

# Laparoscopic extensive colectomy with transanal Soave pull-through for intestinal neuronal dysplasia in 17 children

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**Background:** Open colectomy has been preferred for intestinal neuronal dysplasia type B (IND) due to its low morbidity rate and good functional results. The aim of this study was to investigate the feasibility and results of laparoscopic colectomy with transanal Soave pull-through for the treatment of IND in children.

**Methods:** Seventeen infants and children suffering from IND were treated by laparoscopic extensive colectomy with transanal Soave pull-through. The diagnosis of IND was made via anorectal manometry, X-ray contrast enema, suction biopsies, and laparoscopic full-thickness biopsies with hematoxylin-eosin staining. The technique used four or five abdominal ports. The sigmoid, transverse, and right colon up to the last ileal cove were mobilized laparoscopically in the extended form of IND. A modified Soave's anastomosis was performed. The patients' data, surgical procedures, operative data, postoperative complications and clinical outcomes were analyzed.

**Results:** Five patients underwent laparoscopic left colectomy with modified transanal Soave procedures, and the other 12 were treated by laparoscopic subtotal colectomy and required a Deloyers' maneuver for the Soave pull-through. The proximal margin of barium stagnation in patients with left colectomy was restricted to the distal end of the descending colon, sigmoid colon, and that in patients with subtotal colectomy was restricted to the proximal end of the descending colon, transverse colon, hepatic flexure,

and ascending colon. Postoperative complications included anastomotic leakage, severe perianal erosions, postoperative enterocolitis, and soiling. During a mean follow-up of 4 years, bowel frequency was 4-10 times per day in 3 months postoperatively in patients with subtotal colectomy. The clinical results were good, with no stool incontinence or constipation.

**Conclusions:** Laparoscopic procedure for left colectomy and subtotal colectomy with transanal Soave pull-through in infants and children with IND is safe, feasible, and effective. The location of barium stagnation in proximal margin may be used as a method to predict initially the proximal margin of the resected bowel segment.

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**Key words:** intestinal neuronal dysplasia; laparoscopy; subtotal colectomy

## Introduction

The advancement of immunohistochemical and molecular technologies has enabled a better understanding of the intestinal neuronal dysplasia type B (IND); however, the treatment of the disease is still controversial. It has been considered that if patients with IND do not recover from conservative treatment for several months or several years, the operation procedure is unavoidable. Sphincteromyectomy and classic radical correction for Hirschsprung's disease (HD) have shown a higher incidence of postoperative constipation, which may be caused by misjudgment of the length of affected bowel, leaving a dysganglionic segment in place.<sup>[1]</sup> The determination of proximal resection margin is most difficult in patients with IND. Recently, open extensive colectomy with Duhamel pull-through has been preferred for IND,<sup>[2]</sup> with a low morbidity rate and good functional results.<sup>[3]</sup> Since the introduction of laparoscopic-assisted transanal Soave pull-through in 1995,<sup>[4]</sup> experience in

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laparoscopic HD surgery has increased. Reports<sup>[5-7]</sup> have demonstrated that the laparoscopic procedures with transanal Soave pull-through for long-segment disease or total aganglionosis are feasible and safe. The current study was carried out to analyze the results of children who had undergone the laparoscopic Soave pull-through, and to find an adequate method to evaluate the proximal margin of a resected bowel segment based on histomorphology and functional evaluations.

## Methods

Between October 1999 and March 2007, 17 children underwent laparoscopically assisted colectomy with Soave pull-through. Medical treatment with laxatives and enemas was attempted as an initial approach. The pull-through was performed 6 months to 1 year later if the patient still showed persistent constipation and redo biopsy showed persistent dysganglionic pattern of bowel innervation. The patients with disseminated form such as total colonic involvement and total gastrointestinal involvement were excluded from this study if they underwent a previous colectomy. Of the 17 patients, 11 were male and 6 female, with a median age of 5 years and 10 months (range from 10 months to 13 years). All the patients had a history of constipation, abdominal distention and had been treated with laxatives and enemas. Only 2 patients failed to pass stool in the first 24 hours of life. All 17 patients had retractable chronic constipations and mild to moderate abdominal distention. Rectal examination did not demonstrate a tight anal sphincter with an empty rectum. The diagnosis was made by anorectal manometry, X-ray contrast enema, suction biopsies, and laparoscopic full-thickness biopsies in all patients.

