

Serious Short Bowel Syndrome Case Successfully Managed with Home Parenteral Nutrition

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Short bowel syndrome (SBS) is a serious condition that often limits daily activities. This paper reports a case with serious SBS where the residual intestine is limited to the esophagus, stomach, duodenum, sigmoid colon, and rectum (there is no small intestine). Although the patient largely depends on HPN, she has been successfully rehabilitated for 9 years postoperatively and obtained a high QOL. She arrived at our hospital in 1993 in an ambulance due to abdominal pain and hypotension (shock state) after overeating when she was 29 years old. She was diagnosed with general peritonitis and had an emergency operation. After she discharged from hospital, a subcutaneous implantable reservoir port for HPN was placed. She has been allowed to intermittently administer HPN ad libitum with the infusion pump at night. The port had to be replaced twice due to infection and once due to the occlusion of the catheter in 9 years. The following tests were performed to determine her digestion and absorption ability were low, indicating poor digestion. The duodenum and sigmoid colon was enlarged to compensate for the loss of the intestine. For the delivery of infusion preparations for traveling, it is useful to use home delivery services with refrigerated cars for domestic travel and the postal service for travel abroad. The applicable regulations differ between countries.

Introduction

Short bowel syndrome (SBS) is a serious condition that often limits daily activities. It has been reported that SBS patients with residual small intestines of 60 cm or less cannot withdraw from total parenteral nutrition (TPN) even when the large intestine is normal¹⁾, and that children require residual small intestines of at least 20 to 30 cm to withdraw from TPN²⁾³⁾.

This paper reports a case with serious SBS

where the residual intestine is limited to the esophagus, stomach, duodenum, sigmoid colon, and rectum. Although the patient largely depends on home parenteral nutrition (HPN), she has been successfully rehabilitated for 9 years postoperatively and obtained a high QOL. She can even travel abroad on business now.

Case report

Patient: 38-year-old woman

History of current disease: The patient ar-



Fig. 1 Radiographic image of the digestive tract with contrast medium at discharge. The residual intestine is limited to the esophagus, stomach, duodenum, and sigmoid colon.

rived at our hospital in 1993 in an ambulance due to abdominal pain and hypotension after overeating when she was 29 years old. She was diagnosed with general peritonitis and had an emergency operation. The perioperative findings included marked distention of the intestine and necrosis of most parts of the small and large intestines. The intestine was removed from the jejunum at about 5 cm from the Treitz's ligament to the descending colon. The jejunum was treated with jejunostomy and the stump of the sigmoid colon was closed. The patient's condition was serious with postoperative multiple organ failure and sepsis, but she recovered. Then, she had a secondary operation (duodenum-sigmoid colon anastomosis) after her general condition became stable.

Figure 1 shows a radiographic image of her digestive tract with a contrast medium when she was discharged 40 days after admission. Since then, the patient has been receiving nutritional

Table 1 Composition of the infusion preparation

Daily requirements of the basic infusion	
(Commercially available infusion preparation was used)	
Volume of the infusion	2,000 ml/day
Calories taken	1,420 kcal/day
Total nitrogen	8.55 g
Non-protein calorie/N ratio	139
Vitamin complex preparation	2 A/day
H ₂ blocker (cimetidine)	800 mg/day
Aspartate K	15 mEq/day
Auxiliary preparations	
Fat emulsion preparation (10% 200 ml): 2 V/week	
Trace element preparation (Fe, Mg, Zn, Cu, I): 2 A/week	
Extracellular fluid supplement solution	
(Used depending on the amount of sweat)	

support with HPN.

Treatment with HPN: A subcutaneous implantable reservoir port for HPN was placed. She has been allowed to intermittently administer HPN ad libitum with the infusion pump at night. Table 1 lists the components of her current infusion preparation.

A single-bag preparation was selected because of its simple clean operation and stability. An H₂ blocker is administered at 800 mg/day, and trace element and fat emulsion preparations are administered twice weekly. Extracellular fluid is also infused to prevent dehydration during the summer.

