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Modern Pharmacotherapy of the Patients with Short Bowel Syndrome. Regional Experience in Patients Management

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Abstract

Introduction: Short bowel syndrome (SBS) is a rare disease, however, the awareness of current trends in SBS patients treatment approaches, including the example of pediatric patients management in the Tyumen region of Russia, is of significant scientific and medical interest due to improvement of nursing patient after extensive bowel resections, as well as development of parenteral nutrition opportunities, including home based options.

Basic Provisions: Patients with SBS require a multidisciplinary approach. The traditional tactic for treating such patients includes lifelong parenteral nutrition or intestinal transplantation, as a last resort. Rehabilitation measures for children with SBS should take into account the requirements for correcting malabsorption and rehabilitating nutritional status; relief of diarrhea, prevention and treatment of dysbacteriosis; control and correction of general digestive disorders.

The Discussion of the Results: The experience of management of five children with SBS in the Tyumen region of Russia was evaluated. It was found that the analogue of glucagon-like peptide 2 (GLP-2) - teduglutide - makes it possible to reduce the amount of parenteral nutrition, the time of infusions, as well as to gain a complete transition to enteral autonomy.

Conclusion: The method of treatment with teduglutide is the most promising for patients of this category.

Keywords: *Short Bowel Syndrome; Intestinal Failure; Intestinal Adaptation; Parenteral Nutrition; Teduglutide*

Introduction

A condition that occurs after extensive intestinal resections for various reasons is commonly referred to as Short Bowel Syndrome with Intestinal Failure (SBS-IF). This is a quite rare, debilitating, and potentially life-threatening disease in which the loss of intestinal absorption prevents people from absorbing enough nutrients from the food they eat, so they require parenteral nutrition/intravenous fluid infusion (PN/II, also known as. parenteral support) [1].

Disease coding by IDC-10

K92.1, K89.1- impaired absorption after surgery, not classified elsewhere.

The IDC-11 includes updated codes: DA96.04 Short bowel Syndrome (description: having less than 200 cm of residual small bowl with or without colon in an adult and for children (< 18 years), less than 25% of the normal length of intent for their responsive age. There is also a separate code for newborns - Short bowl syndrome in neonate (KB89.1).

IDC-11 is not used in the Russian Federation yet.

The incidence of SBS in general population ranges from 2 to 5 cases per 1 million [1].

The incidence of SBS among children is:

- 24.5 per 100,000 live births
- 353.7 per 100,000 preterms.

Mortality directly in SBS varies from 11 to 37.5%. The main cause of deaths, regardless of treatment tactics, is infectious complications [2].

The main causes of SBS are widely described in the literature and lead to the development of a number of pathological processes caused by extensive intestinal resection. Clinical manifestation depends on resection volume and level, in severe SBS with discharges through the stoma, electrolytes and minerals such as potassium, sodium, iron, zinc and magnesium are lost. Also, the one of the manifestations of short intestine syndrome is malabsorption with the loss of one or more nutrients from digestive tract accompanied with diarrhea and dehydration [3]. For many years, it was believed that only parenteral nutrition as replacement therapy can be used in SBS patients management. It should be noted that long-term parenteral nutrition can lead to septic complications, liver disorders, etc. In addition, such therapy is accompanied by a significant decrease in the patient's quality of life [4].

Currently, the rehabilitation of SBS patients is based on a multidisciplinary approach, the joint work of doctors of different specialties, which allows an objective assessment of the patient's condition and the subsequent effectiveness of treatment. For the past 20 years, the direction of hormonal treatment of patients with SBS has been actively developing, leading to intestinal adaptation and rehabilitation. Relatively recently, it has been found that glucagon-like peptides (GLP) analogues can reduce apoptosis of epithelial cells, as well as enhance nutrient absorption, that contributes to disease rehabilitation. GLP-2 (glucagon-like peptide type 2) is a natural gastrointestinal hormone secreted in ileac and colonic L cells in response to unabsorbed nutrients in the intestinal tube. The first and only pathogenetic medicine indicated for the treatment of SBS is teduglutide, a recombinant analogue of human GLP-2. It was approved in the European Union and the United States in 2012, registered in the Russian Federation in 2021 for the treatment of SBS patients aged 1 year and older. Due to increased nutrient absorption from the intestine, the need for parenteral nutrition decreases and, as a result, the volume and time of its administration reduce [5].