Barium enema was given to all patients for a 24-hour delayed evacuation of barium from the colon. The proximal margin of barium stagnation was restricted to the distal descending colon and sigmoid colon in 5 patients, to the proximal descending colon in 6 patients, to the splenic flexure of the colon and transverse colon in 4 patients, and to the distal ascending colon in 2 patients. Fifteen patients had a recto-anal inhibition reflex and a reflex wave with a shape of W or U. The diagnosis was made in all patients by mucosal suction biopsy, and histochemical assay was performed using hematoxylin-eosin (HE) staining for evaluation of the extent of the disease. IND was defined as described in the literature.<sup>[8]</sup> In IND involving the intestinal segment, an increase of ganglionic (nerve) cells was observed in the ganglion with more than eight nerve cells per ganglion; nerve cells were grouped into spherical giant ganglia. Hyperplasia of the submucous plexus and increased AChE-positive

nerve fibers in the lamina propria mucosae and in the adventitia of submucosal arteries were considered the mandatory criteria for diagnosis of IND.<sup>[3,8]</sup>

All operations were performed by the same surgical team. Laparoscopic left colectomy was performed using three or four trocars. Laparoscopic subtotal colectomy was performed using four or five trocars. The abdomen was insufflated with CO<sub>2</sub> at a pressure of 8-12 mmHg. It was essential to establish the proximal margin of the resected bowel segment at the beginning of the intraabdominal procedure. Multiple full-thickness biopsies were obtained from the distal end to the proximal end and analyzed with HE staining to confirm the extent of disease. Perforation or bleeding at the biopsy site could be closed with a figure-of-eight braided suture. A marking stitch was placed at the biopsy site, where the ganglionic bowel was confirmed by the pathologist, and used as a landmark during the transanal portion of the procedure.

Once the proximal margin of the resected bowel was firmly established, the mesocolon was dissected. The mesocolon should be divided adjacent to the colon in the dysganglionic portion of the bowel. If the dysganglionic segment was longer than the mid-sigmoid colon, the colon to be pulled through was mobilized from the lateral wall by dissecting the peritoneal attachment, thereby medializing the left colon. The splenic flexure may also be taken down using harmonic scalpel in a similar fashion to avoid tension on the anastomosis. If the diseased bowel was longer, the gastrocolic ligament, hepatic flexure, ascending colon, and ileocecal junction were mobilized. The middle colic artery was interrupted while the descending branch of the arteriae colica dextra was preserved. The remaining colon necessitated a Deloyers' maneuver rotated through 270 degrees and brought down, and the appendix was excised. The proposed segment to be pulled through was then brought into the pelvis to gauge whether a tension-free anastomosis was made.

The pneumoperitoneum was then evacuated, and attention was turned to the transanal portion of the procedure. When the submucosal plane and muscular layer were identified, a fine-tipped hemostat and needle-tip cautery made the endorectal mucosal dissection easier and preciser. The endorectal dissection was continued proximally in a circumferential manner until the muscular cuff of rectal wall was intussuscepted freely. The posterior wall of rectal cuff was then divided 1 to 2 cm below the endorectal mucosal dissection. Once a free plane was found, the division of the muscular rectal wall was continued circumferentially up the intraabdominal colon from the muscle sleeve. The muscular cuff was inverted and the posterior wall of which was split by a V incision,

and the cuff was shortened leaving an anal stump of 3 to 4 cm. The dysganglionic colon was pulled down through the muscle sleeve and out trans-anally until the previously marked ganglionic portion of the colon was identified. At this point, a primary anastomosis between the neo-rectum and the anus was performed with interrupted fine absorbable sutures.

