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Surgical Approach to Short-Bowel Syndrome

Experience in a Population of 160 Patients

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Objective
The authors reviewed their experience with short-bowel syndrome to define the surgical approach to this problem in 160 patients.

Methods
Forty-eight adults and 112 children were evaluated over a 15-year period.

Results
Seventy-one patients (44%) adapted to resection and took enteral nutrition alone. Forty-four patients (28%) were supported by parenteral nutrition (PN). Forty-five patients (28%) have had 49 surgical procedures. Fifteen patients with adequate intestinal length (>120 cm in adults) but dilated dysfunctional bowel underwent stricturoplasty (n = 4) or tapering (n = 11). Thirteen patients (87%) demonstrated clinical improvement. Fourteen patients with shorter remnants (90–120 cm) and rapid transit time received an artificial valve (n = 2) or a reversed segment (n = 1). All patients' conditions improved initially, but the reversed segment was revised or taken down. Fourteen patients with short remnants and dilated bowel underwent intestinal lengthening. Twelve patients' conditions improved (86%), one underwent transplantation, and one died. Sixteen patients with very short remnants (<60 cm) and complications of PN underwent solitary intestine (n = 4) or combined liver–intestinal transplantation (n = 13). One-year graft survival was 65%. There have been five deaths.

Conclusions
The surgical approach to short-bowel syndrome depends on the patient's age, remnant length and caliber, intestinal function, and PN-related complications. Nontransplant procedures have a role in the treatment of selected patients. Intestinal transplantation is emerging as a potential therapy for patients with significant PN-related complications.

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Extensive resection of the small intestine results in malabsorption and diarrhea, a clinical syndrome termed "short-bowel syndrome." The intestinal remnant undergoes structural and functional adaptation in the months after resection, which may permit maintenance of nutritional status by enteral feedings alone. Adapta-

Table 1. CONDITIONS NECESSITATING EXTENSIVE INTESTINAL RESECTION IN ADULT PATIENTS

Condition	No.
Irradiation/cancer	17
Mesenteric vascular disease	10
Crohn's disease	5
Other benign disease	16
Total	48

Table 2. CONDITIONS NECESSITATING EXTENSIVE INTESTINAL RESECTION IN PEDIATRIC PATIENTS

Condition	No.
Necrotizing enterocolitis	37
Midgut volvulus	36
Intestinal atresia	23
Other benign disease	16
Total	112

tion is influenced by many factors, including site and length of the intestinal remnant, presence of the ileocolonic junction (ICJ), underlying intestinal disease, age of the patient, and status of the other digestive organs. However, many patients with short intestinal remnants may remain dependent on parenteral nutritional support.

Although long-term survival is possible in patients with short-bowel syndrome who receive parenteral nutrition (PN), the expense and morbidity of this therapy has maintained interest in various surgical options.[1] Intestinal transplantation and various restorative procedures have been used.[1,2] These procedures have been associated with significant potential morbidity and therefore need to be used judiciously. Our aim in the current study was to define a surgical approach for patients with short-bowel syndrome based on our experience with a population of 160 patients.

METHODS

We retrospectively reviewed the records of 48 adults and 112 children with short-bowel syndrome evaluated at the University of Nebraska Medical Center (University Hospital, Children's Hospital, and Omaha Veterans Administration Medical Center) between 1980 and 1994. Adult patients were included if they had less than 180 cm of small intestine remaining with evidence of malabsorption. Pediatric patients (younger than age 16 years) were included if they had intestinal remnant less than 120 cm and/or required PN support on discharge from the hospital.[3,4] Patients who died in the hospital after massive resection were excluded from the study.

There were 48 adult patients with short-bowel syndrome, ranging in age from 16 to 92 years. The conditions necessitating resection are shown in Table 1. The most frequent indication for resection was cancer and its treatment with irradiation. Eleven (23%) patients were older than age 70 years. The remnant length was less than 60 cm in 15 patients, 60 to 120 cm in 13 patients, and 120 to 180 cm in 20 patients. These patients have been followed up for a mean of 51 months, with 26 (54%) being followed up for more than 5 years.