According to the preclinical and clinical data, the effects of GLP-2 and analogues (the first drug in the class is teduglutide) are the following:

- Sprouts (structural intestinal adaptation): induction of crypt cell proliferation, enhanced crypt cell survival, increased villous length and crypt depth, inhibition of enterocyte apoptosis, growing intestinal mass.
- Functional (functional intestinal adaptation): enhanced intestinal and portal blood flow, inhibition of HCl secretion in the stomach, slowing down intestinal motility, increased intestinal absorption capacity, including chylomicron formation and glucose absorption through SGLT-1 and GLUT-2, improving intestinal barrier function, reducing inflammation, increasing VIP production, reducing IL-1 and IL-2 expression [6].

Purpose of the Study

The purpose of the study is to consider and describe the main results of modern pharmacotherapy of patients with short intestine syndrome in world practice and in the Tyumen region.

Materials and Methods

PubMed, Clinical Trials and eLIBRARY databases were used for search by key words: short bowel syndrome, intestinal failure, intestinal adaptation, parenteral nutrition. Based on the analysis of 25 publications on the topic under study, information from 9 sources of scientific literature was included in the review. After the literature review formation, an analysis of the medical cases of five children diagnosed with SBS, who are under the supervision of specialists from the Center for Palliative Care for Children "Nadezhda" in Tyumen, was carried out.

Results and Discussion

The effect of teduglutide on the pediatric SBS patients was assessed based on an analysis of two clinical studies of the efficacy of teduglutide for children (a 12-week open-label multicenter study and a 24-week randomized double-blind study), which included children with SBS aged from 1 to 17 years who are in need of parenteral nutrition, with the exception of the patients who are unable to enteral nutrition [6]. Throughout a 24-week study among the children with SBS aged from 1 to 17 years, in addition to standard treatment, two doses of teduglutide were used: 0.025 mg/kg/day (n = 24) and 0.05 mg/kg/day (n = 26). 9 patients were included in the control group (standard treatment only). There were no significant clinical and laboratory abnormalities during the study. All patients in the standard treatment group and 98% of patients in the teduglutide groups experienced more than one adverse event, the majority were from mild to moderate severity. It should also be noted that neither polyps nor neoplasms were detected during colonoscopy and fecal occult blood test. 13 patients (54%) treated with 0.025 mg/kg and 18 (69%) treated with 0.05 mg/kg teduglutide achieved 20% reduction in parenteral nutrition volume on the 24 week of the study. Efficacy results - reduced parenteral nutrition volume and infusion time - are presented in table 1. Three patients in the 0.05 mg/kg/day group and 1 in the 0.025 mg/kg/day group. achieved independence from the PN. After 4-week washout period, two of these patients resumed PN and two more remained independent from PN. In addition, the level of citrulline in plasma increased, which indicates an increase in the mass of enterocytes [6].

Index	Teduglutide 0.025 mg/kg/day	Teduglutide 0.005 mg/kg/day	Standard treatment
Reduced parenteral nutrition ≥ 20 %, n (%)	13 (54 %)	18 (69%)	1
Volume of parenteral nutrition, ml/kg/day	↓16,2 (± 10,52)	↓23,30 (± 17,50)	↓6,0 (± 4,55)
Time of infusion, days/week (hours/day)	↓ 0,9 (± 1,78)	↓1,3 (± 2,24)	0
	↓ 2,5 (± 2,73)	↓3,0 (± 3,84)	↓0,2 (± 0,69)

Table 1: *The results of the effectiveness of drug treatment by the 24th week of the study.*

The study Kokoshis 2019, involved 17 patients with short bowel syndrome aged from 5 to 16 years, who were on parenteral nutrition for more than two years and had less than 80 cm of residual colon. They received a daily dose of teduglutide 0.05 mg/kg. On the week 12, 15 children had a decrease in parenteral nutrition by 20%, and calorie demand decreased by 29%. On the week 24, 7 patients reduced the dose of parenteral nutrition by 39%, on the week 36, 2 patients refused it completely [7]. The study found that patients with preserved colon could most easily be excommunicated from parenteral nutrition. This can be explained by the fact that their baseline needs for the PN use are usually lower than that of the patients with colectomy [8].

Experience of Tyumen region (Russian Federation)

9 children with short bowel syndrome are currently under the supervision at the Tyumen Regional Children's Palliative Care Center (hereinafter referred to as the Center). Patients' age varies from 3 months to 12 years. These children are recognized to be in need of palliative medical care by the decision of the medical commissions of the medical organizations where they were treated or observed, based on the prognosis of the disease, the severity of functional disorders in the gastrointestinal tract, the presence of severe comorbidity and painful symptoms [9].

