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SHORT BOWEL SYNDROM IN CHILDREN

Short bowel syndrome (SBS) is a symptom complex caused by the absence of most of the small intestine with the development of mal digestion and malabsorption. The aim of the study is to describe our own clinical case of a child with SBS who achieved enteral autonomy without the use of a synthetic analogue of glucagon-like peptide-2.The presented clinical case shows that as a result of timely initiation of treatment, it was possible to stabilize the boy's condition and transfer him to enteral feeding without the use of a synthetic analogue of glucagon-like peptide-2.

Keywords: short bowel syndrome; parenteral nutrition; children; teduglutide.

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Introduction. Short bowel syndrome (SBS) is a symptom complex caused by the absence of most of the small intestine with the development of mal digestion and malabsorption [8]. In the Russian Federation, a registry of children with SBS has been created; in 2020, it included 220 patients [5].

The aim of the study is to describe our own clinical case of a child with SBS who achieved enteral autonomy without the use of a synthetic analogue of glucagon-like peptide-2.

Pathological physiology. The most common causes of SBS in children are congenital and perinatal diseases: necrotizing enteral colitis (NEC), middle gut incorrect rotation, gastroschisis and intestinal atresia, cytomegalovirus infection [16]. NEC is the basis for almost 30% of all cases of SBS. In most cases, patients are forced to be on parenteral nutrition (PN), which leads to various complications from various organs and systems [14]. After bowel resection, the body responds with a process called intestinal adaptation, which consists of adaptive changes in the preserved portion of the intestine; in children, this process begins soon after bowel loss and continues for several years [6, 15]. The first period is characterized by diarrhea with massive loss of fluid and electrolytes, which requires parenteral administration of these substances [8]. Later, epithelial hyperplasia occurs [17]. Patients with colon resection are at significant risk of dehydration [10]. Most sodium and water are absorbed through the colon, but nutrients with fermented carbohydrates are also absorbed. Patients with SBS can receive up to 50% of their nutritional needs through the colon [2, 9].

Clinic. The clinic is manifested by a violation of physical, psychic development, water-electrolyte and acid-base balance due to insufficient intake of micro- and macronutrients. The main signs of SBS include the following: diarrhea, abdominal pain, bloating, weight loss, dehydration, steatorrhea, edema, weakness, drowsiness, anemia, vitamin deficiency.

Treatment and prognosis. After the stage of surgical interventions, children are transferred to PN, first in the hospital, then at home. Survival after extensive bowel resections after 3 years is 87-89%, but most patients are forced to receive PN. The further principle of treatment is to achieve complete independence from PN. For this purpose, the use of a synthetic analogue of glucagon-like peptide-2 (GLP-2) is recommended [13]. GLP-2 is an intestinal growth factor produced by L-cells of the ileum and distal colon [10]. A synthetic analogue of GLP-2, teduglutide (T) increases the size of villi and the depth of crypts in the intestinal epithelium. In 2021, T was officially registered in our country and approved for use in children with SBS over 1-year-old [3]. It has been established that T allows to reduce PN, infusion time and even achieve a complete transition to enteral autonomy [7]. The ileum is more capable of increasing the surface area of the villi, the height of the villi and the depth of the crypts, as well as developing processes of increasing the length, diameter and motor function [11]. This part of the intestine, compared to the jejunum, specializes in the absorption of vitamin B12, bile acids, fluid and is able to effectively increase its absorption capacity [4].

Clinical case. Boy R., born on November 24, 2021 from the 3rd pregnancy, 2 births. The pregnancy proceeded against the background of chronic viral hepatitis C, chronic cervicitis, mild iron deficiency anemia, at 31 weeks' mild acute respiratory viral infection. Surgical delivery at a gestational age of 40 weeks. The reasons for the cesarean section were premature detachment of a normally located placenta of a severe degree without signs of external bleeding and acute fetal distress.

