



Nutritional Therapy in Adult Hirschsprung Disease Post-Total Colectomy and Ileostomy with Severe Protein-Energy Malnutrition: A Case Report

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Abstract

Adult Hirschsprung disease, also known as congenital aganglionic megacolon, is a rare congenital disorder characterized by the absence of ganglion cells in the distal segment of the large intestine, diagnosed in adulthood. Impaired nutrient absorption due to the dysfunctional intestinal segment can lead to malabsorption and malnutrition. A 23-year-old female presented with severe protein-energy malnutrition after total colectomy surgery for suspected adult Hirschsprung disease. She had a history of chronic constipation from childhood, abdominal pain, and abdominal distension. She was referred to a clinical nutritionist 2 days post-surgery. The nutrition therapy started with oral and parenteral nutrition, gradually increasing macronutrient requirements based on clinical condition and gastrointestinal tolerance. On the 12th day post-total colectomy surgery, she developed complications of obstructive ileus, requiring ileostomy. Laboratory tests showed signs of inflammation. Nutritional support for adult Hirschsprung disease focuses on meeting energy requirements, enhancing nutrient absorption after abdominal surgery, and minimizing post-operative inflammation.

Keywords: Hirschsprung Disease, Colectomy, Ileostomy, Malnutrition

Abstrak

Adult Hirschsprung Disease (AHD) adalah gangguan kongenital langka yang ditandai tidak adanya sel ganglion di distal usus besar yang terdiagnosis saat dewasa. Kasus ini sering sulit didiagnosis karena presentasinya yang atipikal dan dapat keliru didiagnosis sebagai gangguan saluran pencernaan lain. Penyerapan nutrisi yang terganggu akibat segmen usus yang tidak berfungsi dengan baik menyebabkan malabsorpsi dan malnutrisi. Seorang wanita, umur 23 tahun dengan malnutrisi protein energi berat post operasi total kolektomi et causa suspek *Adult Hirschsprung Disease*. Keluhan konstipasi kronik dirasakan berulang sejak anak-anak, nyeri perut dan dirasakan membesar dalam 1 bulan terakhir. Pasien dilakukan total kolektomi, terapi gizi klinik di mulai dengan nutrisi melalui oral dan parenteral, nutrisi ditingkatkan bertahap sesuai dengan kondisi klinis dan toleransi saluran cerna. Post operasi total kolektomi, terjadi komplikasi ileus obstruksi sehingga dilakukan tindakan ileostomy. Terapi nutrisi berfokus pada pemenuhan energi, meningkatkan penyerapan nutrisi post operasi, serta menurunkan inflamasi akibat operasi.

Kata kunci: *Hirschsprung, Kolektomi, Malnutrisi, Ileostomi*

Introduction

Hirschsprung disease is a rare congenital disorder characterized by the absence of ganglion cells in the distal colon. This condition creates a functional obstruction in the affected segment, causing abdominal distension and chronic constipation. The etiology of Hirschsprung disease involves genetic and environmental factors.^{1,2}

The prevalence of Hirschsprung disease is approximately 1 in 5,000 births worldwide, typically detected in the neonatal period, with a male-to-female ratio of 4:1. Premature babies and those born to mothers with obesity are at higher risk for Hirschsprung disease. Although Hirschsprung disease is most commonly diagnosed in children, it can also be found in adults. Adult Hirschsprung disease refers to cases diagnosed in individuals over ten years old.³ Adult Hirschsprung Disease (AHD) may be misdiagnosed as other intestinal disorders. This condition presents as chronic constipation, abdominal distention, and intestinal obstruction. Adult Hirschsprung disease is challenging to diagnose due to the variability of clinical presentation and intermittent signs and symptoms.^{2,4}