After the diagnosis of SBS, during hospitalization at the RCCH (Russian Children's Clinical Hospital, Moscow), the children were provided with permanent central venous access, and individual parenteral and enteral nutrition regimens were selected for them. Reconstructive surgical treatment was applied on 7 children (sequential transverse enteroplasty - STEP). Previously, mothers during hospitalization were trained long-term PN technology, the basis of which is strict adherence to aseptics [9].

Our Center has implemented interdisciplinary strategies of patients' management with SBS in providing PMC (Palliative Medical Care) according to the scheme presented in figure 1.

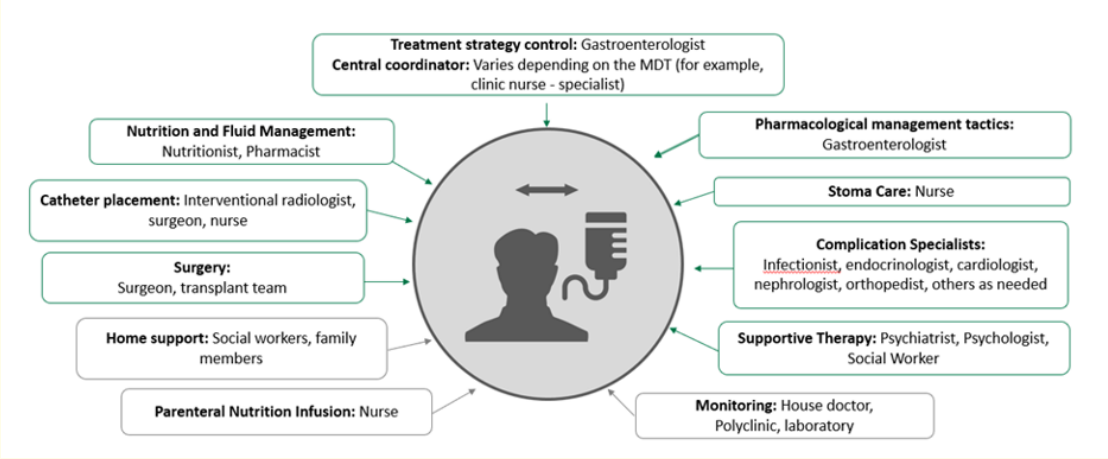

Figure 1: *Interdisciplinary management of patients with SBS in the provision of palliative care.*

A distinctive feature of the integrated approach to the provision of palliative care for children with SBS in Tyumen region (Russian Federation) is the provision of specialized items of medical nutrition (SPMN), enteral and parenteral, as well as consumables intended for the functioning of the Broviak tunneled catheter, in full, according to individual recommendations and needs, at the expense of annual regional subsidies.

The program for providing children with a palliative profile, including those suffering from SBS, started in the region in 2020. The implementation of the program is carried out according to the order of the Health Department of Tyumen Region dated 09/09/2020. No. 576 "Provision arrangement for children's receiving palliative care in Tyumen region with specialized medical food, consumables and medical products" (as amended and supplemented from 2021). The list of SPMN purchased to provide children with SBS in the region includes: Peptamen Junior dry mixture, Neokate Junior dry mixture, SMOFcabeven central, SMOFlipid, Vitalipid H children's emulsion, Soluvit N lyophilizate, Addamel H 10 mL, Aminoven, Sterafundin, Glucosteril, Liquidgen oil.

According to clinical recommendations, a child with SBS older than 1 year old, after reconstructive surgical treatment, who is on PN, is a candidate for treatment with an analogue of glucagon-like peptide 2 (GLP-2). Therefore, we started therapy with teduglutide with 5 children meeting these selection criteria [9].

The results of dynamic observation demonstrated the effectiveness of teduglutide, which was expressed in the change of some assessed indicators of the children's somatic state who have taken the drug for a year (from August 2021), as well as for six months (from January 2022) under the supervision of the specialists from the Center (Figure 2-6). The article provides data on the somatic indicators of two female children taking teduglutide for a year and a half, and three male children taking teduglutide for a year. The following items were evaluated:

1. Body weight (kg)
2. Height (cm)
3. Weekly/daily parenteral feeding volume (ml)
4. Daily stool volume (ml), as well as its consistency and frequency
5. Multiplicity of meals per day
6. Stoma volume (ml) per day
7. Clinical manifestations.

The heterogeneity of the collected and assessed parameters is primarily associated with the heterogeneity of the patients themselves, significant differences in the reasons for the resections, the nature of the existing concomitant diseases, the presence/absence of central venous access, and the presence of stoma.

The data of a female child born in 2009 receiving teduglutide since August 2021 are presented in figure 2.

