

Case Report

The Right Colon Patch Graft Procedure for Extensive Intestinal Aganglionosis

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Extensive intestinal aganglionosis is rare and very difficult to diagnose and treat. The condition is often fatal. A 6-month-old boy who had undergone ileostomy for extensive intestinal aganglionosis was referred to our department. We applied an aganglionic right colon onlay patch to the aganglionic intestine to enhance absorption of water and electrolytes. Three months after the ileocolostomy, the definitive operation, a Swenson-type procedure, was performed. The mesocolon to the onlay patch could be divided because blood supply was adequate from the ileal mesentery via the intestinal wall. Postoperatively, the onlay patch segment appeared normal on colonoscopy and bowel habit was improved. Although the patient still requires parenteral nutrition support due to the short bowel, the right colon onlay patch procedure enables him to be cared for at home and provides an opportunity for normal growth and development.

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Introduction

Aganglionosis involving the entire colon occurs in approximately 10% of patients with Hirschsprung's disease.^{1,2} Extensive aganglionosis involving the small intestine (extensive intestinal aganglionosis or EIA) is thought to be much rarer, affecting less than 5% of such patients.³ It is characterized by the absence of ganglion cells extending from the anus to the lower small intestine. Diagnosis and treatment are relatively difficult,⁴ and morbidity and mortality in children suffering from this disease remain high because of chronic intestinal obstruction, enterocolitis and the requirement for long-term total parenteral nutrition. Unfortunately, complete excision of the involved colon and small intestine has resulted in intractable diarrhea and electrolyte imbalance. Martin⁵ first reported preservation of the aganglionic left colon as a longitudinal side-to-side anastomosis to the ganglionic small intestine. However, there are several reports of associated enterocolitis,⁶⁻⁹ and laboratory studies using the right colon demonstrated superior reabsorption of water and sodium.^{10,11} Kimura et al.¹² and Boley¹³ advocated the use of aganglionic right colon as an antimesenteric patch.

Twenty years ago we performed the left colon onlay patch procedure on a 1-year-old boy with EIA who had suffered from intractable diarrhea, enterocolitis, melena and anastomotic ulcer between the colon and ileum. We report here the second EIA patient we experienced, for whom we used a right colon patch for the first time.

Case Report

A 3200 g male infant was delivered vaginally at 38-week-and-2-day gestation at another medical center. Prenatal ultrasonography raised a suspicion of intestinal atresia with polyhydramnios. One day after his birth, surgical exploration was performed for vomiting and abdominal distension, revealing obstruction in ileum accompanied by torsion, and ileoileostomy was performed. Subsequently, abdominal distension was prolonged and second laparotomy was performed 10 days after his birth. This revealed narrowed ileum at the anal side of the ileal anastomosis. A loop ileostomy was created at the ileal anastomosis, but bowel obstruction was not improved. Thirty-three days after his birth, he required ileal resection and ileostomy

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above the caliber change at 55 cm from the duodenojejunal flexure. Histological study of the resected specimen showed normal ganglion cells. Postoperatively, he received parenteral nutrition and mother's milk.

Six months after his birth, he was referred to our department for further surgical treatment. Since the diagnosis of aganglionic was not definite then, laparotomy was performed and multiple frozen sections were taken from the rectum, transverse colon, ascending colon and terminal ileum. Ganglion cells were absent in all regions except the proximal side of the ileostomy. The length from the duodenojejunal flexure to the initial ileostomy had extended to 80-cm length from 55 cm at 33 days after his birth. He was diagnosed with EIA extending to 50-cm length from ileocecal valve. Eight months after his birth, we performed an isoperistaltic side-to-side anastomosis of the 3-cm aganglionic ileum proximal to the ileocecal valve and 10-cm right colon to the distal end of the ganglionic ileum, creating an approximately 13-cm long onlay patch as seen in Figures 1 and 2.

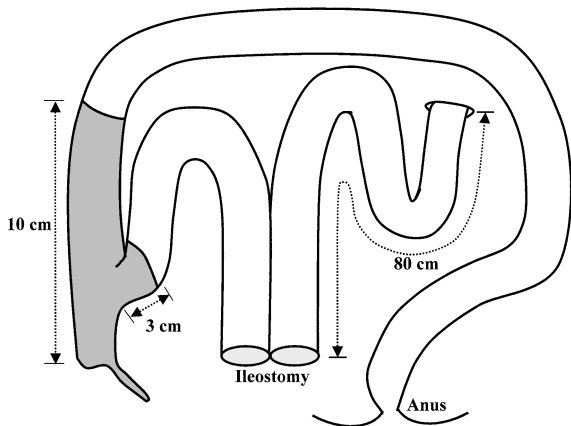


Figure 1. The aganglionic ascending colon (10 cm) and terminal ileum (3 cm) were used for onlay patch graft. The ileostomy was placed 80 cm from the duodenojejunal flexure.

