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Case Report

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Nutritional and dietary approach in the post-surgical follow-up of Hirschsprung disease: a case report

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ABSTRACT

Hirschsprung disease (HD) is a congenital condition caused by abnormal development of the enteric nervous system during the embryonic period, characterized by colonic aganglionosis. Total colonic aganglionosis (TCA) refers to the involvement of the entire colon extending to the terminal ileum, accounting for 3–12% of all HD cases. A 45-month-old male patient presented to the clinic with complaints of frequent vomiting and more than 10 watery diarrhea episodes per day. The patient's medical history revealed the development of short bowel syndrome (SBS) following TCA. Physical examination indicated a poor general condition, decreased skin turgor and tonus, along with growth and developmental delay. Laboratory findings demonstrated low sodium (Na) and potassium (K) levels with electrolyte imbalances. A low-carbohydrate, high-protein diet was planned alongside hydration therapy. Additionally, the family was educated on nutritional management and avoiding harmful foods. Following 17 days of treatment and dietary interventions, the patient's weight increased by 1.5 kg, and complaints of diarrhea and vomiting subsided. During a two-year follow-up period, a reduction in hospitalization frequency and improvements in the patient's overall clinical condition were observed. The individualized treatment and nutritional plan significantly enhanced the patient's quality of life. The management of SBS, which may develop after HD surgery, requires individualized nutritional plans, a multidisciplinary approach, and continuity in patient-family education, all of which are critically important.

Keywords: Hirschsprung disease, pediatric surgery, nutrition and diet, short bowel syndrome

INTRODUCTION

Hirschsprung disease (HD) is a congenital condition caused by abnormal development of the enteric nervous system during embryonic development, characterized by colonic aganglionosis.[1] It occurs approximately in 1 out of every 5000 live births and is one of the most common congenital disorders of the lower gastrointestinal tract.[2] Children with HD typically present with symptoms related to bowel obstruction, such as delayed passage of meconium, abdominal distension, and bilious vomiting within the first 24-48 hours of life. Diagnosis is usually made within the first six months of life. The congenital defect underlying HD involves the complete absence of ganglion cells in the most distal part of the gastrointestinal system.[3] Due to this aganglionosis, there is a lack of peristaltic movements and the inability of the smooth muscles to relax, leading to functional narrowing of the affected bowel segment. Approximately 80% of patients have aganglionic segments localized to the rectosigmoid region; however, in rare cases, the condition may involve the entire colon or even a portion of the small intestine.[2] The involvement of the entire colon extending to the terminal ileum is referred to as total colonic aganglionosis (TCA), accounting for 3–12% of all HD cases.[4] Postoperative complications frequently observed in patients with TCA include stoma prolapse, chronic constipation, fecal incontinence, anastomotic leakage, and Hirschsprung-associated enterocolitis (HAEC).[5,6] Furthermore, the development of short bowel syndrome (SBS) is a potential postoperative concern in cases of TCA.[7]

SBS is a condition characterized by malabsorption resulting from the resection of a portion of the small intestine or due to congenital malformations.[8,9] The degree of malabsorption varies depending on the extent of resection or the remaining intestinal segment.[9] Shortened bowel length leads to accelerated intestinal transit and insufficient absorption of nutrients.[10] Factors such as gastric acid hypersecretion, bacterial overgrowth in the intestine, impaired absorption of fats and bile salts, and reduced fluid absorption collectively contribute to the development of high-osmolarity diarrhea.[7,11] In addition to refractory diarrhea, symptoms such as steatorrhea, weight loss, dehydration, and electrolyte imbalances are commonly observed.[11,12] Managing levels

of magnesium, calcium, and potassium can be particularly challenging in individuals with SBS.[11] Furthermore, these patients often experience nutritional deficiencies and growth and developmental delays.[4]

