One-stage laparoscopy-assisted endorectal pull-through for late presented Hirschsprung's disease—Case series

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A B S T R A C T

INTRODUCTION: Children with late-presenting Hirschsprung's disease (HD) are classically treated by a staged operation with enterostomy. An alternative may be one-stage laparoscopy-assisted endorectal pull-through, which has cosmetic advantages. This case-series report describes the outcomes of children with late-presenting HD who underwent this procedure.

PRESENTATION OF CASES: Eight older (>3 years) children (five males, three females) underwent one-stage laparoscopy-assisted endorectal pull-through in 2010–2012. A retrospective review revealed their median age was 9.9 (range, 3.4–14) years. The transitional zone was rectosigmoid junction in 4 patients, and was rectum in 4 patients. For bowel preparation, five patients required rectal irrigation under general anesthesia. The median operating time was 263 min. There were no intraoperative or early post-operative complications. Patients started a diet a median of 5 days after the operation and were discharged a median of 11.5 days. During the median follow-up period of 37 months, seven (87.5%) had acquired voluntary bowel movements and 12.5% had grade 1 soiling. However, five (62.5%) of the patients still had constipation. The constipation was manageable with diet or laxatives in four patients but one patient continued to require regular enemas.

DISCUSSION: One-stage laparoscopy-assisted endorectal pull-through in late-presenting HD was feasible, even in patients with large fecaloma with obstruction. Rectal irrigation under general anesthesia and the use of laparoscopy and a bipolar coagulator help to overcome the technical difficulties of this procedure.

CONCLUSION: One-stage laparoscopy-assisted endorectal pull-through in children with late-presenting short segment HD is feasible and safe.

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1. Introduction

Hirschsprung’s disease (HD) is a congenital malformation that is mainly found in early infancy. The incidence is approximately 1 in 5000 live births [1]. In developed countries, about 90% of all patients with HD are diagnosed within 1 year after birth [2,3]. The remaining cases are usually diagnosed in childhood but can even be diagnosed in adulthood. The incidence of late-presenting HD cases is higher in developing countries [4,5]. Late-presenting cases usually have complications including severe bowel distension with hypertrophy. The usual therapeutic approach is a staged operation with an enterostomy. However, several studies have shown that one-stage transanal endorectal pull-through (TAERTP) has similar long-term functional outcomes as the staged operation [6–8]. In our hospital, we have been doing one-stage laparoscopy-assisted endorectal pull-through for late-presenting HD to improve the cosmetic outcome. The short-term outcomes of this approach are described in this report.

2. Presentation of case series

2.1. Subjects

All consecutive children who were older than 3 years at the time HD was diagnosed and underwent one-stage laparoscopy-assisted endorectal pull-through at the Asan Medical Center and Haemundae Paik Hospital in 2010–2012 were identified by retrospective review of the medical records. The following data were extracted: age, gender, body weight, operative findings, operation time, and post-operative bowel habits. The patients were followed up at outpatient clinics and the parents were interviewed regarding the functional outcome of surgery, namely, frequency of bowel movements, soiling, and constipation. The functional outcomes were categorized according to the Krickenberg classification system [9].

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Voluntary bowel movements were defined as the urge to defecate, the capacity to verbalize this feeling, and the ability to hold the bowel movement. Soiling was categorized according to a three-grade system: grade 1 was defined as occasional soiling (1–2 times per week); grade 2 was defined as daily soiling but associated social problems were absent; and grade 3 was defined as constant soiling with social problems. Constipation was also categorized according to a three-grade system: grades 1 and 2 were defined as constipation that could be managed by changing the diet or laxative treatment, respectively. Grade 3 constipation was resistant to laxatives and diet.

The study protocol was approved by our Institutional Review Board (AMC IRB 2013-0977). Informed consent to participate in this case series report was obtained from the parents of the patients. The report is consistent with CARE criteria for reporting case reports [10].