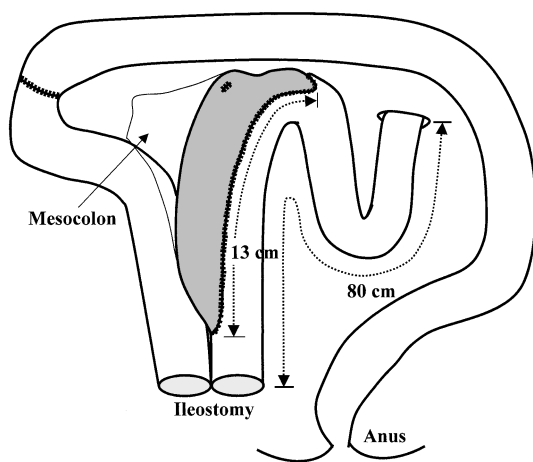


Figure 2. The right colon onlay patch including the terminal ileum was attached to the ganglionic ileum in side-to-side fashion, preserving the ileocolonic vessels.

The anastomosis was performed at both antimesenteric sides with autostapling devices (Endo-TA60 stapler for 60 mm×2 mm staples; Auto Suture Company Division, United States Surgical Corporation, Norwalk, CT) and interrupted uncoated polyglactin 910 braided surgical sutures (Ethicon Inc., Somerville, NJ) to the outer layer, as shown in Figure 3. The right colon and terminal ileum were supplied blood from the ileocolic and right colic arteries. Postoperatively, diarrhea from the ileostomy was decreased to 300-800 mL/day from 900-1300 mL/day observed preoperatively.

The definitive operation was carried out 3 months after the creation of ileo-colonic patch graft. The ileocolostomy was mobilized after severing the mesocolon attached to the colonic patch graft. After the confirmation of adequate blood supply to the colonic-patched portion, the distal colon and rectum were resected. The distal end of the ileocolostomy segment with ileal spout was anastomosed to the end of the rectum in a video-assisted Swenson-type procedure, as shown in Figure 4. The posterior wall of the aganglionic rectum was di-

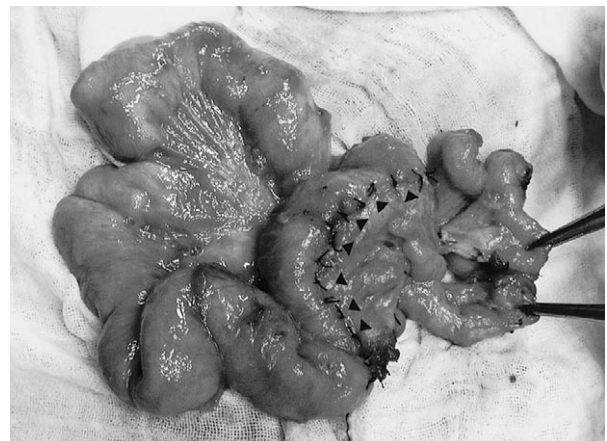


Figure 3. Closed triangles (▲) show side-to-side anastomosis between the right colon onlay patch and the ganglionic ileum.

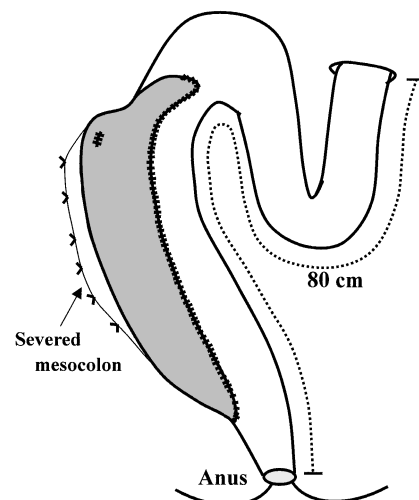


Figure 4. The ileocolostomy dissecting the mesocolon was pulled through to the anus.

vided to the level of the dentate line to prevent postoperative anal achalasia. The patient was in the hospital for 80 days after this definitive operation because of abdominal distension and obstinate diarrhea. Enterocolitis due to functional obstruction of sphincter was suspected; however, it was subsequently relieved by daily rectal irrigation and manual stretching of the sphincter. Colonoscopy performed 6 months after the operation demonstrated normal ileal and colonic mucosa and no ulceration in the ileocolostomy (Figure 5). Since 12 months after the operation, he has had 5 to 6 bowel movements per day and has been growing normally. Because of short residual jejunum and ileum, oral and home parenteral nutritional assistance has been required.

