Laparoscopic-assisted endorectal pull-through for Hirschsprung's disease. *A retrospective study*

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ABSTRACT

Objectives: To summarize the efficacy of the laparoscopic-assisted transanal-endorectal pull-through procedure for Hirschsprung's disease (HD).

Methods: Between May 2006 and May 2013, 22 children with HD undergoing laparoscopic-assisted endorectal pull-through procedures were retrospectively analyzed. The operative time, pathology, intraoperative blood loss, recovery time for gastrointestinal function, postoperative hospital stay, complications, and defecation functions of the patients were analyzed.

Results: The procedure was successfully completed in all 22 patients without conversion to open surgery. The operative time was 105-190 minutes (mean, 160.4 minutes), and the intraoperative blood loss was 20-50 ml. The mean time for the recovery of gastrointestinal function was 22 hours, and the mean postoperative hospital stay was 8 days. All patients had 1-3 defecations per day at 6 months postoperatively without constipation, soiling, or stoma stenosis.

Conclusion: The laparoscopic-assisted transanal-endorectal pull-through procedure is a safe and feasible technique for patients with HD.

The mainstream treatment for Hirschsprung's disease (HD) is surgery. The basic principle of definitive surgery is the removal of poorly functioning aganglionic bowel and the creation of an anastomosis of a normally innervated portion of the colon to the anus. It is known that a good outcome cannot be guaranteed despite complete resection of the aganglionic bowel segment. Postoperative bowel dysfunction presents as enterocolitis in 6-30% of patients and as constipation and soiling in 11-35% of patients.[1,2] One possible reason for this dysfunction is that the proximal bowel remains "abnormal", or it could be due to inaccurate preoperative screening or inappropriate surgical technique.[3] In this study, we summarized our experience with the laparoscopic-assisted endorectal pull-through procedure (LAEPT).

Methods. This is a retrospective study, which was approved by the review board of Xiangyang Central Hospital, Xiangyang, China. Patients with HD who underwent laparoscopic endorectal pull-through procedure at the General Surgical Department of Xiangyang Central Hospital, Xiangyang, China, between 2006 and 2013 were identified in this study. Patients with the long segment, total colonic aganglionosis, intestinal neuronal dysplasia (IND), severe enterocolitis, poor general health, or previous pelvic surgery were excluded.

Hirschsprung's disease was diagnosed based on clinical symptoms, barium enema, entire gastrointestinal barium meal combined with multi-temporal abdominal films, rectal suction biopsy, and anorectal manometry.[4] All children presented with abdominal distension or progressively worsening intractable constipation. All patients had a well-defined transitional zone in mid sigmoid colon (common segment type for 20 cases, short segment type for 2 cases). The diagnosis was confirmed using intraoperative frozen biopsy. All patients underwent laparoscopic surgery, and the length of resected bowel was determined intraoperatively.

All patients underwent a strict preoperative protocol. A warm saline enema was performed once a day for 1-2 weeks. Normal diet was permitted until 6 hours before surgery. Preventive antibiotic treatment was performed 3 days before the operation.

Operative procedure. The veress needle was inserted through the umbilicus to establish the pneumoperitoneum. A 5-mm 30° laparoscope was used through the umbilicus. Two 5-mm ports were placed in the right upper and lower quadrants. In some cases, a port in the left lower quadrant was needed. These ports were 5 mm in diameter. After placement of the trocars, the transition zone and the dilated colon could be identified by morphologic changes. The hypertrophic dilated bowel was stiff in shape and slightly paler in color, while the dilated colon without hypertrophy was

normal in color, and its haustra coli was obvious. When the dilated colon without hypertrophy was clamped by an atraumatic grasper, it had elasticity and normal thickness.

The seromuscular biopsy specimens of the transitional zone, thickened dilated colon, and non-thickened dilated colon were taken under laparoscopy, and the range of the aganglionosis was assessed. Originally, the peritoneal reflection was incised circularly with the ultrasonic scalpel, and mobilization started at the rectosigmoid. This process was performed until the colon pedicle was tensionless to the pelvis. The rectal anterior wall should not be dissected, while the posterior plane needed to be dissected 2-3 cm into the pelvis. The anterior and posterior planes were then joined, hugging the rectal wall. Once the pathologist had confirmed the presence of ganglion cells of the dilated colon without hypertrophy, dissection of the colon pedicle began. The mesenteric dissection was extended proximally, and the marginal artery was preserved carefully. The splenic flexure needed no mobilization. Once the endoscopic dissection of the colon and rectum had been completed, the trans anal dissection started. Six to 8 traction sutures or the Lone Star Retractor (Lone Star Medical Products, Inc, USA) was used to retract the anus and expose the rectum (**Figure 1**).