In the postoperative recovery period of SBS, parenteral nutrition therapy should be initiated to maintain fluid and electrolyte balance and optimize carbohydrate, protein, and fat intake. Following a period of adaptation after resection, moderate enteral nutrition should be introduced.[13] During this phase, semi-elemental or elemental formulas may be utilized. Subsequently, the transition to age-appropriate solid foods with a low-carbohydrate and high-protein composition is recommended.[14]

Surgical intervention forms the cornerstone of treatment for HD; however, postoperative follow-up and care are crucial for the patient's long-term health and quality of life.[1] Issues such as bowel dysfunction and surgical complications that affect quality of life necessitate long-term monitoring. A multidisciplinary approach and individualized treatment plans are essential for improving the quality of life in these patients.[4] This article discusses a case aimed at evaluating the nutritional and dietary approach in the postoperative period of a patient operated on for HD.

CASE

A 45-month-old male patient presented to the pediatric surgery outpatient clinic with complaints of vomiting and more than 10 episodes of profuse diarrhea daily. The patient's medical history revealed an absence of meconium passage within the first 24–48 hours during the neonatal period. Following a preliminary diagnosis of HD based on findings from a colon radiograph and biopsy, a colostomy was performed. During surgery, frozen biopsy results identified TCA. Subsequently, an ileoanal pouch was constructed using the Duhamel–Martin procedure, and the patient was monitored with an ileostomy for six months before its closure. It was noted that the patient continued to experience frequent enterocolitis episodes, leading to SBS symptoms characterized by impaired nutrient absorption due to persistent diarrhea.

Due to persistent diarrhea and SBS, the patient exhibited growth and developmental delays, weight loss, and impaired absorption of vitamins, sodium, and potassium. On physical examination at the time of admission to our clinic, the patient was in poor general condition, with decreased skin turgor and tonus. Abdominal examination revealed old scar tissue from previous surgeries, and bowel sounds were hyperactive. On palpation, no tenderness, guarding, or rebound was noted. Laboratory findings showed decreased sodium (Na) and potassium (K+) levels, while the rotavirus and adenovirus antigens were negative (Na: 135 mEq/L, K+: 3.62 mEq/L, Cl: 96.5 mEq/L, Mg: ALT: 65.5 U/L, AST: 50.1 U/L, CRP: 56.09 mg/dl, albumin: 47.8 g/L, Hgb: 14.0 g/dl).

Dietary habits revealed frequent and excessive consumption of foods such as tea, biscuits, chips, olives, raw meatballs, and raw minced meat. Anthropometric measurements indicated a body weight of 11.5 kg (3rd percentile) and a height of 90.8 cm (3rd percentile).

The patient's treatment plan included intravenous (IV) hydration, metronidazole, ondansetron, and pantoprazole. Due to insufficient oral intake, a nasogastric (NG) tube was inserted, and nutritional support was initiated with an isocaloric enteral formula (**Table 1**).

Table 1. Nutritional plan

Days 1-2:	1300 kcal/24 h (enteral product 1.0 kcal).
Days 3-6:	1300 kcal/24 h enteral product+continued oral intake (small amounts of water/light tea, table salt intake).
Day 7:	960 kcal/24 h enteral product+supported with oral intake (soup, meatballs).
Days 8-10:	720 kcal/24 h enteral product+improved oral intake but still insufficient based on hospitalization.
Days 11-12:	540 kcal/24 h enteral product+increased oral intake (able to consume 2 eggs, meatballs, and ½ bowl of soup comfortably).
Day 13:	270 kcal/24 h enteral product.
Days 14-17:	Various enteral products were trialed (strawberry, vanilla, and unflavored). Oral intake was good.

During the initial days of hospitalization, the patient experienced at least 10 episodes of watery, voluminous diarrhea and 4 episodes of vomiting daily. By the second day of admission, the patient's general condition began to improve, vomiting subsided, and the frequency and consistency of diarrhea improved progressively, decreasing to 4–6 semi-formed stools per day. With the implementation of an adjusted oral feeding and nutrition plan, the patient showed improvement in blood values, oral intake, clinical findings, and physical activity levels. Over the 17-day hospitalization, the patient gained 1.5 kg in body weight and was subsequently discharged.