2.2. Pre-operative bowel preparation to overcome severe fecal impaction

Three methods of pre-operative fecal disimpaction were used, namely, fasting and conventional enema, oral polyethylene glycol, and rectal irrigation under general anesthesia. Fasting and repeated enema was used first. If there was no improvement, the patients who lacked an intestinal obstruction ate oral polyethylene glycol. If the patient had obstructive symptoms, rectal irrigation under general anesthesia was performed. The enema usually included extraction of fecaloma and massive irrigation of the colon. The main operation was delayed at least 3 days to allow the intestine to recover from edema.

2.3. Surgical technique

2.3.1. Laparoscopic approach: determining the level of the bowel resection

After general anesthesia was provided, the patient was placed into the lithotomy position. A full-thickness rectal biopsy was obtained to confirm the diagnosis. After CO₂ insufflation of the peritoneal cavity, three 5-mm trocars (one for the camera and two for the working ports) were inserted. After identifying the transition zone, the level of pull-through was tagged after confirmation of the presence of ganglion cells by frozen biopsy. It is helpful to mobilize the splenic flexure and divide the superior rectal vessels and mesocolon by using an ultrasonic scalpel. Deep pelvic dissection was extended circumferentially just outside the rectal wall, distal to peritoneal reflection.

2.3.2. Perineal approach: overcoming massive bleeding

A circumferential incision was made in the mucosal layer 1 cm above the dentate line. Submucosal dissection above the dentate line was performed by using a bipolar coagulator to achieve effective hemostasis. After the submucosal dissection had been completed, the rectal muscle was incised circumferentially. The rectal muscular cuff was split longitudinally both anteriorly and posteriorly. The rectum and sigmoid colon were then pulled down through the rectal sleeve until the tagged bowel emerged. The colon was transected above this point and anastomosed to the anal mucosa by using absorbable sutures.

2.4. Demographics, diagnosis, and preparation of the bowel

There were eight patients with late-presenting HD who underwent one-stage laparoscopy-assisted endorectal pull-through during the study period. Five were male and three were females. The median age was 9.9 (range, 3.4–14) years, the median weight was 27.6 (range, 14–50) kg, and the median height was 134.3 (range, 91–159.2) cm. All patients presented with prolonged constipation and abdominal distension. The constipation usually started at birth and worsened when the patient started on a solid diet. The constipation became more serious as the children aged. All children had severe fecal impaction, as shown by simple abdominal X-rays.

To diagnose the HD, barium enema, rectal manometry, and rectal biopsy were performed. All children underwent barium enema except one: the exception was due to severe fecal impaction. Four children underwent rectal manometry; in three cases, the rectal inhibitory reflex was absent. All children underwent rectal biopsy. It was either suction biopsy (n = 3) or a full thickness rectal biopsy (n = 5). Only one child achieved satisfactory bowel decomposition by conventional enema and fasting. Two children achieved satisfactory fecal disimpaction after oral polyethylene glycol. The five patients required rectal irrigation under general anesthesia. The pre-operative bowel preparation took 6–11 days.

2.5. Outcome

All patients underwent laparoscopy-assisted endorectal pull-through. The transitional zone was rectosigmoid junction in 4 patients, and was rectum in 4 patients. A median of 35 (range, 23–44) cm of colon was resected. The median maximal diame-

| Patient | 1 | 2 | 3 | 4 | 5 | 6 | 7 | 8 |
|---------|---|---|---|---|---|---|---|---|
| Age at diagnosis | 6 y | 3 y 5 m | 14 y | 12 y | 5 y 5 m | 13 y | 11 y | 6 y 10 m |
| Gender | M | M | M | F | F | F | M | F |
| Weigh (kg) | 19.7 | 14.0 | 50.0 | 29.1 | 18.0 | 40.0 | 32.0 | 26 |
| Height (cm) | 115.8 | 91.0 | 159.2 | 148.4 | 110.0 | 150.0 | 137.5 | 131 |
| Diagnosis | Colon study | Y | N | Y | Y | Y | Y | Y | Y |
| Manometry | Y | Y | N | N | Y | Y | N | N |
| Rectal Biopsy | Y | Y | Y | Y | Y | Y | Y | Y |
| Pre op fasting, days | 7 | 6 | 6 | 11 | 6 | 6 | 6 | 4 |
| Enema under GA | Y | Y | Y | Y | N | N | N | Y |
| Transition zone | R | RSJ | R | RSJ | R | RSJ | R | RSJ |
| OP time (min) | 270 | 190 | 484 | 238 | 283 | 230 | 295 | 255 |
| Intra-op transfusion | N | N | N | N | N | N | N | N |
| Time to diet (days) | 6 | 4 | 6 | 4 | 4 | 5 | 8 | 5 |
| Time to discharge (days) | 11 | 9 | 24 | 12 | 5 | 10 | 14 | 20 |
| Follow up Period (m) | 53 | 51 | 56 | 37 | 26 | 26 | 23 | 17 |