Problems and complications after post-discharge :

(1) Immediately after her discharge, the patient could not go out in the morning due to severe collapses, fatigue, and trembling knees. The sudden collapses were eliminated by slowing down infusion for the last hour and reducing the dropping speed by half and then in half again, instead of simply turning off the infusion pump. Since the patient sometimes had very bad diarrhea, she was instructed not to go to places that were not equipped with toilets. She felt very thirsty and had difficulty breathing for some time. Although the patient initially drank 10 liters

or more of water every day, her degree of thirst gradually improved, and she has recently been drinking an average of 3 liters of water every day. Drinking a large amount of water (from 1 to 2 liters) before going to bed was effective to increase her urine. Drinking water in a standing position quickly resulted in diarrhea, but drinking water while lying down seemed to enable the patient to absorb the water well. She discharged urine only after she got out of bed, and had almost no urine discharge in the daytime.

(2) The control of the subcutaneously implantable reservoir port for HPN required that the port had to be replaced twice due to infection and once due to the occlusion of the catheter in 9 years. This was in spite of giving her and her family members thorough instruction in sterile operation procedures including setting up the infusion line, mixing and injecting vitamin compounds, side injections of other drip infusions, physiological saline and heparin locks for catheters, and puncture site disinfection.

(3) Her high medical expenses were handled by successfully applying for welfare for the physically handicapped. She was determined to be physically handicapped because she corresponded to Class 1 of small intestine dysfunction due to the resection of the small intestine (making daily activities difficult) according to the Disabled Person Welfare Act.

Evaluation of organ functions and nutritional condition 9 years after discharge :

(1) Biochemical testing showed no notable abnormalities except for increased transaminase and reduced serum Ca (Table 2).

(2) All the trace elements (selenium, copper, and zinc) and vitamins were in normal ranges except for selenium at 10.1 $\mu\text{g}/\text{dl}$, which was slightly lower than its normal range. The low selenium value was considered to have resulted from the lack of the element in the general trace

Table 2 Blood biochemical test results(2002. 1)

WBC	4,300 / μl	LDH	184 IU/l
RBC	428×10^4 / μl	ALP	338 IU/l
Hct	36.8 %	γ -GTP	70 IU/l
Hgb	12.5 g/dl	T-bil	0.5 mg/dl
Plt	22.3×10^4 / μl	BUN	14.4 mg/dl
TP	6.6 g/dl	Cre	0.77 mg/dl
Alb	3.7 g/dl	Na	137 mEq/l
AST	67 IU/l	K	3.4 mEq/l
ALT	108 IU/l	Ca	7.8 mg/dl
T-cho	94 mg/dl	TG	78 mg/dl

Table 3 Blood concentrations of trace elements

Selenium	10.1 $\mu\text{g}/\text{dl}$
Copper	92 $\mu\text{g}/\text{dl}$
Zinc	77 $\mu\text{g}/\text{dl}$
Folic acid	15 ng/ml
S-Fe	90 $\mu\text{g}/\text{dl}$
TIBC	238 $\mu\text{g}/\text{dl}$
Ferritin	28 ng/ml
T-bile acids	3.3 ng/ml
Vit B ₁₂	680 pg/ml
Fibronectin	520.5 $\mu\text{g}/\text{dl}$

element preparation used. Considering that the measured value of selenium was only slightly lower than its normal range, and that she has not had complications with serious arrhythmia or cardiomyopathy, it is suggested that the duodenum may be involved in selenium absorption, although the duodenum is not sufficient for such patients with extraordinary short intestines as in the present case (Table 3).

(3) Figure 2 shows a radiographic image of the remaining intestine with a contrast medium. The duodenum was probably enlarged to compensate for the loss of the intestine. The retention time of the contrast medium was prolonged. These results also suggest that the duodenum may be involved in absorption.

(4) Although the CT showed moderate fatty liver, it did not show any tendency to become worse.



Fig. 2 Recent radiographic image of the digestive tract with contrast medium
The duodenum is enlarged to compensate for the loss of the intestine.

Digestion and absorption ability 9 years after discharge :

(1) Blood sugar increasing pattern by loading diet shows in Table 4.

(2) D-xylose absorption test (25 g) was 2.8 g/5 hr (normal: 5~8 g/5 hr). This result indicates very poor absorption.