Physical development at birth: weight 2850 grams, length 50 cm, chest circumference 33 cm, head 34 cm. Apgar score: 1-3 points. The condition is severe due to respiratory disorders against the background of asphyxia, since birth he was on intravenous pulmonary bypass. In the neurological status, the syndrome of central nervous system depression, responded to the examination with tonic convulsions.

On the 3rd day of life, the dynamics are negative due to the abdominal syndrome (intestinal paresis of 2-3 degrees, gastric bleeding), hemodynamic disorders, and increasing respiratory failure. According to radiography, the presence of free air in the abdominal and pleural cavities were detected. A diagnosis of NEC stage 3B, peritonitis was made. After preoperative preparation, an operation was performed - drainage of the abdominal cavity in both iliac regions. The condition in the postoperative period was extremely severe.

After relative stabilization, a repeat operation was performed: laparotomy, revision of the intestine, stomach, total resection of the jejunum, subtotal resection of the colon, duodenal-ileum anastomosis "end to end", end ileostomy, sanitation, drainage of the abdominal cavity. During

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the operation, transfusion of filtered red blood cells (FRBC) was performed. In the postoperative period, the condition remained extremely severe for a long time, unstable, massive intensive therapy was carried out, the child suffered severe multiple organ failure. On the 12th day, enteral loading with water was started, from the 14th day - with the formula "Nutrilon pepti gastro" with a gradual expansion of the volume. At the age of 1 month 20 days, for further treatment, nursing of the child in the postoperative period, he was admitted to the early childhood department. The condition upon admission was severe due to intoxication, severe malabsorption syndrome, gross neurological symptoms, anemia, metabolic and trophic disorders. For a long time, the child's condition remained unstable, severe, the loss of intestinal contents through the stoma progressed. The dynamics showed an increase in intoxication, a sharp increase in acute phase proteins, an increase in leukocytosis in the blood, a decrease in the hemoglobin level to severe anemia, which required a FRBC transfusion, continuous infusion therapy for the purpose of partial PN. Against the background of intensive therapy, it was possible to expand nutrition to 2/3 of the volume of the physiological need, relieve the intoxication syndrome; positive dynamics were achieved in weight, a tendency towards an increase in the hemoglobin level was noted.

Considering the relative stabilization of the condition, the child was prepared for reconstructive surgical correction of the stoma. After preoperative preparation, on 03/10/2022, a reconstructive operation was performed: laparotomy, intestinal lysis, reconstructive resection of the ileum, hardware ileum connection "side to side". In the postoperative period, he was in the anesthesiology and intensive care department, received PP, pain relief, antibacterial therapy, and a FRBC transfusion was performed due to severe anemia. After relative stabilization of the condition, on March 17, 2022, he was again transferred to the early childhood department for further treatment and care. The condition in the postoperative period remained severe for a long time. The volume of nutrition was gradually expanded to the physiological need, which the child began to assimilate, positive weight dynamics were outlined. Cholestasis syndrome persisted for a long time, and was relieved by taking liver protectors during treatment. Neurological symptoms in the form of movement disorder syndrome persist. Therapy and nutrition were constantly corrected, the condition gradually stabilized. The child did not receive T, since in the Russian Federation the drug was registered only in the year of the patient's birth and was approved for use after 1 year, and he had not yet reached this age. The parents refused to raise the boy, and he was transferred to the Orphanage. Upon reaching the age of 1 year, from which the use of T is permitted, it was decided to abandon the use of the drug, since the child had already achieved enteral autonomy without it.

Currently, the patient is 3 years old. He is on complete enteral nutrition, gradually expanding his diet. Medications constantly receive enzymes, amino acid formula, iron preparations. Moderate delay in physical and psychic development persists. He continues to be raised in the Orphanage.

Conclusion. A distinctive feature of this case is that as a result of timely initiation of treatment, it was possible to stabilize the boy's condition, transfer to enteral feeding without the use of a synthetic analogue of glucagon-like peptide-2.

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