Abbreviations: R, rectum; RSJ, rectosigmoid junction; GA, general anesthesia; M, male; F, female; D, day; y, years; m, months; Y, yes; N, no.
Table 2
Functional outcome according to Knickerbank classification.

| Voluntary bowel movements | Soiling | Constipation |
|---------------------------|---------|--------------|
| No (%)                    | 1 (12.5%) | 7 (87.5%) | 3 (37.5%) |
| Yes (%)                   | 7 (87.5%) | 5 (62.5%) |             |
| Grade 1                   | 1 (12.5%) | 2 (25%)   |             |
| Grade 2                   | 1 (25%)   |           |             |
| Grade 3                   | 1 (12.5%) |           |             |

ter of the resected bowel was 6 (range, 3–13.5) cm. The median diameter of the proximal bowel that was anastomosed to the anus was 5 (range, 2.5–6) cm. It is usually acceptable when the circumference of the bowel is twice that of the anus. The median operating time was 263 (range, 190–484) min. None of the patients required a transfusion during the operation. The patients started a diet a median of 5 (range, 4–8) days after the operation and were discharged a median of 11.5 (range, 7–24) days post-operatively. There were no major intraoperative and early post-operative complications, including anastomotic leakage, wound infection, and enterocolitis (Table 1).

The median follow-up period was 37 (range, 17–56) months. In the first month, the frequency of defecation was up to 20 times per day. Within 3 months, the frequency of bowel movement recovered to below five times per day. At last follow up visit, all but one patient (87.5%) could feel and verbalize their bowel movements. Only one patient had grade 1 soiling. Five patients (62.5%) still had constipation. For two patients, the constipation was manageable by dietary modification. Another two patients treated with laxatives. However, one patient required regular enemas to manage the constipation (Table 2).

3. Discussion

While most cases of HD manifest during the neonatal period, some of HD is diagnosed during childhood or adulthood [11,12]. Such late-presenting HD cases are usually present with severe constipation, abdominal distension, vomiting, and recurrent enterocolitis. Many have been treated for constipation with enemas and laxative from early infancy [11,11]. However, most physicians fail to differential diagnosis between functional constipation and HD.

HD is diagnosed by barium enema, anorectal manometry, and rectal biopsy. Barium enema is not helpful for very short segment HD and total colonic aganglionosis, because the transitional zone tends to be very short in late-presenting HD cases [11,13,14]. The success with which rectal manometry diagnoses HD also varies, ranging from 75% to 100% of cases [14–16]. In our study, rectal manometry showed 25% of false negative. Therefore, suspected HD in older children should be confirmed by pathology [11,13,14]. We usually employ the suction biopsy technique but it can be difficult to get an adequate specimen that includes submucosal tissue in children who have a thickened bowel wall. In such cases, a full thickness rectal biopsy under general anesthesia is helpful [17]. In our opinion, combining rectal irrigation with a rectal full thickness biopsy is an good option as it allows the simultaneous diagnosis of HD and preparation of the bowel.