(3) The following tests were performed to determine her digestion ability. The results obtained were low, indicating poor digestion. Fecal chymotrypsin measurement was 39.8 U/g (normal: >5 U/g). Fecal Sudan 3 staining was negative. PFD test (BTPABA test) was 26.3% (normal: 6 hr 73.4~90.4%).

(4) Bone mineral quantitative determination test (YAM) : The test result was 78%, indicating reduced bone mineral. The reduced bone mineral may have been induced by calcium deficiency due to long-term digestion and absorption failure, hepatopathy, and HPN effects including excessive amino acids, sodium load, and hyperglyce-

Table 4 Blood sugar increasing pattern

Loading diet	Blood sugar (g/dl)	Insulin * (U/ml)	C-peptide ** (ng/ml)
Before	70	3.4	3.9
30 min	88	7.8	4.6
60 min	96	7.3	4.8
120 min	94	11.0	4.8
180 min	62	10.0	5.4

* Insulin was measured as immunoreactive insulin (IRI). (normal: >17 U/ml)

** C-peptide was measured as C-peptide immunoreactivity (CPR). (normal: 0.6 ~ 2.8 ng/ml)

mia.

History of extending daily activities after discharge :

(1) Table 5 shows the medical examinations and drug prescriptions for 1 year after discharge. The patient recovered 1 year after discharge and had extended her daily activities. At that time, she only had to be careful to prevent dehydration. Although it was difficult to deliver a large amount of infusion preparations, she can now use a home delivery service that keeps them cool and delivers them at the appointed time. Using this service, she can go anywhere in Japan.

(2) Tables 6 and 7 list the problems associated with traveling abroad on business. She always uses the postal service to send her infusion preparations because it is easier and quicker to deliver items requiring customs procedures. The preparations are packed into an ordinary box, which has an invoice listing the contents. A "Handle with care" label is attached. Since it is prohibited to send sterilizing ethanol abroad or even carry it into the air plane, she always uses disposable alcohol cottons.

Discussion

Short bowel syndrome (SBS) is defined as malabsorption associated with a massive resection of the small intestine. Although there is a standard of residual small intestine of 150 cm or

Table 5 History of extending her daily life

From discharge to now	Medical examination and drug delivery	Physical condition
Discharge ~ 1 month Field of activity : home ~ hospital	Medical examination : once a week Drug delivery : about 22 kg for 1 week A family member had to stay with her for every medical examination, burdening her family.	Easily tired and vertigo Sudden vomiting
1 month ~ 1 year Within 1-hour trip by train	Medical examination : twice a month The preparation was prescribed outside the hospital. Home delivery service with refrigerated cars	hypoglycemia, vomiting, thirst frequent diarrhea
1 year after discharge and later She became able to stay out overnight.	Medical examination : once a month Home delivery service with refrigerated cars : twice a month Pharmacist : twice a month	Care only for dehydration

Table 6 HPN practice abroad

A) Delivery of infusion preparations
1) When the patient carries them for themselves while traveling by air: The suitcase containing the preparations should be packed in a large plastic bag. The preparation and preparation pump can be carried and used on the airplane.
2) When the preparations are delivered: It is necessary to have a place where they are to be delivered and to tell someone there how to take care of them. It is useful to use the postal service to deliver them abroad.
3) Disinfecting ethanol It is prohibited to send disinfecting ethanol abroad or to carry it on airplanes.
B) To secure storage conditions for the preparation
C) Usage environment for the infusion pump Power transformers can be used.
D) Communication between physicians and pharmacists in case any problems occur A medical history sheet in English should be prepared beforehand to present to physicians abroad.