Treatment for Hirschsprung disease involves surgery to remove the nonfunctional intestinal segment and restore normal intestinal flow. Surgical procedures in AHD referred to removing the aganglionic segment of colon, including total colectomy, subtotal colectomy, low anterior resection, and colonoscopic decompression for emergency intestinal obstruction.⁵ Preoperative patients with Hirschsprung disease are recommended to follow a high-fiber diet and maintain adequate hydration to ensure regular defecation. Meanwhile, postoperative patients with total colectomy and ileostomy due to Hirschsprung disease may be at risk for malnutrition due to malabsorption. Colectomy leads to insufficient absorption of water and electrolytes, whereas ileostomy affects the absorption of essential macronutrients and micronutrients.^{2,6}

An interdisciplinary approach should be implemented among digestive surgeons, gastroenterologists, and clinical nutritionists to treat cases of adult Hirschsprung disease, aiming to avoid complications, shorten hospital stays, and improve quality of life.

Case presentation

A 23-year-old woman consulted the clinical nutrition department for nutrition management after total colectomy due to Adult Hirschsprung disease. The patient was instructed on oral nutrition with a clear liquid diet every two hours by the digestive surgery department and received parenteral nutrition from the anesthesiology department. The patient had decreased oral intake over the past year, caused by regular abdominal pain and constipation. She also complained of nausea and vomiting, but had no difficulty swallowing. She lost 9 kilograms during the year. Upon hospital admission, she had experienced constipation for 29 days and reported normal urination via a catheter.

Food habits and food recall 24 hours

She had decreased oral intake over the past year due to bloating after eating, abdominal pain, and constipation. The patient had regularly consumed vegetables and fruits over the last 6 months. No food allergies were reported. Over the previous 24 hours, the patient received

parenteral nutrition totaling 293 kcal, with 27 grams of protein, 50 grams of carbohydrate, and no lipids.

Physical examination, anthropometric data, and additional examination

The patient's anthropometric data revealed a height of 155 cm and an ideal body weight of 49.5 kg; however, actual body weight could not be obtained. Subjective Global Assessment (SGA) screening classified the patient as severely malnourished (Score C).

Vital signs were normal for blood pressure, pulse, and temperature. Respiratory support was administered via a nasal cannula at a rate of three liters per minute. Loss of subcutaneous fat was observed on the chest and temporal region of the face, whereas muscle wasting was evident in all extremities.

Laboratory examinations showed hypoalbuminemia, mild hypokalemia, leukocytosis, immune system depletion, and an increased neutrophil-to-lymphocyte ratio (NLR) (Table 1). A chest X-ray (thorax photo) revealed elevation of the left diaphragm. Histopathological examination confirmed the presence of aganglionic cells in the intestinal segment.

Tabel 1. Laboratory result during care

Laboratory test	Before surgery	POD 5 total colectomy	POD 9 total colectomy	POD 1 ileostomy	Reference	Value
Day of nutritional care	D-2	D-5	D-9	D-12		
Haemoglobin	12.2	8.4	10.8	10	12.0 - 16.0	gr/dL
MCV	77.6	76.2	76.2	79.3	80 - 97	fL
MCH	26.2	25.6	25.6	26.2	26.5 - 33.5	pg
MCHC	33.8	33.6	33.5	33	31.5 - 35.0	gr/dL
Leukocyte	23.1	11.5	17	28	4 - 10	10 ³ /uL
TLC	713	1161	1314	1698	1.5 - 4	10 ³ /u L
NLR	30	8.2	11.3	14.4	0.78-3.53	-
Platelets	356	120	547	714	150 - 400	10 ³ /uL
Blood glucose	159	92	not checked	not checked	70 - 200	mg/dL
Natrium	138	132	131	134	136 - 145	mmol/l
Potassium	3.3	2.6	2.9	3.5	3.5 - 5.1	mmol/l
Chloride	112	103	104	107	97 - 111	mmol/l
Albumin	2.1	2.2	2.7	not checked	3.5 - 5.0	gr/dL

POD: Post Operation Day, MCV: Mean Corpuscular Volume, MCH: Mean Corpuscular Hemoglobin, MCHC: Mean Corpuscular Hemoglobin Concentration, TLC: Total Lymphocyte Count, NLR: Neutrophil-to-Lymphocyte Ratio.