In the rectal mucosa, a circular incision was performed with a 0.5-1 cm distance to the dentate line. The exposed mucosal edge was pulled with fine silks in the proximal lip, then an enclosing submucosal plane was dissected with a fine-tipped hemostat. Electrocautery was only used for coagulation by touching the hemostat in order to prevent injury to the mucosa and internal sphincter. The plane was extended proximally until the muscular cuff was inverted and pulled out of the anus. It was then shortened, leaving a muscular cuff of 2-3 cm; with the posterior wall V-shaped partial resection, the pointed end of the V should reach the level of the anastomosis. The rectum and colon were pulled down through the anus in continuity until the dilated bowel presented in the anus. Then the laparoscope was used, and the bowel was inspected to avoid internal herniation or twisting. The aganglionic and thickened dilated segments were resected. Coloanal anastomosis between the dilated colon and anus was fashioned with interrupted fine absorbable sutures. Since the diameter of the rectum was relatively larger than that of an anus, the 4 quadrants were sutured in advance to avoid an uneven anastomosis.

The specimen was examined histologically post operation. All patients maintained gastric decompression for 12 hours post operation. A diet was provided after
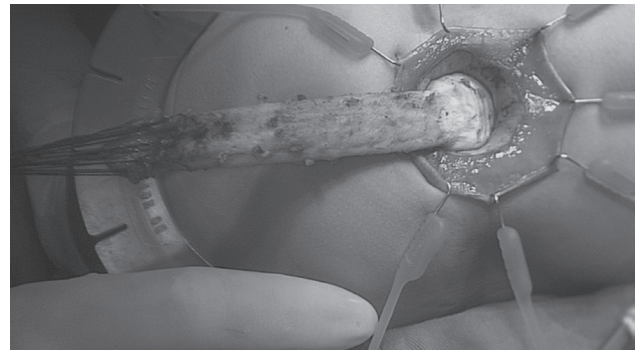

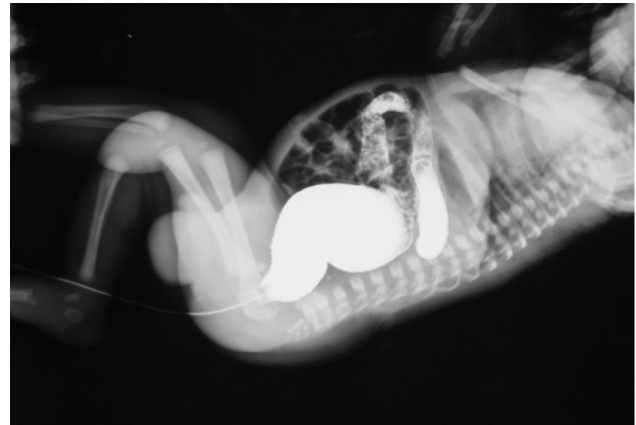**Figure 1 -** Using the lone star retractor to retract the anus and expose the rectum.

Figure 2 - Digital examination of the anus and anal dilatation were performed 2 weeks after surgery.

the bowel function recovered. Digital examination of the anus and anal dilatation were performed 2 weeks after surgery (**Figure 2**). If there was no anastomotic stenosis, anorectal dilatation was performed once or twice per week for 1-3 months. If there was anastomotic stenosis, anorectal dilatation was performed once or twice a day until the stenosis disappeared. Barium enema was performed 3-6 months postoperatively.

Data collection. The data of clinical characteristics, surgical procedures, postoperative complications, and outcomes were collected retrospectively from hospital electronic medical records. Data were analyzed using STATA 14.0 (StataCorp. USA), and summary statistics were presented as medians (ranges).

Results. During the period of May 2006 to May 2013, 22 patients with HD undergoing LAEPT were identified (**Table 1**). Among them, 17 were boys and 5 were girls. Their age at surgery ranged from 9 months

Table 1 - Clinical characteristics and outcome of 22 children with Hirschsprung's disease undergoing laparoscopic-assisted endorectal pull-through procedures.

Charactarestic	n	% or (range)
Median age (years)	6.5	(1-14)
Boy:Girl	17:5	77:23
Conversion to open surgery	0	0
Median operation time (minutes)	160	(105-190)
Median blood loss (ml)	34	(20-50)
Median time to stool and flatus (hours)	22	(7-43)
In-hospital deaths	0	0
Median postoperative hospital stay (days)	8	(6-13)
Defecation frequency postoperatively in first 3 months (time/day)	10	(4-16)
Defecation frequency postoperatively after 6 months (time/day)	2	(1-3)
Enterocolitis	1	4.5

to 14 years, and the median age was 6.5 years. The median follow-up was 9 months (range, 6-12 months). The LAEPT procedure was performed on 20 patients with common segment type HD and on 2 patients with short segment type HD.