Table 7 Problems associated with delivering the infusion preparations abroad

1) Treatment of the infusion preparation The delivery of the infusion transfusion is not regarded as an import. Therefore, it is not necessary to obtain import approval. It is necessary to attach an invoice listing the preparation contents to the box containing it.
2) Explaining the difference between the preparation, stimulants, and other substances at the customs office The invoice is enough to explain that the infusion preparation is not a stimulant or other such substance. The package insert of the preparation should be carried in order to explain that it is essential nutrition necessary for medical reasons, and that it must be used every day.
3) Protecting privacy Attaching a medical certificate (the problem of where the certificate will be opened)
4) Disinfecting ethanol It is prohibited to send disinfecting ethanol abroad or to carry it on airplanes. Although sending disinfecting ethanol is prohibited, sending alcohol cotton is possible.

less for adults and 75 cm or less for children⁴⁾, the “massive” or “extensive” resection should not

be examined with the length of the residual intestine, but with the ratio of the resected or remain-

ing intestine to the whole intestine. Patients with SBS with residual small intestines of 60 cm or less are considered to have difficulty in withdrawing from TPN even when their large intestines are normal, and they rarely survive long. Therefore, the present case is very rare as she has survived for 9 years in spite of serious SBS in which the small intestine was resected and only the sigmoid colon remains in the large intestine.

HPN is indicated for patients who have stable primary diseases, can only take nutrition with parenteral nutrition, and cannot be controlled with oral or enteral nutrition. The guidelines of the Japanese Study Group for Intravenous and Enteral Nutrition specify the following conditions as the indications for HPN : ① massive resection of the intestine, ② some malignant tumors under chemotherapy, ③ inflammatory intestinal diseases, such as Crohn's disease, ④ intestinal dyskinesia, such as chronic idiopathic pseudo-obstruction, ⑤ radiation enterocolitis, ⑥ digestion and absorption disorder syndrome, ⑦ intractable diarrhea, ⑧ sclerotic peritonitis by CAPD.

A diet load test to examine the pattern of the increase in blood sugar (blood sugar, insulin concentration, and C-peptide) and a D-xylose absorption test (25 g) were performed in order to investigate digestion and absorption. Fecal chymotrypsin was measured and fecal Sudan 3 staining and a PFD test (BTPABA test) were performed to investigate digestion. The results showed all the values obtained were low, indicating that the digestion and absorption of the residual intestine was very poor. Bone mineral density has been measured over time to investigate possible osteopathy. The results of the bone mineral quantification test (YAM) in these several years have been stable at around 78%. Although bone mineral is low, there is not any tendency for it to become worse.

Nutritional therapies consisting only of chemi-

cally pure substances, such as total parenteral nutrition (TPN) or elemental diet (ED), have been widely applied to diseases associated with digestion and absorption disorders. As the patients chronically treated with these nutritional therapies have increased, an increasing number of studies have reported deficiencies in various trace elements such as zinc, copper, selenium, and chromium, resulting in the investigators' attention to the nutritional importance of these trace elements in humans⁵⁾.

The reduced level of selenium, a trace element contained in grain, meat, and fish, has been reported to induce serious arrhythmia and cardiomyopathy. Selenium is considered to be an antioxidizing agent that neutralizes the effect of free-radicals. Selenium deficiency may damage intracellular genetic materials and induce carcinogenesis⁶⁾. Copper and zinc are included in the general trace elements, while selenium is not. Therefore, some investigators recommend administering a selenious acid injection (2.5 $\mu\text{mol/l}$) and using fresh frozen plasma (FFP)⁶⁾.

Although it is possible that the enlarged duodenum compensating for the loss of the intestine may be involved in absorbing selenium in such patients with severe SBS such as the present case, the patients should be strictly followed for a long time for the deficiency of trace elements or unknown substances that cannot be supplemented by commercially available trace element preparations.

To appropriately control the present SBS case with TPN, the infusion preparation used was selected considering preventing fatty liver. It is normally thought that the preparation for SBS should have the following specifications: a non-nitrogen-sourced calorie/nitrogen ratio of 180 to 200; energy administered 0.9 to 1.1 \times BEE; and amino acids from 0.9 to 1.3 g/kg/day. A single bag preparation was chosen for simple clean op-

eration and stability. The preparation is a mixture of a basic solution and amino acid preparation in which the Meillard reaction from the mixture of glucose and amino acid is prevented by adjusting the pH and adding lactate. The preparation contains 35 mEq/l of L-lactate and is adjusted to have a low pH from 3.4 to 4.8. Although the patient had a mild increase in transaminase and fatty liver as demonstrated by the CT for these 9 years, these increases will be carefully followed without specific treatments because neither increase showed any signs of exacerbation.

