

腹腔镜辅助下治疗先天性巨结肠： 文献综述和技术报告

Laparoscopic-Assisted Procedures for the Treatment of Hirschsprung's Disease: Review of the Literature and Technical Report

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1 概述

先天性巨结肠(Hirschsprung's disease, HD)又称肠管无神经节细胞症,是由神经嵴细胞迁移发育成肠神经系统过程停顿所致。大多数情况下,无神经节细胞段和正常神经支配结肠之间的移行段位于结肠与直肠交界处或以下,约占 75%;而长段型和全结肠型无神经节细胞症并不罕见^[1]。1948 年,Swenson 和 Bill 最先描述了该疾病的外科治疗策略,明确指出切除无神经节细胞的直肠而并非扩张的近端结肠的必要性^[2]。从 20 世纪中期起相继报道了腹腔镜与先天性巨结肠的三个标准术式联合运用的成功病例。腹腔镜具有伤口小、术后疼痛轻、肠道功能恢复快、术后恢复时间短以及良好美容效果等优点。本文就运用腹腔镜治疗先天性巨结肠的文献进行综述,并介绍我们应用腹腔镜技术经肛门拖出的手术方法。

2 Review of the Literature

The first published discussion on the use of laparoscopy in the treatment of Hirschsprung's disease was in 1994, when Curran and Raffensperger from the Children's Memorial Hospital in Chicago, Illinois, USA published the results of an animal study in which the Swenson pull-through procedure was performed on 13 dogs^[3]. The anastomosis was fashioned in the same manner as in the open procedure. These animals were all found to have formed stools and were continent of stool and urine, and at autopsy and it was determined that the suture lines were intact with no evidence of hemorrhage or abscess. The authors concluded that a laparoscopic approach could be safely attempted in humans. These surgeons subsequently reported the successful practice of this procedure in three of four patients in an addendum to the original manuscript^[3]. One patient required conversion to an open procedure. A follow-up manuscript published the following year reported a decreased length of stay, decreased blood loss, and decreased exposure to nosocomial pathogens in those undergoing the laparoscopic-assisted

approach, however none of these factors were quantified^[4]. Curran and Raffensperger later published a comparison between eight patients who underwent laparoscopic Swenson procedures to ten who underwent the open procedure in an overlapping 19 month period^[5]. Statistically significant decreases in post operative length of stay (5.25 versus 8.8 days) and time until oral food intake (2.75 versus 5 days) were seen in the laparoscopic group. No early complications occurred in the laparoscopic group, but one patient later required re-operation for an adhesive small bowel obstruction. The mean cost associated with the laparoscopic procedure was about twice the mean cost of the open procedure. The follow-up was deemed too short to compare functional results, but the authors concluded that the laparoscopic procedure was safe and resulted in decreased morbidity in their small sample of patients. Despite the fact that the operation was more costly in this cohort, it is generally felt that overall the laparoscopic procedure is cost-effective due to decreased hospital stay^[6]. Bufo et al from Memphis, Tennessee, USA quantified the difference in cost between 15 patients who underwent a one stage laparoscopic assisted Soave or Duhmael procedure and 18 patients who underwent two-stage (colostomy, then open pull-through at a later date) Duhamel procedure^[7]. The overall costs for the laparoscopic technique were nearly half those for the open procedure. The authors concluded that the laparoscopic approach was more cost-effective despite the obvious limitations of the study. Hoffmann et al from Germany also reported a series of seven patients who underwent the laparoscopic-assisted Swenson's procedure^[8]. The patients were aged four weeks to 13 years. There were no intraoperative or early postoperative complications, but three patients developed anastomotic strictures.

The first description of a laparoscopic-assisted endorectal pull-through procedure was published by Georgeson et al from the Children's Hospital of Alabama, USA in 1995^[9]. They described their

technique used in 12 patients ranging in age from three days to six years. The technique is the basis for that described in detail below, and is based on the Soave procedure which involves a mucosectomy. They successfully used either the Harmonic scalpel or bipolar cautery to divide the superior rectal vessels and the inferior mesenteric artery if needed (depending on the length of the aganglionic segment). No case required conversion to open, and operative time was just over two hours which was similar to their times for their open operation. One patient required a blood transfusion intraoperatively. In their 12 patients oral feeding was resumed between one and three days postoperatively. Rectal dilation was performed by parents using increasingly larger Hegar dilators, until a number 14 dilator could be passed. Follow up was deemed was too short (mean six months) to assess bowel function and continence, but no patient developed enterocolitis or anal strictures. The authors concluded that their technique was safe with good early results.