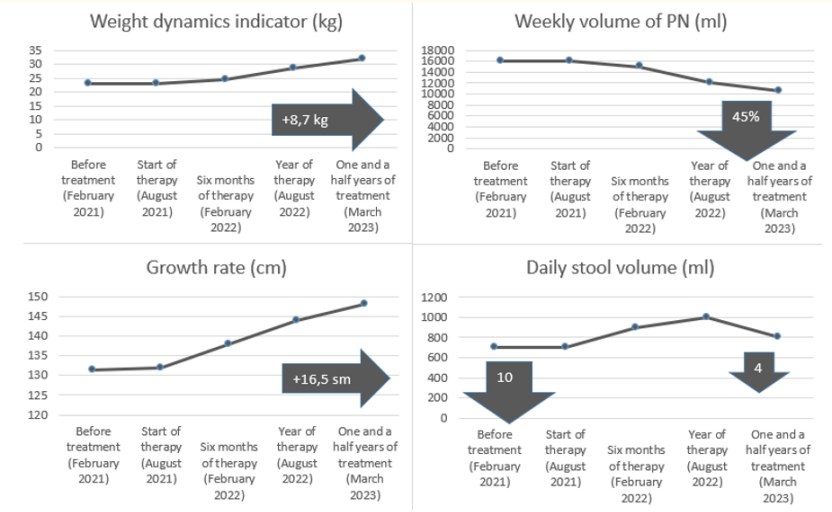

Figure 2: *Dynamics of clinical manifestations of SBS of a female child born in 2009 who has been taking teduglutide since August 2021.*

Clinical diagnosis (main): Recurrent stenosis of entero-colonic anastomosis. Short bowel syndrome, 200 cm variant after two autologous STEP reconstructions. Condition after multiple operations for neonatal ulcerative necrotizing colitis.

Complications of the underlying disease: Malabsorption syndrome. Grade 1 nutritional insufficiency. Grade 1 growth retardation.

Companion: Broviac 6.6 Fr Tunneled central venous catheter Carrier. Ileostoma carrier. Condition after surgical treatment of recto-vestibular fistulas.

From medical history: repeatedly operated for ulcerative necrotizing enterocolitis prematurely. A total of 5 operations were performed. The remaining length of small intestine is 50 - 70 cm, total colectomy with formation of entero-colonic anastomosis. In the outcome: short intestine syndrome, scar stenosis of the entero-colonic anastomosis, recto-vestibular fistula. 16.10.2013. STEP Extension Enteroplasty. 29.03.2016. repeated STEP, elimination of the recto-vestibular fistula. 04.09.2017 g. Surgery: posterior-sagittal proctoplasty.

Since October 2014, the child has been in the home parenteral nutrition system. It should be noted that the girl during the year of treatment with teduglutide added 16.5 cm in height and 8.7 kg in body weight. There were periodic interruptions in parenteral nutrition, which was carried out daily before the start of therapy, the volume was reduced by 25% (from 16 liters per week to 10,5). The stool volume increased (from 700 to 1000 mL per day) and its consistency thickened after stoma closure, however, as treatment continued and due to an increase in the volume of diuresis (due to an increase in the absorption capacity of the intestinal wall), the stool volume decreased (to 800 mL per day). The number of acts of defecation (from 10 to 4 per day), as well as the volume of stoma separated (from 1500 mL per day to 1100 mL) before it was closed, decreased significantly. The volume of enteral nutrition significantly decreased (from 600 ml per day of additional nutrition to 100 ml of Peptamen Junior), while the diet expanded, the child switched from a specialized individual diet to a common table. In July 2022, that is, 11 months after the start of therapy with teduglutide, the child underwent surgery to close the colostoma. Currently, the girl has independent defecation up to 4 times a day of the consistency of thick porridge without pathological impurities.

As one of the significant criteria for the effectiveness of the therapy, the volume of PN required steadily decreases (from 16 to 10.5 liters per week).

The data of a female child born in 2012 with teduglutide for a year are presented in figure 3.

Clinical diagnosis of the main: Short bowel syndrome: a variant without a large intestine, 100 cm small after autologous reconstruction according to the STEP method.

Complication of the underlying disease: Malabsorption syndrome. Chronic enteritis due to the syndrome of increased contamination of the small intestine with pathogenic flora. Violation of venous port function.

From the history of the disease: initially operated on the first day of life for multiple atresias (sausage form) of the small intestine and atresia of the right half of the large intestine with perforation of the transverse colon. 31.01.2013. enteroplasty was performed according to the STEP method with intestinal intubation through gastrostomy.