Important complications in controlling SBS include hepatic insufficiency associated with TPN and catheter infection associated with HPN. Hepatic insufficiency associated with TPN occurs due to the complex mechanism based on various factors including fatty degeneration from excessive nutrition, methionine dysbolism, abnormal proliferation of Enterobacteriaceae, cholestasia mainly from fasting, and hepatopathy due to inflammatory mediators derived from the bacterial translocation by enteritis⁷⁾.

Since the present case is the first case treated only with TPN, it is necessary to provide nutrition while paying full attention to the possible development of hepatic insufficiency. Catheter infection can occur through 2 routes: one is exogenous infection from the catheter puncture site, infusion line, or infusion preparation, and the other is endogenous infection from bacterial translocation etc. Since some studies have reported that catheter infection is particularly likely to occur in patients with SBS⁸⁾⁹⁾, it is important to prevent endogenous infection. Giving thorough instruction to patients and their families is important to appropriately manage HPN. Important procedures include sterile operation procedures such as setting up the infusion line, mixing vitamin preparations, side injections of other drip infusions, physiological saline and heparin locks of

catheters, disinfection of the puncture sites, and emergency actions for circulation disorders from dehydration and catheter infections.

For the delivery of infusion preparations for traveling, it is useful to use home delivery services with refrigerated cars for domestic travel and the postal service for travel abroad. The applicable regulations differ between countries. The delivery of infusion preparations, for example, has not been approved in Germany, but the situation is improving.

In the future, while maintaining the reliable relationship, we will try to closely investigate the cause of hepatopathy with a histological examination and liver biopsy, and to examine the condition of the intestinal villi with a biopsy of the duodenum and large intestine.

Conclusions

(1) We examined a long-term survivor with very serious short bowel syndrome in whom the remaining intestine is limited to the esophagus, stomach, duodenum, sigmoid colon, and rectum.

(2) She has been able to have a good social life with home parenteral nutrition. She can even travel abroad on business.

(3) Although her intestinal digestion and absorption are very poor, her nutritional condition has been maintained relatively well with a commercially available infusion preparation.

(4) It is useful to use home delivery services with refrigerated cars for domestic travel and postal services for travel abroad in order to have the infusion preparation while away from home.

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在宅静脈栄養 (Home Parenteral Nutrition: HPN) が著効している 重度短腸症候群 (Short Bowel Syndrome: SBS) の1例

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短腸症候群は大腸が正常でも残存小腸が60cm以下では完全静脈栄養(TPN)からの離脱は困難とされる重症病態で、それにともない日常生活範囲も制限されることが多い。我々は残存腸管が食道・胃・十二指腸・S状結腸・直腸の重度短腸症候群で、在宅静脈栄養(以下HPN)に大部分が依存しているが、術後9年間にわたり海外出張をこなすまで社会復帰しているQOLの高い1症例を経験したので報告する。患者は38歳、女性。29歳時、過食後の腹痛・血圧低下により救急搬送され、汎発性腹膜炎の診断で緊急手術を施行した。2期的手術の後、現状となり退院後はHPNによる栄養管理を行い現在に至っている。HPNは皮下埋め込みリザーバーポートを留置し、輸液ポンプを用いて夜間間歇的投与で自由摂食としている。輸液は清潔操作の簡便性と製剤の安定性よりシングルバッグ製剤を用いており、H₂-blockerを800mg/日、微量元素製剤および脂肪乳剤を週2日投与している。また夏季には脱水予防のため細胞外液輸液を追加投与している。退院後9年目の現在、代償性に肥大した十二指腸、軽度骨密度低下を認めるが、栄養状態は良好である。合併症としてTPNの観点からの肝機能障害(脂肪肝)とHPNの観点からのカテーテル感染症があるが、どちらも重症化することなく対処している。国内の長距離移動にはクール宅急便を、海外には郵便輸送によって輸液製剤を搬送しているが、大きなトラブルもなく良好な社会生活を営んでいる。