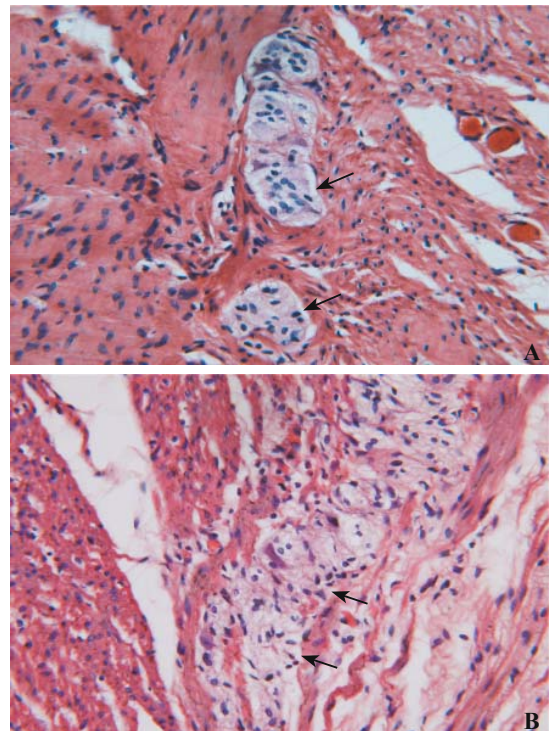
The data about age, gender, symptoms, diagnostic evaluation, surgical procedures, postoperative complications, and clinical outcome were analyzed.

## Results

In the 17 patients, the proximal margin of resected colon was identified by rapid frozen section analysis with HE staining (Fig. 1). In this series, 5 patients had left colectomy, whose barium stagnation over 24 hours was located in the distal segment of the descending colon (3 patients) and sigmoid colon (2 patients) (Fig. 2); the average operative time was 145 minutes (range, 115-190 minutes). The remaining 12 patients had subtotal colectomy, whose barium stagnation over 24 hours was located in the proximal segment to the descending colon (6 patients), transverse colon (4 patients) and ascending colon (2 patients) (Fig. 3); the length of the colon preserved was 11.5 cm (range, 7-13 cm), and the average operative time was 188 minutes (range, 145-240 minutes). The average volume of bleeding was about 40 ml (range, 20-80 ml). In two patients who had been subjected to previous operative management, one patient received sphincteromyectomy at the age of 12 months with recurrence of constipation 6 months after operation, and the other patient received perineoanoplasty because of low imperforate anus. Histologically, the proximal margin of the resected bowel segment showed normal innervation in 16 patients and pathological innervation in 1 patient.

Postoperative complications are summarized in Table. One patient with subtotal colectomy developed leakage on the fifth postoperative day and required temporary bowel diversion. The mean postoperative follow-up of the 17 children was 4 years (range, 1-7 years). Chronic bleeding, fecaloma formation, or anastomotic stricture was not found in any patient. Severe perianal erosion caused by frequent bowel movement occurred in 10 patients. One patient had postoperative enterocolitis (5.8%), which was treated conservatively. Soiling was seen in 2 patients (5.8%) who had undergone subtotal colectomy, with a steady improvement of continence in the second year.

Defecation frequency was 1-3 times per day in 3 months postoperatively in patients who received left colectomy, and 4-10 times per day in 3 months



**Fig. 1.** A: The normal myenteric plexus found in the normal segment of colon (arrow); B: A hyperplastic change in the myenteric plexus found in the resected colon (arrow) (HE, original magnification  $\times 200$ ).



**Fig. 2.** A 1-year-old boy with HD associated IND, barium stagnation located in the distal segment to descending colon (arrow). He underwent a laparoscopic left colectomy with modified Soave procedures.



**Fig. 3.** A 2-year-old girl with IND had the location of barium stagnation in the ascending colon (arrow). She was treated by laparoscopic subtotal colectomy and required a Deloyers' maneuver for the pull-through.

**Table.** Complications and bowel movements

Complications	Subtotal colectomy (12 patients)	Left colectomy (5 patients)
<b>Complications</b>		
Anastomotic stricture	0	0
Anastomotic leakage	2	0
Severe perianal erosions	9	1
Postoperative enterocolitis	1	0
Soiling	2	0
Fecal incontinence	0	0
Constipation	0	0
<b>Defecation frequency (postoperatively)</b>		
<2 wk	5-30 times per day	3-8 times per day
3 mon	4-10 times per day	1-3 times per day
6-12 mon	2-5 times per day	1-3 times per day
12-24 mon	2-3 times per day	1-2 times per day

postoperatively in patients with subtotal colectomy (Table). The overall functional outcome was good in all the patients with no stool incontinence or constipation during the follow-up period.

## Discussion

Clinical experience<sup>[9,10]</sup> has indicated that IND is one of the most common causes of chronic retractable constipation. Although there is controversy over the surgical treatment in patients with IND,<sup>[11]</sup> aggressive surgical management should be adopted because IND has an unpredictable clinical course, and that its management should be dependent on the conditions of each patient.

