malabsorption.
Keywords: gluten enteropathy, gluten, intestines, food, diet, fermentopathy, children.

Guy-Herter-Geibner disease (gluten enteropathy) is characterized by atrophy of the mucous membrane of the small intestine due to congenital intolerance to gluten - the gluten protein of cereals and, in particular, one of the main components of gluten – gliadin. The disease manifests itself after the introduction of gliadin-containing products (semolina, milk mixtures, bread, etc.) into the child's diet.

It is manifested by a violation of absorption, which is caused by gluten intolerance. Genetic factors play an important role in the development of the disease; immune mechanisms and fermentopathy are also assumed to be involved.

Shortening of villi, expansion of crypts is detected in the intestinal biopsy. Both clinical and morphological signs of the disease disappear against the background of a gluten-free diet and resume after eating gluten-containing foods. Symptoms can appear at any age, but most often — at 9-18 months. With artificial feeding, the disease manifests itself earlier.

Clinical manifestations of gluten enteropathy in children in the acute period are well known, while the pathogenesis, polymorphism of the clinical picture in the long-term period, the relationship with the dietary factor still remain a little-studied area. In the literature, there are only isolated data on prospective studies of children with gluten enteropathy devoted to the assessment of clinical manifestations, the dynamics of morphological and immunological parameters, depending on the severity of compliance with a gluten-free diet [1,2].

Further progress in the treatment of patients with gluten enteropathy is impossible without a clear understanding of the mechanisms of long-term pathophysiological processes caused by the action of gluten.

The main method of treatment and prevention of recurrence of gluten enteropathy is strict, lifelong adherence to a gluten-free diet. The Guy-Herter-Geibner disease does not disappear with age and against the background of treatment, although clinical symptoms may become less pronounced if the diet is not followed [3,4].

There are two main "key" points in pathogenetic therapy - the composition and nature of the gluten-free diet. Although labeling a diet as gluten-free should imply that the diet is gluten-free, this is not always the case. In some countries (USA, Canada, Russia), gluten enteropathy is based on foods that completely exclude gluten. In others (Scandinavia, UK), gluten enteropathy may include foods (e.g. wheat starch) that contain small amounts of prolamins. The results of experimental and clinical trials of oat toxicity in patients with gluten enteropathy are ambiguous [5]. Most international experts, however, prohibit the use of oats. To date, there is no clear scientific data on the amount of prolamins that can be harmless to patients with gluten enteropathy, therefore, in Russia, the use of oats by patients is prohibited [6].

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There is a fairly large list of products that include the "hidden" gluten. In accordance with international standards, based on the FAO/WHO Codex Alimentarius, gluten-free products can be classified when the gluten level in them is below 10 mg per 1 kg of product; in EU countries - 200 mg/kg of product in terms of a dry sample [7]. However, even in specialized gluten-free products, the gluten content may be higher than these values, which is associated with contamination on production lines [8].

There are a number of socio-psychological difficulties in the lifelong observance of a gluten-free diet in the Republic:

- traditionally, the widespread use in our country of products from wheat and other cereals, which are the basis of the diet of uzbeks;

- the inability to organize proper nutrition for a sick child in children's institutions - kindergarten, school, sanatorium, which dictates the need to create specialized groups in kindergartens of large cities and specialized sanatoriums for schoolchildren and adolescents;

- the use of industrial products in nutrition, the packaging of which does not list the ingredients that are fully included in their composition, that is, there are impurities of "hidden" gluten.

Patients with celiac disease may, for one reason or another, refuse to follow a strict gluten-free diet and switch to a regular diet. In this case, not everyone and not so quickly returns to the clinical manifestations of the disease and morphological changes in the mucous membrane of the small intestine, characteristic of the initial period of gluten enteropathy. So, in the experimental works of American scientists, it was shown that in children with celiac disease after the introduction of gluten (at the rate of 0.2-4.2 g / day. 0.02-0.26 g/kg)) the appearance of clinical symptoms was observed after 5-51 weeks from the moment of loading [9].

Thus, there are still many issues in the pathogenesis, diagnosis and dietary treatment of Guy-Herter-Geibner disease in children, the solution of which will contribute to improving the quality of life and prognosis of patients, as well as the prevention of a wide range of chronic autoimmune and oncological diseases in adults [10,11].

The purpose of the study: was to determine the effectiveness of a gluten-free diet for children with an established diagnosis of Guy-Herter-Geibner disease (gluten enteropathy, celiac disease).