Nutritional diagnosis, nutritional therapy, follow-up, and evaluation.

The medical diagnosis was Hirschsprung disease with total colectomy. The nutritional status assessment revealed severe malnutrition, characterized by inadequate intake, hypoalbuminemia, and electrolyte imbalance. Nutritional therapy aimed to provide a diet that was tolerated by the gastrointestinal system, with long-term goals of improving nutritional status, enhancing recovery, and reducing the length of stay. The patient received 1,800 kcal via a combination of oral and parenteral nutrition, consisting of 1.7–2 g/kg of body weight of protein (14.5% of total calories), 247 g of carbohydrates (55% of total calories), and 61 g of fat

(34.6% of total calories). The nutrition plan was gradually increased until the total calorie goal was achieved.

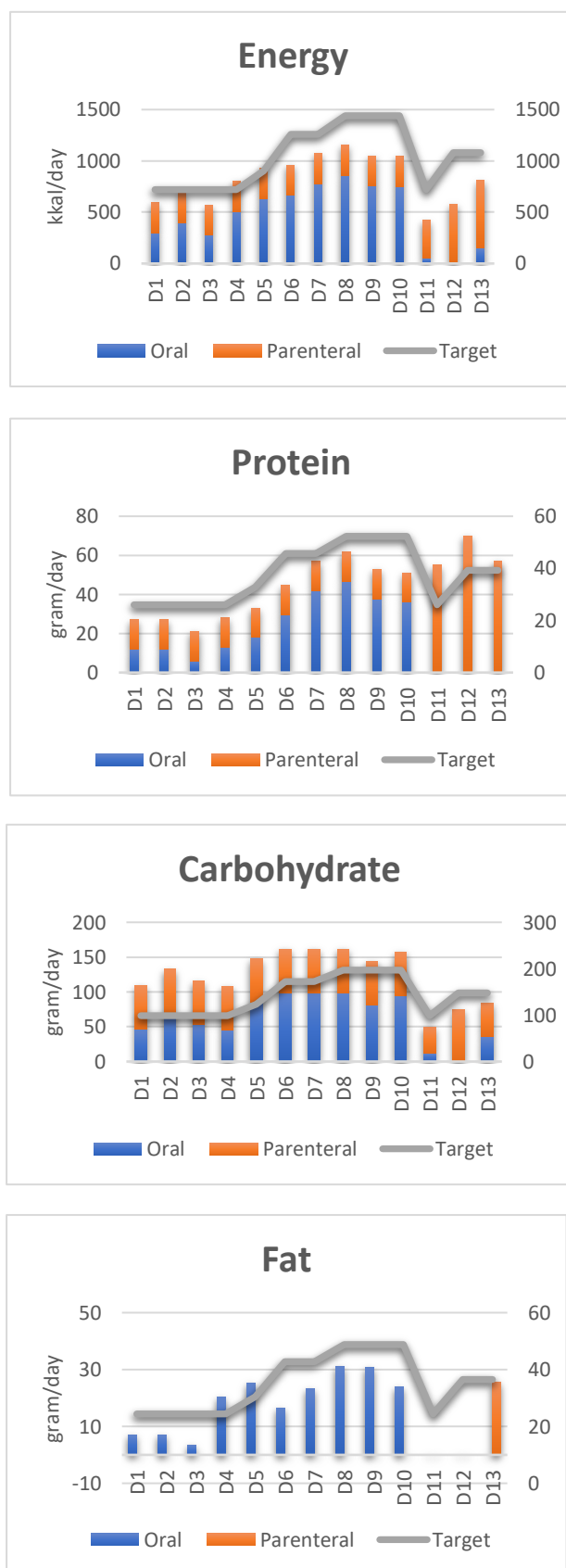


Figure 1 – 4. Observation Energy, Protein, Carbohydrate, and Fat Intake During Care

During treatment, the patient's intake fluctuated, but they were able to achieve up to 66% of the total calorie requirement, 100% of the target protein, 100% of the target carbohydrate, and 50% of the target fat (Figure 1-4).