Another early series using the laparoscopic-assisted Soave procedure was published by Rothenberg and Chang from Denver, Colorado, USA^[6]. Fifteen patients, ranging in age from seven days to eight years (all but three patients were less than eight months old) underwent the procedure. Eleven had primary pull through while the remaining four had undergone previous colostomy. Three or four ports were used and the hook attachment for the Harmonic scalpel was used for the abdominal dissection. The average operating time was two hours 50 minutes. The average time to oral feeding was 1.3 days and the postoperative length of stay was 3.4 days. Daily anal dilation was started at four weeks. One case of anal stenosis was reported in a patient who was non compliant with follow up for some time. This patient required dilation of the colo-anal anastomosis under anesthesia. Follow up was between two and 28 months. The patients were all continent, and were passing two to six stools per day. The authors enthusiastically endorsed this technique, and barring problems with long term bowel control, advocated it as the recommended operation for the treatment of Hirschsprung's disease.

In 1999 Georgeson et al reviewed their experience to date with the procedure referred to in their original report^[10]. The results of 80 patients from six centers ranging in age from three days to eight years were included. Seventy of the patients were under six months of age. The average postoperative stay was 3.7 days, although most of the patients were discharged within the first three days. Most of the children were too young to evaluate fecal incontinence, but 18 of the older children were found to be continent. The series included 11 patients with a transition zone proximal to the sigmoid colon and one with total colonic agangliosis. Average operative time was two hours twenty seven minutes and two of the cases were converted to an open procedure. One patient required an intraoperative transfusion. Seventy-four patients

(92.5%) had a bowel movement within 24 hours after surgery. Four patients required re-operation with diversion; two for anastomotic leaks, one for severe enterocolitis, and one for an associated congenital malformation. Half of the patients underwent routine postoperative anal dilation, while only one required forceful dilation under anesthesia due to chronic constipation. Six patients reported chronic diarrhea (greater than 6 months). Two were found to be lactose intolerant and were successfully treated. Two were found to have not formed a rectal reservoir, and their diarrhea resolved after Duhamel procedures were performed to enlarge their rectal capacitance. The last two patients were successfully treated with a constipating diet and anti motility agents. In 2004, Georgeson and Robertson reviewed their technique again and suggested that in patients with extended or colonic forms of Hirschsprung's disease a laparoscopic Duhamel procedure is preferred to the Soave as it results in a larger reservoir in these patients who will have a short or absent colon^[11]. As well, they outlined the contraindications to a primary laparoscopic-assisted pull-through procedure; associated life threatening anomalies, deteriorating general health, severe enterocolitis, and severe dilatation of the proximal bowel. The authors recommended that these patients undergo a leveling colostomy. The authors also addressed the role of primary endorectal pull-through without laparoscopic guidance. This procedure omits the laparoscopic biopsies and preparation of the distal mesocolon before the transanal dissection. They believe that identification of the normally innervated bowel above the transition zone should be achieved before beginning any mesenteric or rectal dissection and the laparoscopic procurement of seromuscular biopsies greatly facilitates this aim. An additional advantage of the laparoscopic assistance of the endorectal pull-through includes the versatility afforded by this technique for performing almost all surgical strategies for correcting Hirschsprung's disease. Laparoscopic assistance allows for precise dissection that can be used to develop a mesocolic pedicle when indicated, and allows for the easy release of tethering ligaments, which can diminish the tension on the coloanal anastomosis. It also allows for an evaluation for rotational abnormalities in the pull-through segment.

The first case report of a laparoscopic assisted Duhamel procedure was published by Smith et al in 1994 from Memphis, Tennessee, USA^[12]. Bonnard et al from France subsequently reported a series of seven patients with extended (two patients) or colonic (five patients) aganglionosis who were treated with laparoscopic-assisted Duhamel procedures^[13]. All patients had a diverting colostomy within 45 days of age. The average time between diversion and the Duhamel procedure was 11 months. Three complications were noted; one wound infection, one complicated fever course, and one bowel perforation with peritonitis requiring diversion. No soiling or incontinence was present in any of the patients after a

median 12.9 months of follow up. De Laguisse et al also from France reported a series of six primary laparoscopic-assisted Duhamel pull through procedures^[14]. The patients ranged in age from two days to five years. No cases were converted and the average operating time was three hours and 10 minutes. One patient developed a retro-rectal abscess that required surgical drainage on the 12th postoperative day. The two day old patient developed generalized sepsis. The four patients without early complications were discharged between five and seven days postoperatively. No enterocolitis was seen in this series. One patient had soiling three months postoperatively while the others were continent at last follow up.