Transanal Soave pull-through is the best procedure for short-segment and rectosigmoid HD. When a long-segment or total aganglionosis is found, laparoscopically assisted approach should be employed. The isolated colonic segment of IND can be resected as that of HD. If the IND section of the colon reaches the splenic flexure and can thus not be pulled down to the perineum, the colon is resected up to the hepatal flexure, ascending part and ileocecal junction. In our study, 12 patients requiring subtotal colon resection received the laparoscopic procedure without additional skin incision. The arterial branches of the ascending colon have to be preserved and laparoscopic Deloyers' maneuver is needed. The colon needs to be rotated by 270 degrees (not 180 degrees) as an open procedure,<sup>[2]</sup> which is an important concept because it is very difficult to rotate the colon to avoid the pull-through with any twisting laparoscopically. It should be tried again and again and completed by the cooperation of abdominal maneuver and perineal pull-through.

Anastomotic leakage occurred in 2 patients with

subtotal colectomy in our study, but it is difficult to affirm its relationship with ischemia, high tension, or poor nervous alimentation in anastomotic stoma because of a small series of patients. Mattioli et al<sup>[3]</sup> reported a high adhesion rate in IND patients undergoing an open pull-through procedure. They speculated that the major causes are hypomotility of the entire bowel in IND patients. Because there was no intestinal adhesion in our series, laparoscopic procedures showed marked superiority in patients with IND. Adhesion and bowel obstruction are high risk factors for postoperative enterocolitis.<sup>[12]</sup> The incidence of postoperative enterocolitis is as high as 12% to 33%.<sup>[13]</sup> The relatively low incidence of postoperative enterocolitis (5.8%) in the current series may be related to the lower adhesion rate.

Soiling and increased frequency of bowel movements in patients may be due to over stretching of the downwardly pulled colon. The 5.8% soiling rate in this study was within the 4%-17% range reported in literature.<sup>[3,9,12]</sup> Patients with subtotal colectomy have frequent bowel movements after surgery, and 40 or more bowel movements a day at most, but it can recover to 2-3 times per day after about 6 months to 1 year for most cases due to preservation of the ileocecal valve and the cecum. Long-term amelioration of this condition has been reported.<sup>[14]</sup> There is a high incidence of perianal erosions in patients who had subtotal colectomy. Meticulous nursing care and keeping the perianal area dry with blower can decrease the pain and speed up the recovery.

It is more difficult to diagnose IND than HD before operation because of their non-typical clinical characteristics. Histochemical techniques in mucosal biopsies and full-thick specimens for accurate diagnosis of IND have been improved markedly.<sup>[8,15-17]</sup> Biopsies at proximal level may be performed via laparoscopy or a small umbilical incision before bowel dissection. This is especially true in IND, and the extent of dysganglionic bowel is most crucial to preoperative planning, particularly in a single-stage repair. In patients with HD, the anticipated level of aganglionosis can be determined by the radiographic transition zone on preoperative contrast enema,<sup>[18]</sup> but contrast enema studies in patients with IND have shown no typical manifestations (without obvious caliber change) and the transition zone is a poor predictor of dysganglionic segment.<sup>[2]</sup> Interestingly, there was a delayed evacuation of barium for 24 hours in all patients with IND in our studies. The 5 patients with left colectomy had barium stagnation located in the distal segment to the descending colon, and the 12 patients with subtotal colectomy had barium stagnation located in the proximal segment to the descending colon, hepatic flexure, transverse colon, and ascending colon. The histological finding of the proximal margin of the

resected bowel segment correlated accurately with the level of the ganglionic segment in 94% (16/17) of the patients, a good explanation for patients' bowel segment resected more proximal to the operatively confirmed level of dysganglionosis, especially in patients with subtotal colectomy. In a patient with postoperatively retained IND segment, barium stagnation was located in the distal segment to the ascending colon, the length of the colon preserved was only 8 cm, and persistent constipation did not appear during the follow-up period. Probably, the short colonic segments with pathological innervation have sufficient contractility to pass stools through the remaining rectum after pull-through.<sup>[3]</sup> Thus, our data indicate that the location of delayed evacuation of barium can predict initially the level of dissection of the colon, particularly for colectomy by one-stage laparoscopically assisted pull-through. Further studies are needed to evaluate data on morphologic and functional maturation in large series of IND cases.