There were 112 pediatric patients with short-bowel syndrome, ranging in age from newborn to 14 years. The conditions necessitating resection are shown in Table 2, of which necrotizing enterocolitis and volvulus were the most common. All but three patients developed short-bowel syndrome in the 1st year of life. Two of the patients who underwent transplantation had near-total enterectomy performed at the time of intestinal transplantation. Remnant length was less than 30 cm in 43 patients, 30 to 60 cm in 23 patients, and more than 60 cm in 46 patients. These patients have been followed up for a mean of 70 months, of whom 63 (56%) have been followed up for more than 5 years.

Surgical procedures for short-bowel syndrome included only those designed to improve intestinal function (Fig. 1). Ostomy formation and procedures designed to restore intestinal continuity were excluded. "Intestinal tapering" refers to reduction of the circumference of the intestine by either imbrication or excision of redundant bowel wall along the antimesenteric border. Stricturoplasty was performed by means of a Heineke-Mikul-

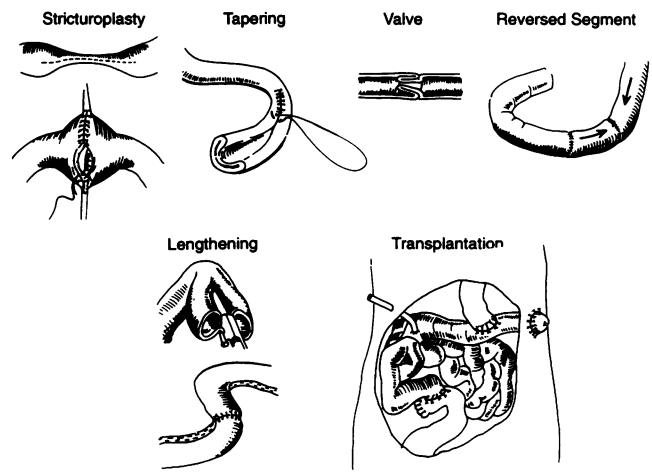

Figure 1. Surgical procedures for improvement of intestinal function in short-bowel syndrome.

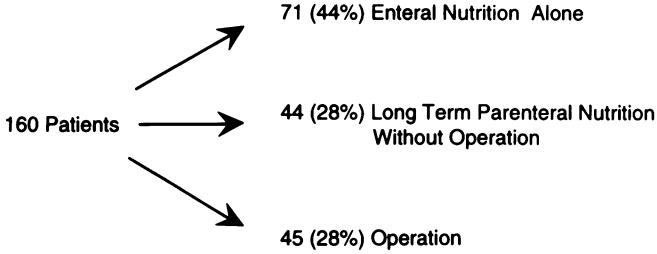

Figure 2. Outcome of short-bowel syndrome.

icz–type enteroplasty at a stenotic region of the intestine. Artificial intestinal valves were created by distal intussusception of a segment of small intestine. Intestinal lengthening involved longitudinal transection of the intestine between the mesenteric and antimesenteric edges and anastomosis of these parallel intestinal segments, as described previously.[5] Intestinal transplantation, either solitary or combined with liver transplantation, was performed with immunosuppression that involved either cyclosporine or tacrolimus.

Statistical comparisons were made with a chi square test, and a probability level of less than 0.05 was considered significant.

RESULTS

Overall, 71 patients (44%) adapted sufficiently to the intestinal resection to maintain adequate nutrition with enteral feeding alone (Fig. 2). This occurred more often in children compared with adults (51% *vs.* 29%, $p < 0.05$). Forty-four patients (28%) were maintained on long-term PN without attempted surgery. Ten of these patients are awaiting transplantation. Forty-nine operative procedures have been performed on 45 patients to improve intestinal function (Table 3). Children were more likely to undergo operation compared with adults (33% *vs.* 17% $p < 0.05$).

Table 4. FACTORS AFFECTING MANAGEMENT OF ADULT PATIENTS

	N	Enteral Nutrition Alone	Require Parenteral Nutrition	Operation Performed
Remnant length				
<60 cm	15	0*	14	1
60–120 cm	13	5	4	4
120–180 cm	20	9	8	3
Ileocecal junction				
Present	6	2	1	3*
Absent	42	12	25*	5
Patient age				
<70 yr	37	11	20	6
>70 yr	11	3	6	2
Disease				
Mesenteric vascular	10	3	7	0*
Irradiation/cancer	17	6	9	2
Other benign	21	5	10	6

* $p < .05$ *vs.* others.