The procedure was completed successfully in all the 22 patients without conversion to open surgery. No patient died in the perioperative period. The median length of the surgical procedure was 160 minutes (range, 105-190 minutes), and the median intraoperative blood loss was 34 ml (range, 20-50 ml). The patients exhausted stool and flatus after 7-43 hours postoperatively (median 22 hours) and took full enteral feeding by 72 hours. Patients were discharged from 6-13 days postoperatively (median 8 days). Postoperative pathological examination showed normal ganglion cells in the non-thickened or mildly thickened dilated colons, and the aganglionic segment was completely resected.

The defecation frequency postoperatively was 4-16 times per day in the first 3 months. After 6 months, the defecation frequency was 1 to 3 times per day without constipation, soiling, or stoma stenosis. One child suffered enterocolitis at one month postoperatively, which was resolved using conservative treatment within 15 days. After 3 to 6 months of follow up, the dilated colonic segments disappeared in 20 cases using barium enema.

Discussion. Hirschsprung's disease is a congenital disorder characterized by the absence of ganglion cells in the submucosal (Meissner's) and myenteric plexus along a variable length of the intestine. Characteristically, it involves the distal colon and a variable length of the contiguous proximal colon; in some cases, it can involve the entire colon and, in rare cases, the entire intestinal tract.[5] The aganglionic bowel in DH lacks normal motility; thus, the upstream bowel becomes dilated secondary to functional obstruction. Patients usually have difficulty stooling, leading to abdominal distention.

Recently, minimal access techniques have been introduced for dissection of the aganglionic part of the bowel.[6] Some surgeons advocate removing not only the aganglionic segment and transitional segment but also all of the dilated segment.[7] One child with HD admitted to our hospital had to undergo a loop colostomy at diagnosis of intestinal obstruction for decompression of the dilated bowel proximal to the aganglionic segment. His 0.3 cm thick descending colon with a diameter of 12 cm was pulled out during a colostomy. Three months after the 2-stage operation, the original dilated and mildly hypertrophic colon returned to normal and presented normal appearing haustra coli. Another 2 patients with HD, one was transferred from another hospital half a year and the other was transferred one year after sigmoidectomy, underwent secondary Duhamel procedure. During the operation, we observed that the obviously hypertrophic colon could not recover normally.

Generally, when an intestinal obstruction is treated by an operation, it is unnecessary to remove all the dilated bowel, regardless of the dilated small intestine or colon. It is suggested that the secondary dilated colon without hypertrophy or with mild hypertrophy will recover normally if the intestinal obstruction is resolved. For these reasons, when we performed the procedure in these 22 cases, the dilated segment was preserved while the thickened dilated segment was removed. It was not difficult to identify the boundary between the dilated colon, and the thickened dilated colon by gross visual inspection intra-operatively. Frozen examination of seromuscular biopsies obtained laparoscopically between the dilated colon and thickened colon at the transition zone at the beginning of the operation could save expense and time, so it is well-accepted in developing countries. It is not necessary to take down the splenic flexure of the colon. Postoperative histopathological examination showed normal ganglion cells in non-hypertrophic or mildly hypertrophic dilated colons. Dilated colonic segments returned to a normal caliber that was determined on follow-up barium enema examination after 3-6 months postoperatively.

We emphasized that thickened dilated colon should be resected because the smooth muscle cells of this segment were highly proliferative, and the ganglion cells were degenerated and reduced in number. As a result, it is impossible for this segment to restore normally.

Recently, the treatment of HD has become less invasive.[8] In this study, the clinical result was satisfactory. There were no postoperative complications such as constipation, soiling, or stoma stenosis. Surgical treatment should be cautiously considered for patients with IND, severe enterocolitis, or poor performance status. It was reported that 25-35% of HD patients have IND.[9] Some investigators have suggested that the postoperative persistent constipation of HD is caused by IND.[10,11] An entire gastrointestinal barium meal combined with multi-temporal abdominal radiography is a convenient method to assess gastrointestinal motility and to distinguish physiological and pathological changes simultaneously. This method could be helpful for screening patients with IND preoperatively.

Our study has limitations given the retrospective nature and the limited number of patients. Thus, future prospective study with the large size of patients is needed. In conclusion, LAEPT procedure is safe for HD patients with satisfactory results.

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