Since 2013, she has been in the home parenteral nutrition system. In February 2021 venous port is removed due to impaired functioning and catheter-associated infection. The uniqueness of this patient consists in the fact that within six months before the start of targeted therapy, she was deprived of the possibility of PN, due to the removal of an infected catheter and the loss of CVA (central venous access) for a replaceable catheter. In this regard, the parents organized frequent meals for her in small portions (up to 20 times a day every half hour), which was dictated by the child's need in the absence of parenteral support and the presence of dumping syndrome (stool was

observed almost every 1 - 2 meals). A year and a half after the start of therapy, the child managed to reduce the number of meals to 4 times a day (not counting specialized enteral nutrition with the Peptamen junior mixture) with the possibility of transferring to dishes of the common table. In addition to the lack of body weight gain (from 2019 to 2021, the weight was 19 kg), due to frequent acute respiratory diseases (ARD), the girl regularly lost 500 g in weight after each cold episode. Since the start of teduglutide therapy, parents have noted a significant decrease in the frequency of acute respiratory infections, as well as a serious increase in body weight (by 8 kg). In addition, the girl significantly decreased the number of acts of defecation (first to 3-4 per day, to date - to 2) and thickened feces (this can explain the pronounced decrease in stool volume, since the child has no stoma) (Figure 3).

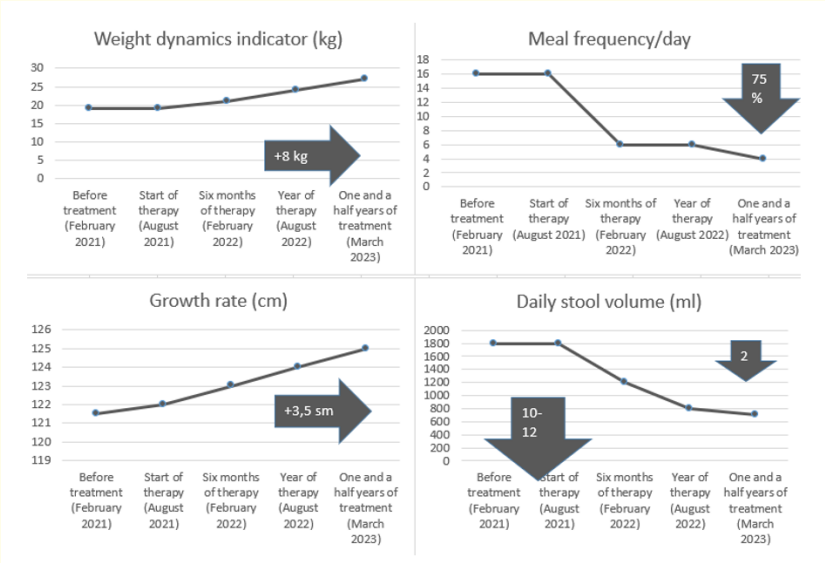

Figure 3: *Dynamics of clinical manifestations of SBS of a female child born in 2012 who has been taking teduglutide since August 2021.*

In addition, parents note a decrease in the frequency and intensity of flatulence as a painful symptom that reduces the quality of life. There was a decrease in the volume of specialized enteral nutrition (from 800 ml per day - a mixture of Peptamen Junior + Neokate Junior, as additional nutrition, to 500 - 600 ml per day - a mixture of Peptamen Junior, as additional nutrition) and the expansion of the diet: the addition of vegetable soups, stews and fruits. Parents also noted the appearance of the girl's opportunity to drink water, without fear of immediately running to the toilet. In addition, an increase in immunity was noted, which was expressed with a decrease in the frequency of intercurrent diseases (from 10-12 to 3-4 per year), an improvement in well-being and an increase in physical activity.

Very significant is the case of a male child born in 2014 (Figure 4).

Main diagnosis: K 91.2 Postresection syndrome of short bowel in the outcome of operative treatment of Ledd's syndrome, small intestine necrosis (18.09.14 - laparotomy, jejuno-ileo resection, anastomosis, 01.10.14 - relaparotomy, small intestine resection, duodeno-ileoanastomosis, 09.10.17 - relaparotomy, sanitation and abdominal drainage).

Complications of the main: Grade 1 nutritional insufficiency. Chronic intestinal failure. Chronic enterocolitis without exacerbation.