Discussion

Patients with Adult Hirschsprung disease (AHD) often present with chronic constipation, abdominal distention, and intestinal obstruction, leading to loss of appetite. Malabsorption also occurs due to motility abnormalities that decrease nutrient absorption. Inflammation stimulates insulin resistance and creates a hypercatabolic condition, including proteolysis and lipolysis, ultimately resulting in severe malnutrition.^{7,8}

The diet prescription was initiated with a liquid diet combined with parenteral nutrition, and the diet consistency was upgraded to a solid diet following observation. Parenteral nutrition was provided during care to meet calorie targets, as the patient was a picky eater and refused the prescribed diet. Nutrition counseling and education were delivered to the patient to address her eating issues.

The total colectomy procedure affects intestinal transit time, typically resulting in faster transit. Semi-elemental formulas have been shown to be better tolerated after colectomy. An oligomeric formula was given to facilitate better absorption and increase intestinal tolerance. However, diarrhea occurred after the total colectomy procedure, with electrolyte imbalance as a consequence of loss of colon function and dysbiosis due to intestinal dysmotility.^{9,10}

Post-surgical inflammation generates hypoalbuminemia, immune depletion, and an increased neutrophil-to-lymphocyte ratio. Surgery-related stress stimulates the production of pro-inflammatory cytokines, including IL-1, TNF- α , and IL-6. C-reactive protein is also activated, initiating the synthesis of acute-phase proteins and leading to hypoalbuminemia.¹¹ To overcome this situation, a protein intake of 2 g/kg body weight/day was provided, consisting of a mix of protein from oral nutrition supplements, amino acid parenteral infusion, and albumin capsules. Additionally, zinc, vitamins B1, B6, and B12 were administered to combat inflammation.

Zinc, as an antioxidant, protects cells from reactive oxygen species (ROS) and reactive nitrogen species (RNS), modulating cytokine production and initiating proliferation of CD8⁺ T-cells. Vitamins B affect inflammation by managing cytokine production, activating natural killer cells, and acting as immunomodulators for cellular immunity.^{12,13}

During follow-up and observation, calorie intake tended to increase. On day 11 of care, the patient complained of bloating due to not having defecated for the previous 2 days. A radiology examination was performed, revealing an obstructive ileus. Postoperative complications of Hirschsprung disease, such as anastomotic leakage and stricture after definitive pull-through, may have occurred. An ileostomy was performed by the digestive surgeon to release the intestinal obstruction. Parenteral nutrition was administered during the early ileostomy period due to the shorter intestinal length, which decreased intestinal absorption.^{14,15}

In this case, several conditions challenged the patient's improvement, namely severe protein-energy malnutrition, total colectomy, and ileostomy. Along with these complicating

situations, we found it difficult to maintain adequate energy intake due to the total colectomy and ileostomy procedures, which decrease the absorption of macronutrients, micronutrients, electrolytes, and water. Moreover, severe protein-energy malnutrition hindered the effectiveness of nutrition therapy. Severe protein-energy malnutrition combined with surgical procedures can lead to inflammation, a poor prognosis, and potentially death.¹⁶ The implementation of perioperative nutrition therapy in patients with severe protein-energy malnutrition before surgery, such as total colectomy and ileostomy, is mandatory to improve nutritional status and quality of life.¹⁷

Conclusion

An adult with Hirschsprung disease post-colectomy may experience complications such as anastomotic leakage and stricture, which can lead to ileostomy. Both complications can decrease intestinal absorption and exacerbate malnutrition. Medical nutrition therapy should focus on fulfilling nutritional requirements, enhancing nutrient absorption, and reducing inflammation. Inter-professional collaboration between digestive surgeons, clinical nutritionists, nurses, dietitians, and pharmacists is essential to improve outcomes for patients with Hirschsprung disease with surgical procedures.

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