Other North American experience is from Canada where Singh et al compared the incidence of enterocolitis between 25 patients who underwent laparoscopic-assisted Swenson's procedure and 27 who had primary trans-anal Swenson's pull-through^[15]. They found that the incidence of enterocolitis was 16% in the laparoscopic group and 7.4% in the primary trans-anal pull-through group. The follow up period was shorter in the primary pull through group, however, and in no case did a patient require reoperation.

Authors from around the world have also published series of laparoscopic-assisted pull through procedures. Kumar et al from Australia reported on the follow up of 29 patients who underwent laparoscopic assisted Swenson procedures^[16]. Sixteen patients with a median age of five days underwent primary repair, while the remaining children had undergone previous diversion. Three of these patients had extended aganglionosis and underwent laparoscopic colectomy in addition to the Swenson procedure. Median operating time was one hour and 45 minutes, and the median time to tolerating a regular diet was 48 hours. No patients required open conversion. Postoperative complications included post-operative ileus in three patients and enterocolitis in two. Median hospital stay was 4 days. Median follow up was 2.2 years, with satisfactory continence seen in 15 of the 19 children older than age three at the time of last visit. The authors used unipolar cautery for the laparoscopic dissection arguing that this is a safe and cheaper alternative to the Harmonic scalpel advocated in some of the previously mentioned studies.

Antao and Roberts from England also reported their results after performing six laparoscopic assisted endorectal pull-through procedures^[17]. Two cases required open conversion due to pelvic adhesions. In all cases the entire necessary mobilization of the colon as well as the acquisition of seromuscular biopsies to localize the transition zone were performed laparoscopically. Median operative time was two hours and fifteen minutes. All children tolerated a regular diet by 48 hours. Median hospital stay was six days. Two cases of enterocolitis that resolved with conservative management were the only immediate

postoperative complications. No patients had incontinence or stool soiling with a median follow up of twelve months.

Most recently Ghirardo et al from Italy retrospectively compared 21 patients who underwent open Duhamel pull through procedures to ten who underwent the laparoscopic assisted operation^[18]. The Harmonic scalpel was used for dissection of the mesentery and mesenteric vessels in the laparoscopic cases. All of the patients had rectosigmoid transition zones. The mean age in the open group was 14.6 months and 14 months in the laparoscopic group and mean postoperative length of stay was 10 days and 6.8 days respectively. Operative times were greater in the open cases; four hours 57 minutes compared to four hours and 13 minutes. Mean time to feeding was 4.9 days in the open group and 3.3 days in the laparoscopic group. Early complications in the open group were one case of ileo-ileal intussusception, while late complications included four cases of constipation. Four of the above five children required reoperation, with one case of constipation managed medically. Early complications in the laparoscopic group included one case of intra-operative transection of the ureter, and late complications were five cases of constipation, four due to overly long residual colonic wall. All other patients in both groups had normal bowel function at the time of last follow up. The authors were encouraged by these results concluding that the incidence of complications was the same in both groups, but the laparoscopic technique takes less time and allows for earlier discharge from the hospital.

The only prospective study on the laparoscopic assisted pull through appears to be by Ishihara et al from Japan^[19]. These authors developed a standard series of forms to evaluate complications including constipation, incidence of enterocolitis, and continence. Continence was categorized as "normal", "good", "fair" or "poor" based on prospectively determined definitions. Thirty-three patients underwent laparoscopic assisted Soave pull through operations. The mean age of the patients was 11 months and mean follow up time was four years. No intraoperative complications occurred. In 20 subjects over three years of age with a follow up period of 12 months or greater continence was found to be "normal" in five patients, "good" in ten patients, "fair" in four, and "poor" in one. Overall three patients developed enterocolitis. None of the patients reported constipation. The authors concluded that this procedure is safe but that longer term follow up is necessary to assess anal continence.