Approximately 2 months after discharge, the patient presented to the clinic again with more severe complaints of diarrhea, nausea, and vomiting, and was readmitted. A significant decrease in sodium levels (Na: 121 mEq/L) was observed, and IV hydration along with NG tube-enteral feeding therapy was initiated. Alongside enteral nutrition, a healthy eating plan was developed and implemented. Upon assessing the patient's daily diet, it was determined that the patient paid little attention to their nutrition, and there was poor adherence to dietary recommendations. The family's socio-economic level contributed to a high level of non-compliance, as they permitted the consumption of unprocessed foods like raw minced meat and encouraged direct consumption of salt, potentially affecting sodium levels. Dietitian collaboration was reinforced with the family, offering nutritional and dietary education both face-to-face and through communication tools (e.g., phone), with frequent monitoring intervals.

Over the course of approximately 2 years, the patient visited the clinic 14 times, with 7 admissions spaced two months apart due to poor clinical condition and low sodium and potassium levels. It was observed that admission durations decreased progressively with each visit (**Figure 1**). At the

most recent visit, the patient's sodium level was 123 mEq/L and potassium level was 3.02 mEq/L. Although the patient's overall condition remained poor, there was a noticeable increase in growth and weight gain, with body weight reaching 18 kg. Additionally, the patient began to comfortably consume previously avoided foods (e.g., eggs). With the latest follow-ups, the foods the patient desired to consume were prioritized, and a personalized nutrition plan was redesigned to maintain fluid-electrolyte balance and prevent diarrhea (Table 2).

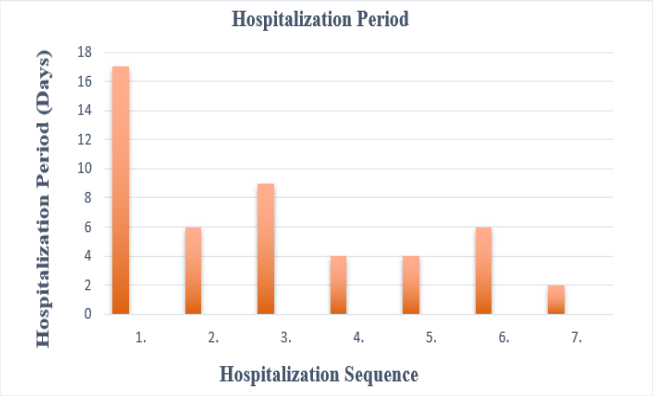

Figure 1. Graph of patient's hospitalization duration

Tablo 2. Sample nutritional plan

Morning
1 cup of herbal tea (linden tea)
1 boiled egg (50-60 g)
1 slice of white cheese (40 g)
4-5 olives
2 thin slices of white bread (50 g)
Mid-morning snack
½ carton of enteral product/plus fiber (1 ml=1.5 kcal) (100 ml)
Lunch
½ bowl of soup (100 ml-½ ladle)
2 tablespoons of yogurt or tzatziki
1 small banana/apple/peach
2 thin slices of white bread (50 g)
Afternoon snack
½ carton of enteral product/plus fiber (1 ml=1.5 kcal) (100 ml)
Dinner
2 portion of meat/chicken/ground beef (60 g) (e.g., boiled chicken, meatball, homemade hamburger)
4 tablespoons of vegetable dish
3 tablespoons of rice pilaf
1 bowl of tzatziki (made with 4 tablespoons of yogurt)
Night snack (2-3 hours before bed)
1 carton of enteral product/plus fiber (1 ml=1.5 kcal) (200 ml)

Upon analyzing the patient's anthropometric measurements, initial admission (at 45 months) showed a body weight of 11.0 kg (3rd percentile) and height of 90.8 cm (3rd percentile). At the latest admission (at 66 months), the body weight had increased to 18.0 kg (25-50th percentile), and height was 106 cm (10-15th percentile) (Figure 2).