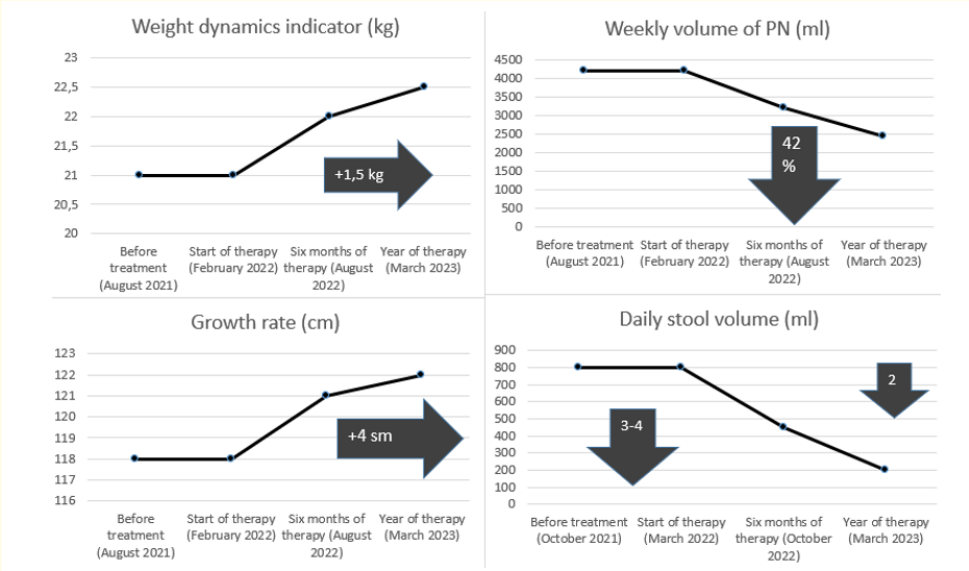

Figure 4: *Dynamics of clinical manifestations of SBS of a male child born in 2014 who has been taking teduglutide since March 2022.*

Comorbidities: Catheter associated history of superior vena cava thrombosis. Carrier of a tunneled Broviac 4.2 Fr catheter in the right internal jugular vein from 01.04.2020. Asthenic-neurotic syndrome of resident-organic genesis. Grade 3 sensorineural hearing loss on the left. Adenoides, state after an adenotomiya of 22.02.19.

Attention is drawn to the radical changes in stool: while before the start of therapy with teduglutide it was very watery and was observed 3 - 4 times a day, then, after a year of treatment, it acquired a thick consistency and decreased to 2 times a day. According to the observations of caregivers, there was a serious dynamics in the somatic state and well-being of the child, since before the start of therapy he often had disorders of the acid-base state (acidosis), which was manifested by the appearance of a shaky gait and sleepiness, and also had laboratory confirmation. These life-threatening conditions required emergency intervention by administration of saline solutions when the child even requested for oral administration of Regidron himself. From July 2022, the acid-base state is laboratory normal and, which is very important, clinical manifestations disappeared, episodes of lethargy and unstable gait stopped. A dramatic improvement in the quality of life of the child and his family members is certain. The boy studies in the 1st grade of a comprehensive school and successfully masters the program. Currently, he receives PN in 2 days after 2 to 700 ml per day, established 6 months after the start of targeted therapy. In April, 2023, He is scheduled to be hospitalized in the RCCH for further correction of PN and treatment.

Also of interest is the dynamics of somatic indicators and clinical manifestations of the disease in a male patient born in 2015, who started taking teduglutide in January 2022.

Clinical diagnosis of the main: Short bowel syndrome: variant 120-140 cm of the small intestine (after 05.08.2020) in the absence of the colon. Gastrointestinal neuromuscular disease: aganglionosis of the colon and distal ileum, focal hypoganglionosis of the small intestine. Status after multiple operations.

Complication of the underlying disease: Gastrostomy carrier, enterostoma carrier, peristomal infiltrate and dermatitis, erosive proctitis, chronic intestinal obstruction, malabsorption, malnutrition, maldigestion, grade 1 nutritional insufficiency, moderate chronic multifactorial anemia, metabolic bone tissue disease, thrombotic catheter associated complications.

Companion: Broviac Tunneled CVC Carrier.

From the history of the disease: repeatedly operated for gastrointestinal neuromuscular disease - ileal and colon aganglionosis with an outcome to short intestine syndrome. For 5 years he receives treatment in the home parenteral nutrition system. The boy could not start therapy for a long time, due to frequent enteritis, including fungal ones. In addition, the child had practically no independent stool (according to the parents, 1 once a month there could be a separation of mucous contents with a volume of 5 ml after increased tension of the abdominal wall). At the same time, up to 1 liter per day was separated by stoma. In addition, the boy deliberately refused oral nutrition during the day, agreeing to take it only in the evenings. During the year of teduglutide therapy, the child's weight increased by almost 3 kg, the height increase was 8 cm (Figure 5).