3 Conclusions

Laparoscopic-assisted procedures for the treatment of Hirschsprung's disease have been performed safely in humans for over ten years. The three established procedures for the treatment of Hirschsprung's: Soave, Swenson's and Duhamel procedures, are all feasible with a laparoscopic-assisted approach. Even in cases of extended or total aganglionosis the successful application

of laparoscopy has been reported. We advocate the use of the laparoscopic-assisted modified Soave technique described below. To the best of our knowledge no randomized controlled trials have been done comparing the open and laparoscopic-assisted techniques, but from our review of the literature, we conclude that the laparoscopic-assisted procedure is as safe as the open approach, with significant reductions in length of stay and return of bowel function. The laparoscopic-assisted approach also provides a definitive method of determining the length of the diseased segment prior to any rectal dissection, which is not possible with the primary trans-anal approach. The above findings, in addition to the general benefits of laparoscopic surgery including improved cosmetic result and decreased pain from laparotomy have made the laparoscopic-assisted pull-through our preferred procedure for the treatment of patients with Hirschsprung's disease.

4 Operative technique

Pre-operatively, infants undergo bowel preparation with rectal irrigations using a 28F red-rubber catheter and 10 cc/kg of normal saline three times daily. Older children undergo a similar preparation, with the potential addition of enteric lavage. Broad-spectrum intravenous antibiotics are administered within one hour prior to the time of incision to prevent surgical site infection. Infants and small children prepared with a povidine-iodine solution circumferentially from the level of the xiphoid process to the toes. Older children are positioned in stirrups in the lithotomy position. A three or four trocar technique is used with the first trocar (5 mm) placed through the umbilicus. Our preference in infants is to use the Veress needle technique through the umbilicus. A second trocar (3 mm) is placed subcostally in the right upper quadrant and a third trocar (3 mm) is placed in the anterior axillary near McBurney's point. The fourth trocar (3 mm) can be placed in the left upper quadrant if needed for traction on the colon. It is imperative to confirm the level of the transition zone at the beginning of the intra-abdominal procedure prior to proceeding with any rectal dissection (Figure 1). A seromuscular biopsy is procured by grasping the serosa with a 3-mm Maryland grasper. The seromuscular wall is tented upward and the endoscopic Metzenbaum scissors are used to incise down to the mucosa (Figure 2). A small flap is lifted upward and the biopsy completed with the scissors. Perforation or bleeding at the biopsy site can be closed with a figure-of-eight or interrupted silk suture. Even in the absence of injury, a marking stitch is placed at the biopsy site where ganglionic bowel is confirmed by the pathologist to be used as a landmark during the trans-anal portion of the procedure (Figure 3). Usually 2 or 3 biopsies are obtained and sent to the pathologist for rapid frozen section analysis. If aganglionosis is noted, the biopsies should be continued as far proximally as necessary to identify normal ganglionic bowel. If the pathologist is unable to detect normal colonic biopsies proximal to the mid-transverse colon, further biopsies are taken but the

procedure is converted to a laparoscopic-assisted colostomy at the level where ganglionic bowel is identified. No dissection of the rectum or its mesentery should be started until the location of the transition zone has been confirmed by the pathologist.

Once the proximal margin of the transition zone is firmly established, dissection of the mesocolon may be initiated. In most small infants, this dissection can be accomplished with a 3-mm hook electrocautery. In children greater than 1 year of age, the Harmonic scalpel is preferred to prevent bleeding when dividing the vessels in the mesocolon; however, the 3-mm port in the right lower quadrant must be up-sized to a 5-mm port. The mesocolon should be divided adjacent to the colon in the aganglionic portion of the bowel (Figure 4). If the aganglionic segment is longer than the mid-sigmoid colon, the colon to be pulled through should be mobilized from the lateral side wall by dissecting the peritoneal attachment thereby medializing the left colon (Figure 5). The splenic flexure may also be taken down using the hook electrocautery or harmonic scalpel in a similar fashion to avoid tension on the anastomosis. This dissection of the mesocolon renders the trans-anal dissection a simpler and faster procedure. The author prefers to extend the colon resection 5 to 10 cm above the transition zone to avoid pulling through a hypoganglionic segment of colon. The proposed segment to pull through is then brought into the pelvis to gauge whether a tension-free anastomosis may be achieved.