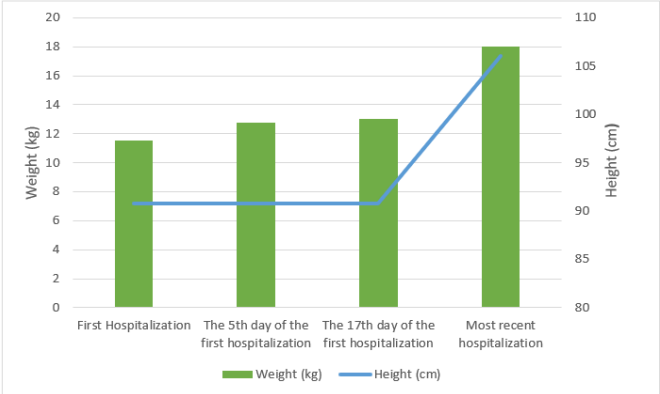

Figure 2. Graph of patient's body weight and height

DISCUSSION

This study aims to comprehensively examine the nutrition and diet approach in cases of TCA secondary to Hirschsprung's disease and associated SBS. TCA is a rare form of Hirschsprung's disease that leads to significant clinical and nutritional challenges early in life.[4] In the literature, TCA cases are frequently complicated by conditions such as enterocolitis, malnutrition, growth retardation, and electrolyte imbalances.[15] This highlights the need for a multidisciplinary approach beyond surgical management, involving close monitoring by pediatric surgeons, gastroenterologists, pediatricians, dietitians, psychologists, and nurses. In cases of Hirschsprung's disease with total colectomy and ileostomy, the importance of medical treatment, as well as care and nutritional support, is emphasized. Multidisciplinary collaboration in such cases is crucial in reducing hospital stays and minimizing the need for additional surgeries.[4]

In Hirschsprung's disease, surgical treatment methods such as Soave, Swenson, and Duhamel-Martin procedures are commonly used, each with distinct advantages and associated risks of complications.[16] The Duhamel-Martin procedure, in particular, has been associated with frequent postoperative complications such as enterocolitis, constipation, and vomiting, as reported in the literature.[17] In our case, similar findings were observed, with postoperative enterocolitis attacks and a progressive course of SBS being documented.

The outcomes of SBS include malabsorption, diarrhea, dehydration, electrolyte imbalances, nutrient deficiencies, and inadequate weight gain or loss, necessitating parental support along with both enteral and solid nutritional support.[8,18] Non-cellulose forms of fiber in grains contribute to an increase in stool volume, while pectin and guar found in fruits and vegetables provide a diarrhea-preventive effect by slowing intestinal transit time.[18] In our case, efforts were made to include these foods in the individualized nutrition plan.

A significant issue in this case was the family's lack of adherence to nutritional treatment, exacerbated by a low socio-economic level, which contributed to this noncompliance. This situation negatively affected the patient's clinical course. Continuous support through a structured follow-up mechanism and collaboration with dietitians helped implement an individualized nutrition plan. This approach is believed to have played a significant role in the patient's progress in growth and development. Literature supports that a multidisciplinary approach positively influences the health outcomes of individuals.[19]

CONCLUSION

As a result, the management of SBS following HD surgery requires individualized nutrition plans, a multidisciplinary approach, and continuous patient-family education. Maintaining direct contact with the patient and family, utilizing communication channels, and ensuring timely hospital admissions when necessary are essential. Early evaluation of blood electrolyte levels and appropriate dietary interventions are crucial for facilitating a smoother recovery and improving the patient's quality of life during this period.

ETHICAL DECLARATIONS

Informed Consent

The patient's family signed a free and informed consent form.

Referee Evaluation Process

Externally peer-reviewed.

Conflict of Interest Statement

The authors have no conflicts of interest to declare.

Financial Disclosure

The authors declared that this study has received no financial support.

Author Contributions

All of the authors declare that they have all participated in the design, execution, and analysis of the paper, and that they have approved the final version.

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