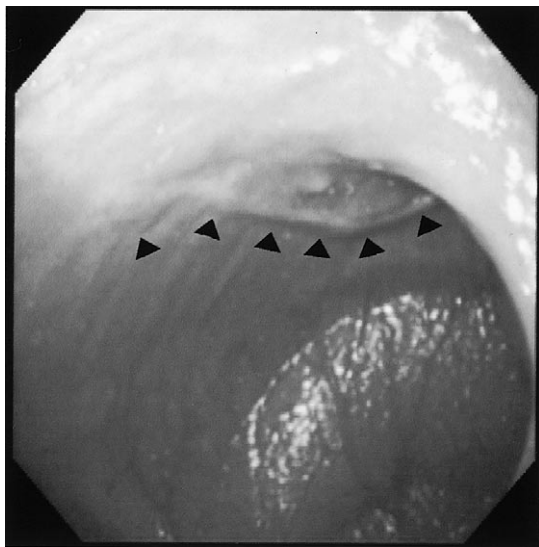


Figure 5. Colonoscopic findings of the ileocolostomy segment. Closed triangles (▲) show the border between the colon and ileal mucosa. Neither redness nor ulceration of the mucosa was observed.

Discussion

Difficult management problems remain in EIA. Complete resection of the entire involved colon and small intestine has resulted in prolonged watery stool and loss of electrolytes; hence, various definitive operations have been proposed for avoiding these problems. In 1968, Martin⁵ first introduced a modification of the Duhamel procedure preserving the aganglionic left colon expecting the facilitation of reabsorption. Subsequently, Kimura et al.¹² and Boley¹³ reported the use of the aganglionic right colon as onlay patch. Kimura described a two-stage procedure of Swenson-type pull-through using a devascularized combined segment created several months previously,¹² while Boley reported a single-stage endorectal pull-through procedure with antiperistaltic right colon patch.¹³ Kimura's devascularized combined segment from the mesocolon exhibited adequate vascular collaterals from the anastomosis.¹² Devascularization was initially undertaken 6 to 10 months after ileocolostomy, but subsequent clinical experiments and evaluations suggest that the interval of only a

couple of months is preferable.^{12,14-16} Nishijima et al.¹⁵ reported that the severed aganglionic colon patch maintained its viability and absorptive ability. Heath et al. reported that both the left and right aganglionic colon patches had the capacity to reabsorb water.¹⁷ However, the right colon patch, compared with left one, exhibited superior reabsorption of water and electrolyte, enabling a relatively short length of use.^{10,11} The ideal length for the colon patch is controversial. Martin reported that the entire aganglionic colon patch provided a greater surface area for absorption and fecal storage.¹⁸ However, there are some reports that extended aganglionic colon patch leads to frequent enterocolitis and other complications.⁶⁻⁹ Similarly, Nishijima et al.¹⁵ suggested that longer patch might develop enterocolitis or bowel obstruction, and accordingly advocated that 10-cm length was appropriate as a right colon patch. A two-stage operation has the advantage of providing a manageable ileocolostomy, which can be pulled down to the anus independent of its blood supply from the mesocolon, and, when severed, is available for the Soave procedure as well as the Swenson procedure.

Twenty years ago, we performed the definitive surgical treatment of the Soave pull-through with aganglionic sigmoid and ascending colon as a 16-cm onlay patch to the ganglionic ileum on a 1-year-old boy with EIA. Despite adequate small bowel length, the left colon onlay procedure could not provide expected reabsorption of water and resulted in recurrent episodes of enterocolitis with abdominal pain, melena, and diarrhea. We used the right colon onlay procedure on the present EIA patient with short bowel for its advantages regarding absorptive capacity and others. Even in this multi-stage reconstruction for EIA, pelvic adhesions were not too severe, and video assistance enabled us to complete the Swenson-type procedure without any difficulty.

Both left and right colon patches have certainly brought great benefits to EIA patients in terms of reabsorption of water and electrolytes, and improved prognosis, but the major prognostic factor is the length of residual unaffected small bowel. Children suffering from EIA with a long segment of involved small intestine often need long-term enteral or total parenteral nutrition (TPN). Fouquet et al.¹⁹ reported that children with EIA in which ileal involvement was less than 50 cm had good prognosis, while those with ileal involvement exceeding 50 cm needed long-term nutritional assistance. Ikawa et al.²⁰ also reported that EIA patients with less than 40 cm of ileal involvement could attain body weight within the normal range. Iron deficiency anemia is also one long-term complication in EIA with ileal involvement; this may not be related to the extent of ileal involvement, because almost all iron is absorbed in the duodenum and the upper jejunum.¹⁵

Innovative surgical techniques and TPN have improved the survival of children with EIA. However, even a surgical technique such as right colon patch often fails in bringing adequate normal bowel function, and TPN is generally necessary. Most patients with EIA are highly dependent on TPN for growth and therefore they will face not only disease-related complications but also those of TPN (including failure of vascular access and progressive liver failure).²¹ Although the postoperative course of the present patient is currently

satisfactory, he will require a careful and long-term support for bowel and nutrition.

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