The pneumoperitoneum is then evacuated and attention is turned to trans-anal portion of the procedure. Identifying the appropriate dissection plane is facilitated by placing circumferential traction sutures (5-0 silk for infants) approximately 0.5 cm proximal to the dentate line. Placement of these sutures may be aided by scoring the mucosa at the appropriate level using the needle-tip cautery. The traction sutures allow for the endorectal dissection to proceed more precisely. The endorectal dissection is continued proximally in a circumferential manner until the muscular cuff of the rectal wall intussuscepts freely, or for approximately 5 cm. The posterior wall of the rectal cuff is then divided 1 to 2 cm below the endorectal mucosal dissection. Once a free plane is found, the division of the muscular rectal wall is continued circumferentially freeing up the intra-abdominal colon from the muscle sleeve. The cuff should be no more than 5 cm long and is divided posteriorly all the way to the level where the endorectal dissection was initiated. Splitting the cuff posteriorly is important to prevent entrapment of the neo-rectum by the contracted muscular sleeve. Furthermore, the contracted muscular cuff may constrict the neo-rectum leading to the inadequate development of a rectal reservoir. The aganglionic colon is pulled down through the muscle sleeve and out trans-anally until the previously marked ganglionic portion of the colon is identified. At this point, a primary anastomosis between the neo-rectum and the anus is performed with interrupted fine absorbable sutures (5-0 Vicryl).

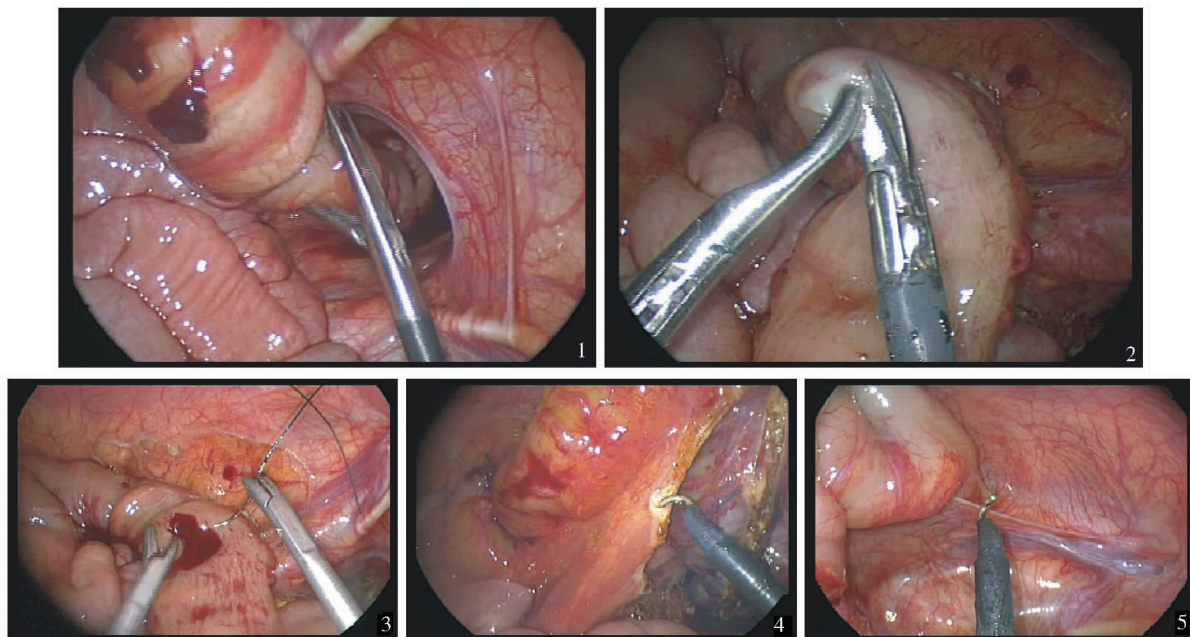


Fig. 1 The transition zone between contracted aganglionic rectum and the more dilated rectosigmoid junction is depicted. The 3-mm bowel grasper is adjacent to the contracted aganglionic rectum. **Fig. 2** A 3-mm Maryland dissector and a 3-mm endoshears are used to obtain a seromuscular biopsy of the sigmoid colon. **Fig. 3** Two 3-mm needle drivers are used to mark the area where the presence of ganglion cells were confirmed by the pathologist. A 2-0 silk suture on a ski needle is used here. **Fig. 4** The 3-mm hook and electrocautery are used to divide the mesocolon adjacent to the ganglionic bowel. **Fig. 5** If necessary, the 3-mm hook with electrocautery can be used to take down the lateral attachments of the left colon to gain length for a tension-free anastomosis.

The pneumoperitoneum is reinstituted and visualization of the pull-through segment is performed to ensure there is no twisting of the mesentery and there is no tension on the anastomosis. The trocars are removed and the pneumoperitoneum evacuated. The umbilical trocar site is closed with an absorbable braided suture for the fascia (2-0 Vicryl) and rapidly absorbing monofilament suture in the skin of the umbilicus (5-0 Chromic). The other trocar sites are closed with strips or absorbable sutures.

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