For many years, one-stage operations were avoided in older patients with HD because of difficulties in decompressing the bowel [7,18,19]. Chronic constipation with a huge load of feces often induces colonic obstruction with fecaloma and gradual dilatation with severe wall thickening. The presence of impacted feces is also an enormous obstacle when performing one-stage endorectal pull-through, which is why many surgeons recommend colostomy as the first stage [4,5,20,21]. In a recent study of 41 children who presented with HD after 1 year of age, 35 (85.4%) underwent colostomy to achieve decompression. Of these, 68.6% patients experienced colostomy-related complications that included skin exorotation (n = 11), stoma prolapse (n = 11), and stoma stenosis (n = 2). 26% of patients suffered from complications after colostomy closure [5]. In our series, half of the patients were adolescents. Enterostomy can devastate the daily life of an adolescent; it can also distort the patient’s self-image and induce psychological damage. Indeed, these considerations caused us to favor a one-stage operation for older children with HD. To overcome fecal impaction, we tried various methods to clean the colon before the operation, including massive colon irrigation under general anesthesia. The results of these methods were encouraging.

Late-presenting HD also associates with thickened mesentery, inflamed mucosa, longstanding dilated colon, and rigid pelvis, all of which impose further technical difficulties. In newborns and younger infants, the fixation of the colon to the retroperitoneum is looser, which makes it easier to resect long segments of colon through the anus [7]. By contrast, in children older than 3 years, the transanal procedure of submucosal dissection is very difficult because of the mucosal inflammation and fibrotic changes resulting from recurrent enterocolitis. Moreover, ligation of the thick mesenteric vessels through the anal canal often results in tearing of the stretched vessels and bleeding. To prevent bleeding complications, we used laparoscopy and bipolar coagulation. The bipolar coagulator effectively induces hemostasis with minimal tissue injury. Moreover, laparoscopic ligation of the mesenteric vessels with an ultrasonic scalpel prior to submucosal dissection can reduce the possibility of massive bleeding. Laparoscopic assistance provides crucial clues for determining the correct level of bowel resection needed to ensure tension-free anastomosis [22]. Also, laparoscopy is essential for protecting the sphincter. Dilated bowel and thickened mesentery can cause over-dilation of the sphincter, which results in a weakened fecal continence mechanism. Laparoscopic-assisted pelvic dissection may prevent damaging over-dilated internal anal sphincters.

In general, patients with late-presenting HD take longer to recover normal bowel habits than neonates because older children have a stretched pelvic floor and a decreased sense of defecation. In our series, all eight patients had frequent stooling during the immediate post-operative period. Later, this dropped to 1–2 defecations per day. To manage constipation after surgery, we used laxatives and regular enemas and diet changes. At the end of the median follow-up period of 37 (range, 23–56) months, one patient still had to undergo regular enemas because of constipation and one patient had intermittent soiling.

Although, surgical treatment of late-presenting HD will be more difficult than in neonates, we found that one-stage laparoscopy-assisted endorectal pull-through was feasible in patients with late-presenting HD, even in cases with a large fecaloma with obstruction. The technical difficulties of this approach are ameliorated by preparing the bowel by rectal irrigation under general anesthesia and by using laparoscopy and a bipolar coagulator to perform the surgery. However, the functional outcome of our patients was still not as satisfactory as that of neonates. Nevertheless, it also takes a long time to restore bowel function after staged operations for late-presenting HD. To promote gradual improvement over time, patients with late-presenting HD require careful follow up with dietary advice.

4. Conclusions

One-stage laparoscopy-assisted endorectal pull-through in children with late-presenting HD is feasible and safe avoiding enterostomy complications.
Conflict of interest

There are no relationships/conditions/circumstances that present a potential conflict of interest.

Funding

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Ethical approval and consent

The study protocol was approved by our Institutional Review Board (AMC IRB, 2013-0977). All patients provided written informed consent for publication of this case series report and the accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contribution

So-Hyun Nam wrote the paper with data analysis.
Min Jeong Cho reviewed the medical record and collected the data.
Dae Yeon Km is a director of this paper.

Guarantor

So Hyun Nam, M.D., Ph.D. and Dae Yeon Kim, M.D., Ph.D.

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