Ten of the adult patients (21%) have died. One patient died of candidal sepsis related to long-term PN. The other deaths were related to underlying malignant or cardiac disease. Fourteen pediatric patients (13%) died: five of liver disease while awaiting transplantation, six after surgery, and three of other underlying conditions.

The management of disease in adult patients was determined by remnant length, presence of the ICJ region, and underlying disease (Table 4). Adults with less than 60 cm of intestine and those with an absent ICJ were significantly less likely to be maintained on enteral feed-

Table 3. RESULTS OF OPERATIVE TREATMENT

	Valve or Segment	Stricturoplasty	Tapering	Lengthening	Transplantation	Total
N	3	4	11	14	17	49
Adults	2	4	0	1	1	8
Remnant length						
<30 cm	0	0	2	5	14	21
30–60 cm	0	0	4	5	3	12
>60 cm	3	4	5	4	0	16
Preoperative nutrition support						
Enteral only	0	1	1	1	0	3
Parenteral	3	3	10	13	17	46
Postoperative nutrition support						
Enteral only	2	4	9	7	10	32
Decreased PN	1	0	2	5	2	10
Parenteral	0	0	0	2	5	7

PN = parenteral nutrition.

ings alone. Patients with an intact ICJ were more likely to undergo surgery whereas those with mesenteric vascular disease were less likely to undergo operation. Remnant length was also an important factor among pediatric patients (Table 5). Children with less than 30 cm of intestine were less likely to adapt to enteral nutrition alone compared with those with remnants longer than 30 cm. Patients with a remnant shorter than 30 cm were more likely to be sustained on enteral nutrition if they had an intact ICJ (50% with ICJ *vs.* 14% without, $p < 0.05$). Overall, patients with an intact ICJ were more likely to adapt enterally ($p < 0.05$). Pediatric patients with a remnant longer than 60 cm were less likely to undergo operation, as were those with necrotizing enterocolitis. The majority of nontransplant operations among children were related to dilated intestinal segments and bacterial overgrowth.

Four adults and 11 children with adequate intestinal remnant length (>120 cm in adults) but dilated bowel underwent surgery to improve intestinal function. The adults had stenotic segments causing partial obstruction and underwent stricturoplasty. Three of these adult patients required PN before surgery, and all four experienced improvement and were able to sustain themselves enterally after operation. The 11 children, age 6 months to 9 years, had dilated segments (>3 cm in diameter) with remnant lengths longer than 30 cm. All of these children with dilated segments had associated bacterial overgrowth and malabsorption, but only three had mechanical obstruction. Enteroplasty was performed on the duodenum (n = 3), jejunum (n = 3), or ileum (n = 5). The length of the tapered segment was longer than 15 cm in 7 of the 11 patients. Tapering enteroplasty resulted in improved function transiently in all patients (Table 3).

Table 5. FACTORS AFFECTING MANAGEMENT OF PEDIATRIC PATIENTS

	N	Enteral Nutrition Alone	Require Parenteral Nutrition	Operation Performed
Remnant length				
<30 cm	43	11*	13	19
30–60 cm	23	8	4	11
>60 cm	46	38	1	7*
Ileocecal junction				
Present	41	30*	2	9
Absent	71	27	16	28
Disease				
NEC	37	25*	9	3*
Volvulus	36	11	6	19
Other	39	21	3	15

NEC = necrotizing enterocolitis.
* $p < 0.05$ *vs.* others.

However, two patients subsequently underwent intestinal lengthening because of recurrent malabsorption. Follow-up ranged from 2 to 10 years, with 5 patients (45%) followed up for more than 5 years.

Two adults and one child with shorter remnants (90–120 cm) and rapid transit time received an artificial valve (n = 2) or a reversed segment (n = 1). An adult with a 100-cm remnant had an intussuscepted valve placed just proximal to her jejunostomy. Her ostomy output decreased and she was able to discontinue PN after 2 months. A 3-year-old child with a 90-cm remnant underwent valve creation combined with tapering and has remained off of PN for 4 years. A 10-cm reversed segment was placed just proximal to a jejunostomy in another adult with a 120-cm remnant. Her ostomy output decreased, but persistent nausea and vomiting led to revision of this segment 14 months later.