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**Contributors:** Tang ST wrote the first draft of this paper. All authors contributed to the intellectual content and approved the final version.

## References

- Tomita R, Munakata K, Howard ER, Fujisaki S. Histological studies on Hirschsprung's disease and its allied disorders in childhood. *Hepatogastroenterology* 2004;51:1042-1044.
- Skába R, Frantlová M, Horák J. Intestinal neuronal dysplasia. *Eur J Gastroenterol Hepatol* 2006;18:699-701.
- Mattioli G, Castagnetti M, Martucciello G, Jasonni V. Results of a mechanical Duhamel pull-through for the treatment of Hirschsprung's disease and intestinal neuronal dysplasia. *J Pediatr Surg* 2004;39:1349-1355.
- Georgeson KE, Fuenfer MM, Hardin WD. Primary laparoscopic pull-through for Hirschsprung's disease in infants and children. *J Pediatr Surg* 1995;30:1017-1022.
- Bonnard A, de Lagausie P, Leclair MD, Marwan K, Languépin J, Bruneau B, et al. Definitive treatment of extended Hirschsprung's disease or total colonic form. *Surg Endosc* 2001;15:1301-1304.
- Travassos DV, Bax NM, Van der Zee DC. Duhamel procedure: a comparative retrospective study between an open and a laparoscopic technique. *Surg Endosc* 2007;21:2163-2165.
- Rückauer KD, von Dobschuetz E. Laparoscopically assisted colorectal resection in Hirschsprung's disease. *Eur J Med Res* 2005;10:361-365.
- Meier-Ruge WA, Ammann K, Bruder E, Holschneider AM, Schärli AF, Schmittenbecher PP, et al. Updated results on intestinal neuronal dysplasia (IND B). *Eur J Pediatr Surg* 2004;14:384-391.
- Schmittenbecher PP, Sacher P, Cholewa D, Haberlik A, Menardi G, Moczulski J, et al. Hirschsprung's disease and intestinal neural dysplasia—a frequent associated with implication for the postoperative course. *Pediatr Surg Int* 1999;15:553-558.
- Zhou XF, Li GS, Pan CL, Wang LT, Liu YC, Xie JL, et al. Diagnosis of 63 cases with Hirschsprung's disease-allied disorder. *Chin J Pediatr Surg* 2004;25:571-572. [In Chinese]
- Martucciello G, Caffarena PE, Lerone M, Mattioli G, Barabino A, Bisio G, et al. Neuronal intestinal dysplasia: Clinical experience in Italian patients. *Eur J Pediatr Surg* 1994;4:287-292.
- Tang ST, Zhou X, Ruan QL, Guo XL, Liu CP, Liu CP, et al. Risk factors for enterocolitis after pull-through for Hirschsprung's disease. *Chin J Pediatr Surg* 2001;4:213-215. [In Chinese]
- Marty TL, Seo T, Matlak ME, Sullivan JJ, Black RE, Johnson DG. Gastrointestinal function after surgical correction of Hirschsprung's disease: long-term follow-up in 135 patients. *J Pediatr Surg* 1995;30:655-658.
- Tsuji H, Spitz L, Kiely EM, Drake DP, Pierro A. Management and long-term follow-up of infants with total colonic aganglionosis. *J Pediatr Surg* 1999;34:158-162.
- Kubota A, Yamauchi K, Yonekura T, Kosumi T, Oyanagi H, Mushiake S, et al. Clinicopathologic relationship of hypoganglionosis. *J Pediatr Surg* 2001;36:898-900.
- Park SH, Min H, Chi JG, Park KW, Yang HR, Seo JK. Immunohistochemical studies of pediatric intestinal pseudo-obstruction: bcl2, a valuable biomarker to detect immature enteric ganglion cells. *Am J Surg Pathol* 2005;29:1017-1024.
- Zhou J, Wei MF, Feng JX, Wang G, Ru LQ, Zhang Y, et al. A rapid method for the intraoperative diagnosis of Hirschsprung's disease and Hirschsprung's disease-allied disorder. *Chin J Gastrointestinal Surg* 2006;6:456-457. [In Chinese]
- Proctor ML, Traubici J, Langer JC, Gibbs DL, Ein SH, Daneman A, et al. Correlation between radiographic transition zone and level of aganglionosis in Hirschsprung's disease: implications for surgical approach. *J Pediatr Surg* 2003;38:775-778.

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