Guy-Herter-Geibner disease surveyed on the basis of a specially compiled questionnaire map. The questionnaire took into account: the level of education of parents, the region of residence and the availability of gluten–free products, the awareness of the next of kin, especially the treatment of patients with Guy-Herter-Geibner disease. The parents of 76 patients took part in the survey.

Results and discussion: The parents of 76 patients took part in the survey. Of these, 64 (84,0%) children were with a typical form and 12 (16,0%) with an atypical form of gluten enteropathy. In turn, out of 64 patients with typical gluten enteropathy, there were 48 children (75,0%) with non-refractory form, 16 children (25,0%) with refractory form. According to the parents, due to the survey data, patients with typical non-refractory gluten enteropathy are hospitalized every 6 months. 23 (48,0%), with typical refractory 10 (62,3%) and atypical 6 (50%) patients, which is 51,3% of all patient groups. Every 3 months, 14.5% of patients with gluten enteropathy from the studied group are hospitalized.

Thus, the remaining 34,2% are not hospitalized on time, that is, they do not receive the necessary treatment. Recommendations for home are partially fulfilled by 45% of patients, 24% of patients do not fulfill and only 31% of patients strictly adhere to the recommendations. Most of the patients are observed by first-level medical workers. During these visits, health care workers may emphasize the benefits of adherence to home recommendations and a strict gluten-free diet throughout life. 33% of patients are observed by the district doctor, the rest are treated only for the purpose of prolonging disability. The control of medication intake and diet is mainly carried out by the parents of children. The survey data showed that 49% of patients strictly follow the diet.

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The main method of treating gluten enteropathy is a lifelong gluten-free diet, which leads to remission in most individuals. Children with an established diagnosis of gluten enteropathy strictly follow a gluten-free diet in 49% of cases, follow a diet with disorders, eat foods containing "hidden" gluten in 32% and do not follow a gluten-free diet in 13% of cases, which makes their prognosis pessimistic. With the violation of the gluten-free diet, an increase in the number of patients with stunted growth and reduced body weight was noted. Here it is necessary to take into account that some parents, without knowing it, violate the diet due to ignorance of foods containing hidden gluten.

Knowledge of the origin of gluten enteropathy combined with experience in identifying gluten-containing products improves the quality of self-treatment. 33% of mothers and grandmothers who cook food for a sick child answered that they do not know recipes for dietary dishes, 51% of parents do not know all products containing hidden gluten.

The duration of strict adherence to gluten-free diet for 5 years was observed in 32% of children, more than 5 years in 68% of children. Mostly children go to school (68%), of which 31% take dietary food from home. The gluten-free diet is observed more strictly by children with early-onset and typically occurring gluten enteropathy, compared with patients with late-onset celiac disease. Younger children follow a gluten-free diet better than teenagers.

Strict adherence to the diet with the timely start of treatment and high-quality care for a sick child allows for the normal development of the child. According to the survey, 20% of mothers have an 8-hour working day, 18% have a 4-hour working day, this indicates a decrease in quality care for a sick child. Perhaps this also affects the dynamics of the course of the disease of patients.

Currently, it is considered proven that in case of Guy-Herter-Geibner disease, lifelong adherence to a diet is required, since departure from it is not only fraught with a possible exacerbation of the process, but significantly increases the risk of developing malignant neoplasms, including intestinal lymphomas.

Diseases of the digestive system, deep metabolic changes (pronounced lag in physical development, deep anemia, hypopolivitaminosis, hypocalcemia, myocardiodystrophy) are widespread in 68% of children with the refractory form of Guy-Herter-Geibner disease, regardless of the severity of the gluten-free diet.

Our observations show that in case of following strict diet and adequate supplemental therapy, children with gluten enteropathy do not lag behind their peers either in physical or mental development.

Judging by the responses of parents after treatment and diet, the condition improved in 81% of children with the non–refractory form of Guy-Herter-Geibner disease. In the group with refractory gluten enteropathy, the condition did not improve in 2/3 of patients. We believe that this may be due to non-compliance with recommendations at home, violation with diet, ignorance of gluten-containing products, irregular patronage by district doctors of polyclinics (survey data).

Thus, patients are subject to constant dispensary supervision with annual examination and examination at least 2 times a year.

A lifelong strict gluten-free diet is the key to normalizing the structure and functions of the small intestine, eliminating metabolic disorders, ensuring normal rates of physical, mental and sexual development of the child.

It is necessary to increase the alertness and education of parents regarding this disease.

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