One adult and 13 children, 3 months to 11 years of age, with short remnants (<90 cm) and dilated bowel (>4 cm) underwent intestinal lengthening. Segments ranging from 5 to 35 cm were lengthened; 9 patients had their intestines lengthened by at least 20 cm. Nutritional status was improved for 12 of these patients (86%) (Table 3). The 3-month-old died of pneumonia during the same hospitalization. Another child continued to require PN and subsequently underwent intestinal transplantation. One child required an additional tapering enteroplasty. Follow-up ranged from 1 to 13 years, with 6 patients (43%) followed up for more than 5 years.

Sixteen patients with very short remnants (<60 cm in adults) and either liver disease or other complications of PN underwent solitary intestinal transplantation (n = 4) or combined liver–intestinal transplantation (n = 13). An adult patient who initially had a combined liver–intestinal transplantation underwent retransplantation after excision of an ischemic intestinal graft. The children ranged in age from 8 months to 9 years, of whom 5 (33%) were younger than 1 year of age. Remnant length was less than 30 cm in 12 patients (80%). The 1-year actuarial patient and graft survival rates were 81% and 65%, respectively. Among the combined transplantations, one allograft was lost to rejection (cyclosporine had been administered) and another was removed as a result of cytomegaloviral enteritis. The five deaths were related to fungal or viral sepsis. Four patients died soon after surgery, but the other patient died 18 months later, and this death was associated with medication noncompliance. One of the patients who died had previously undergone liver transplantation elsewhere and developed recurrent PN-induced liver disease. All three children who underwent isolated transplants were able to discontinue PN within 2 weeks and are at home on enteral feed-

ings. Follow-up ranged from 3 months to 40 months, with 11 patients (69%) having been followed up for more than 12 months.

DISCUSSION

Home PN prolongs life in patients with short-bowel syndrome but is not the ideal solution for patients with anticipated long-term survival. Long-term PN is associated with significant morbidity and mortality, costs as much as $100,000 per year, and affects overall quality of life.[6-10] Our medical center's mortality rate for PN-related complications compares favorably with that reported by others.[3,4,11,12] Overall, 20% of patients developed liver disease, which reflects, in large part, the referral nature of the patient population. Fortunately, the majority of patients, particularly children, adapt and are able to maintain themselves on enteral feedings alone, although life-style may not be optimal due to the necessity of feeding tubes, dietary restrictions, and ostomies.[13] In the current study, 44% of patients were taking enteral nutrition alone. An aggressive approach to enteral feeding may not only hasten adaptation but also increase the proportion of patients no longer requiring PN.[14,15] Recent efforts at stimulating adaptation with various growth factors may also result in a larger proportion of patients maintained on enteral nutrients alone.[16]

Intestinal remnant length and the presence or absence of the terminal ileum and ICJ continue to be important prognosticators of nutritional therapy.[17] Adults with less than 60 cm of intestine remaining and those in whom the ICJ was absent were more likely to require long-term PN. Patient age and underlying disease were less important factors in nutritional outcome. Our findings are consistent with the observation made by Gouttebel et al.[17] that permanent PN is likely if the remnant length is smaller than 30 cm with the ICJ or smaller than 60 cm without.

The significance of remnant length as a predictor of nutritional outcome in infants and children is more controversial. Before the availability of long-term PN support, Wilmore[18] found that infants died if they had less than 15 cm of intestine remaining with the ICJ and less than 40 cm without it. Use of PN support with aggressive enteral feeding permits long-term survival of infants with as little as 10 cm of intestine with the ICJ and 25 cm without it.[12,19,20] Many infants with these short remnants will ultimately be sustained on enteral feedings alone. Twenty-six percent of our patients with a remnant smaller than 30 cm adapted to enteral nutrition alone, and 64% of these patients had an intact ICJ. Remnant length and presence of the ICJ predict duration of PN among these patients.[15] The poor prognosis of infants with an initial remnant length smaller than 10 cm has prompted some to recommend withholding PN therapy