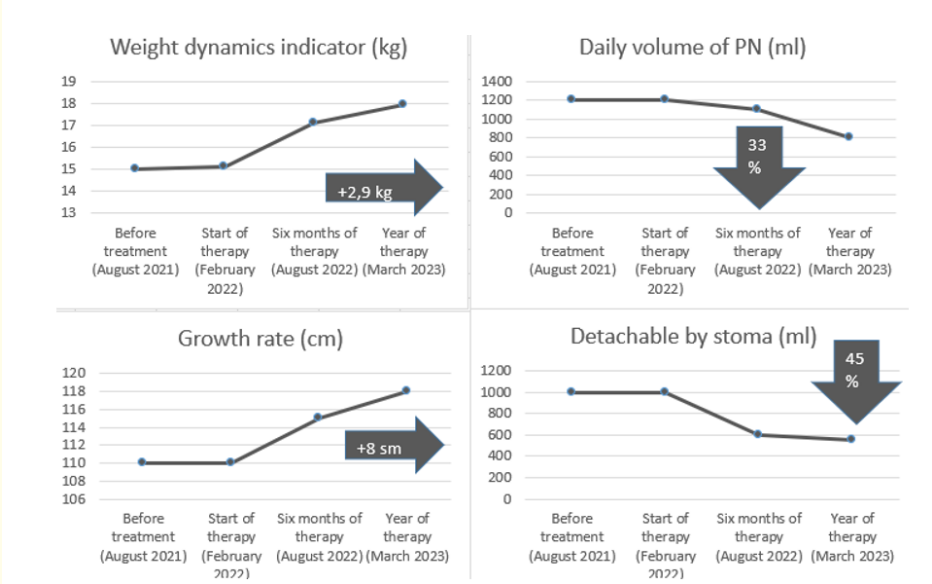

Figure 5: *Dynamics of clinical manifestations of SBS of a male child born in 2015 who has been taking teduglutide since January 2022.*

The volume of parenteral nutrition administered was significantly reduced (from 1200 to 800 ml per day), which made it possible to carry out it with a break of 1 day every three days. At the same time, the volume of stoma separated (from 1000 to 550 ml per day) was significantly reduced and, importantly, daily independent stool appeared (up to 20 ml in the volume of daily defecation). Due to the expansion of the diet and the normalization of the diet (the transition from a specialized individual diet to expansion due to bran and fruits in small quantities), it was decided to reduce the volume of additional enteral nutrition (from 600 ml per day to 100 ml specialized mixture of Neokate Junior).

Consider the data of a male child in 2019, the diagnosis of the main one: K 91.2 Postresection syndrome of the short bowel in the outcome of operative treatment of Ledd syndrome and necrosis of the small intestine (05.08.19 - subtotal resection of the small intestine,

echinostoma excretion, 18.09.19 - adhesiolysis, ileo-jejunal anastomosis application, 04.10.19 - adhesiolysis, juno application coloanastomosis, 17.12.21 - laparotomy, revision of the abdominal cavity, removal of a foreign body from the intestine, suturing of the small intestine, application of gastrostoma, 29.12.21 - restoration of gastrostoma); residual length of small intestine at the time of resection - 2 cm (lean), colon preserved).

Complications of the underlying disease: E 44.0 Grade 2 nutritional insufficiency.

Comorbidities: K 52.8 Erosive eosinophilic left-sided colitis.

The carrier of the tunneled Broviac catheter 4.2 in the VIV to the right of the 18.09.2019, repairing the catheter from the 21.10.21. Cerebroasthenic syndrome of resident-organic genesis. ONR. delayed formation of expressive speech. Retinal angiopathy.

The dynamics of SBS manifestations is shown in figure 6.