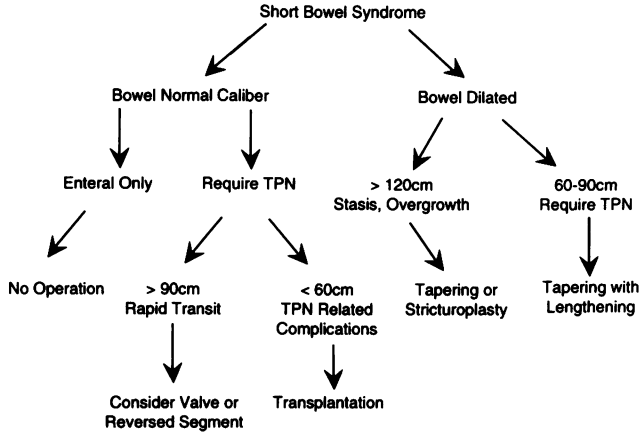

Figure 3. Surgical management of short-bowel syndrome.

in these individuals.[21] However, Kurkchubasche et al.[19] suggested that a remnant smaller than 10 cm warrants early small-bowel transplantation. Seven of our patients who underwent transplantation had remnants in this range.

Nontransplant surgical procedures designed to improve intestinal function have historically produced unpredictable results with significant morbidity.[1] Thus, candidates for these procedures should be in relatively good health otherwise and should benefit greatly if the procedure is successful. We generally reserve such operations for patients who have a high likelihood of discontinuing PN as a result of the procedure. Because patients with short-bowel syndrome often require reoperation for other problems, adjunctive procedures should be considered at that time.[22] Patients at the extremes of age and those with significant underlying disease would not be good candidates. Adult patients with mesenteric vascular disease were unlikely to receive surgery. In the current study, children were more likely to undergo surgery than adults, due in part to the more frequent occurrence of PN complications and dilated intestinal segments. However, three (50%) of our procedure-related deaths involved children younger than 12 months of age.

Overall, 84% of the nontransplant procedures performed in the current study led to long-term clinical improvement. After surgery, 69% of patients were maintained on enteral nutrition, and 25% required less PN. However, there was one death, and 16% patients required another procedure because the initial benefit was not sustained. Operations aimed at prolonging intestinal transit time are especially inconsistent. Only two of our three operations for this purpose were successful. However, when patients have defined problems, such as strictures or poorly functioning dilated segments, the results of surgical procedures are better. We have found stricturoplasty and tapering enteroplasty to significantly improve intestinal function in 87% of patients. Intestinal

lengthening led to improved intestinal function and nutritional status in a similar proportion of patients (86%). The success of these tapering and lengthening procedures has made the caliber of the intestinal remnant an important determinant of surgical therapy. We have not attempted staged procedures to increase intestinal diameter to achieve increased intestinal caliber, however.[23]

Substantial progress has been made in the area of intestinal transplantation in recent years, with improved immunosuppression, graft preservation, and technical advances.[2] The majority of our patients underwent combined liver–intestinal transplantation for PN-induced liver disease. Although there is significant associated morbidity and mortality, this procedure offers the best chance for return to function. Recently, Lawrence and coworkers[24] suggested that liver transplantation alone may be applicable to patients with PN-induced liver failure and short-bowel syndrome. However, overestimating the adaptive capability of the small intestine in this situation has a penalty. One of our patients who underwent liver–intestinal transplantation had previously undergone liver transplantation alone at another institution and again developed PN-induced liver disease within 6 months. He died after undergoing combined transplantation.

More recently, we have performed solitary intestinal transplantation in patients with problems maintaining vascular access or with impending liver failure. This is the most appealing approach to short-bowel syndrome. Our success with these patients suggests that this procedure should be applied sooner to prevent the development of liver disease. Because most of our PN-dependent patients have remnants smaller than 30 cm, isolated intestinal transplantation is likely to have a greater effect on management than would nontransplant procedures.

On the basis of our experience with this large group of patients with short-bowel syndrome, we have developed a management scheme for surgical therapy (Fig. 3). The surgical approach to patients with short-bowel syndrome depends on the patient's age, remnant length and diameter, intestinal function, and PN-related complications. Nontransplant procedures have a role in selected patients for improvement of intestinal function. Intestinal transplantation is emerging as a potential therapy for patients with significant PN-related complications.

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Discussion

DR. JOSEF E. FISCHER (Cincinnati, Ohio): This is a good time to review experiences with short-bowel syndrome. Obviously, Dr. Starzl's paper on Thursday and the success of small-