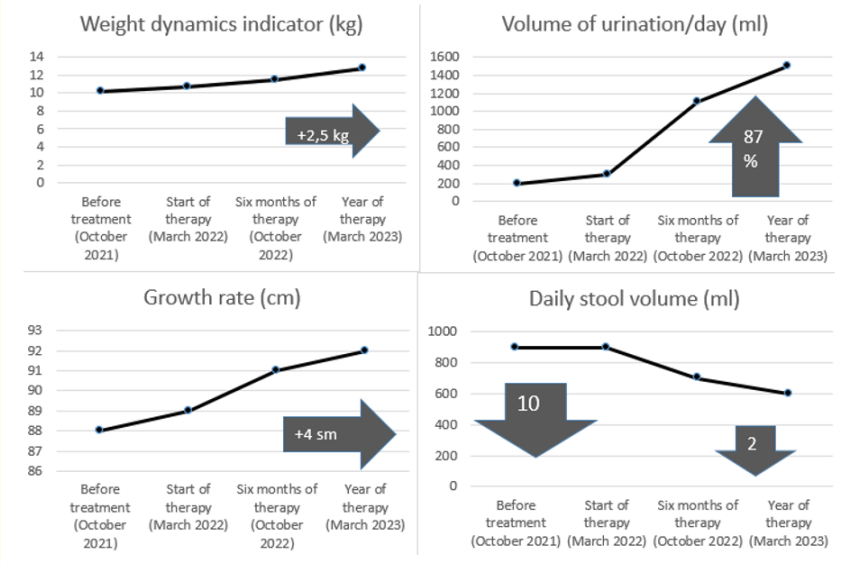

Figure 6: *Dynamics of clinical manifestations of CCM of a male child born in 2019 taking teduglutide since March 2022.*

Due to the fact that changes in the volume of parenteral nutrition are planned for April during hospitalization in the RCCH, the dynamics of the daily volumes of stool and urine was chosen to assess the effect of improving the absorption function of the intestines. As you can see in the figure, the volume of urinary discharge increased significantly (by 87%), while the volume and frequency (from 10 to 2 per day) of defecation decreased significantly. These changes indirectly indicate a significant increase in the absorption capacity of the intestine, which confirms the effectiveness of the hormone therapy. The volume of parenteral nutrition is still injected at the initial level of 760 ml per day.

In addition to the above, over the past year of teduglutide therapy, the child showed the appearance of 120 g of porridge per feeding (earlier than 70g), which he absorbs well. Expanding the diet by adding banana and apple. In addition, the Peptamen mixture is obtained

(only 1 times at night 50 - 70 ml). Parents note an improvement in appetite, and the possibility of adding cookies. An important thing for themselves is a decrease in the time for infusion of PN 13.5 hours (earlier than 15 hours).

Summarizing the results, the following significant changes in the condition of children can be noted:

1. Gradual reduction in volume and calories delivered with parenteral nutrition.
2. Expanding the diet by introducing traditional foods, gradually reducing the need for special cooking, separate from other family members, switching to a common table.
3. Change in the amount and consistency of stool (decrease in the number of defecations, thickening of the consistency of stool, return to the physiological act of defecation (colostomy closure - clinical case 2).
4. Reduction in relapses of D-lactate - acidosis, dehydration (reducing the risk of life-threatening conditions requiring emergency measures).
5. Change in weight-height parameters (weight gain and height, approximation to physiological age standards).
6. Increased body defenses (reduced frequency of intercurrent diseases, increased physical activity, improved general well-being).
7. Improving the patient's quality of life (there is an opportunity to attend school, as well as sports sections and institutions of additional education).
8. Improving the quality of life of parents (including reducing the time required to organize a home PN system, freeing up time for employment and socialization, reducing anxiety, risk of developing depressive conditions).

In all cases, in the first days of teduglutide use, children experienced episodic abdominal pain, which was self-managed after 2 - 3 weeks without subsequent recurrence. No other adverse events were observed in patients during the entire treatment period.

At the same time, it must be borne in mind that each child is individual. Individuality is manifested both in the ability and timing of intestinal adaptation, and in the response to therapy and, accordingly, in the achievement and stability of the result.

The analysis of the presented clinical examples allows us to hope for further successful use of teduglutide in the complex therapy of short intestine syndrome, the main task of which is to achieve enteral autonomy, and as a result, improve the quality of life, which is extremely important primarily for pediatric patients.

Conclusion

Rational treatment tactics for patients with short bowel syndrome and a comprehensive multidisciplinary approach can improve the quality of life of patients with this disease. Studies have proven and in practice confirmed that teduglutide - an analogue of human GLP-2 - allows to reduce the volume of parenteral nutrition consumption and infusion time in children and adults, as well as achieve complete enteral autonomy. Most of the adverse events associated with teduglutide therapy were mild to moderate in severity and were manifested mainly by abdominal pain, which was quite expected in SBS. Thus, numerous studies and real clinical practice have shown the effectiveness of teduglutide in the treatment of the patients with short bowel syndrome. The experience of our center indicates that, when creating certain working conditions in the form of an organization of a multidisciplinary team of specialists, providing dynamic monitoring, as well as meeting the personalized need of patients with SBS for the necessary medications, consumables, specialized medical food products, including training parents/caregivers to work in compliance with the principles of aseptics and antiseptics, as well as strict accounting of new and monitoring of the condition of existing patients in the regional register, patients with such a diagnosis can live ordinary daily life and be fully socialized in human society.

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Conflict of Interest

The authors declare that there is no conflict of interest.

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