

# Laparoscopic Management of Hirschsprung's Disease

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## ABSTRACT

Laparoscopy has improved the outcome and management of Hirschsprung's disease. The most commonly seen Hirschsprung's disease with transition zone in the rectosigmoid is done in the neonatal period as one-stage transanal endorectal pull-through (ERPT) procedure. In all other children, it must be one-stage laparoscopy-assisted transanal pull-through procedure. Advantage of a laparoscopic procedure is that a biopsy report of the level of transition zone and the normal ganglionic segments is obtained before the dissection is begun. It allows peritoneal dissection and isolation of marginal artery under vision. Also adequate length of colon can be dissected free of the attachments under vision.

**Keywords:** Hirschsprung's disease, Laparoscopy, Transition zone.

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## INTRODUCTION

Twenty-five years back in the postoperative ward of our pediatric surgical unit, there would have been at least one child with both legs and arms tied to the four posts of a bed, and a pair of straight arteries, larger than the child, stuck into the anus, and the anxious and frightened mother standing beside the bed with her hand stuck to the chin. Anybody can read what is happening in that mother's mind. This was the situation of a child with Hirschsprung's disease. Thanks to the gastrointestinal stapler, this situation soon changed. In this article, I am reviewing various articles and studies to evaluate, further, how laparoscopy has influenced the treatment of these children.

## AIM

To assess whether laparoscopy has influence on the outcome of treatment of Hirschsprung's disease.

## MATERIALS AND METHODS

Research materials taken are the various articles published in PubMed, ScienceDirect, Journal of Paediatric

Surgery, Indian Journal of Paediatric Surgery, Seminars in Paediatric Surgery, and Annals of Surgery. The research gave a wide range of research material of which relevant articles were selected. The criteria for selection of papers were the number of cases included in the study and the duration of study.

Swenson and Bill<sup>1</sup> described surgery for Hirschsprung's disease. Ever since, surgery has remained the cornerstone for treatment of Hirschsprung's disease. At the outset, the surgical procedure was a three-stage procedure. It involved the formation of colostomy and after a period of colonic washouts, the second stage of anorectal pull-through was done, keeping a protective colostomy and later, in a third stage, closure of the colostomy was done.

Soave<sup>2</sup> described the endorectal pull-through procedure to exclude the disadvantages inherent to pelvic dissection.

Duhamel<sup>3</sup> described his procedure of rectal pull-through and anastomosing with a part of aganglionic rectum longitudinally. Later surgeons started removing the colostomy along with the pull-through procedure and the surgery became a two-stage procedure.

The open abdominal procedure with anorectal pull-through was associated with various complications. As it was the staged procedure and colostomy was done first, the complications inherent to colostomy like stenosis, prolapse, and abdominal excoriation were prone to occur even before the definitive procedure. The complications of laparotomy like wound infections, intra-abdominal and pelvic abscesses, wound dehiscence, leak at the anastomotic site, leading to long hospital stay, and delayed complications like intestinal adhesions and stenoses at the anastomotic site needed repeated hospital admissions. Enterocolitis was another major problem and patients developed enterocolitis before and after surgery.

Fontana et al<sup>4</sup> in a study of 82 infants who had undergone Soave or Duhamel open procedures were reviewed after 20 years. They found only 60 and 67% of the children had an uneventful recovery; 20 and 29% of them needed reoperation, and short-term continence for both was around 50% and became 100% continent about 15 years of age.

So et al<sup>5</sup> published that Hirschsprung's disease in the newborn does not mandate the performance of a preliminary colostomy. They treated enterocolitis by a precise regimen of colonic irrigations and then did the

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endorectal "pull-through" procedure. They found it was safe and effective when performed in the neonatal period.

Laparoscopy in Hirschsprung's disease<sup>6</sup> was being done from 1992, and it was limited to biopsy, followed by a minilaparotomy and pull-through procedure, as the anesthetic complications were very high. Georgeson et al<sup>7</sup> reported laparoscopy-assisted colon pull-through procedure.

Between November 1993 and September 1994, they did 12 primary laparoscopic colon pull-through procedures in infants and children. The patients' ages ranged from 3 days to 6 years. The primary diagnosis in all 12 patients was Hirschsprung disease. All children had their operations without construction of preoperative or postoperative colostomy. They mobilized sigmoid colon and proximal rectum laparoscopically. A submucosal sleeve was developed transanally to meet the dissection from above. The colon was then pulled down in continuity, divided above the transition zone, and secured to the anal mucosa 5 to 10 mm above the pectinate line. Mean postoperative stay was 4 days. Laparoscopic visualization provided clear delineation of pelvic structures even in small infants. He concluded that laparoscopic pull-through required no more time than similar open procedures, averaging just over 2 hours, and morbidities associated with colostomy formation and closure and the inconvenience of colostomy care were avoided with a one-stage technique. Postoperative sequelae like wound dehiscence and wound infection were avoided due to the laparoscopy.

Georgeson et al<sup>8</sup> again reported on laparoscopy-assisted pull-through for 80 children.<sup>9</sup> The age at surgery ranged from 3 days to 96 months. They discussed the outcome of primary laparoscopic pull-through in 80 patients performed at six pediatric surgery centers over 5 years. The average length of the surgical procedure was 2.5 hours. Almost all of the patients passed stool and flatus within 24 hours of surgery. The average time for discharge after surgery was 3.7 days. The transition zone was identified by seromuscular biopsies obtained laparoscopically. The colon pedicle preserving the marginal artery was also fashioned endoscopically. The rectal mobilization was performed transanally. They used the endorectal sleeve technique with the anastomosis performed transanally 1 cm above the dentate line.

Sixty-nine (86%) of their 80 patients had a transition zone in the rectum or sigmoid colon. The remaining 11 patients had a transition zone proximal to the sigmoid colon; one of these patients had total colonic aganglionosis. Seventy (87.5%) of the children were younger than 6 months of age at the time of the pull-through procedure. Operative time averaged 147 minutes. Blood loss was <10 cc per patient; only one patient had blood transfusion. They found that there was rapid postoperative recovery

and bowel movement started within 24 hours except for six patients. The mean time to discharge was 3.7 days.

They did not find any instances of anastomotic stricture, postoperative bowel obstruction, wound infection, prolonged ileus, pelvic or intra-abdominal abscesses, or wound dehiscence. Ten (12.5%) of the 80 patients were readmitted to the hospital for complications. Four of these 10 patients required postoperative diversion of the gastrointestinal tract.

Teitelbaum et al<sup>10</sup> published a paper describing a single-stage primary ERPT procedure. Their study included 78 infants who underwent a primary ERPT procedure. Study duration was from May 1989 to September 1999.

Mean age at the time of ERPT was 17.8 days of life. They compared primary ERPT with a two-stage approach. Their study showed a trend toward a higher incidence of enterocolitis in the primary ERPT group compared with those with a two-stage approach (42.0 vs 22.0%). Other complications they found were either lower in the primary ERPT group or similar, including rate of soiling and development of a bowel obstruction. Median number of stools per day was two at a mean follow-up of  $4.1 \pm 2.5$  years, with 83% having three or fewer stools per day. They found that performance of a primary ERPT for Hirschsprung's disease in the newborn is an excellent option. Results were comparable to those of the two-stage procedure. They also found a greater incidence of enterocolitis as compared with the two-stage procedure, but the multistage pull-through had higher incidence of readmissions than an ERPT. They found the ERPT procedure left no scars and was associated with less postoperative pain and discomfort, and shortened hospital stay. Short-term function was reported to be very similar to that after open or laparoscopic procedures. They suggested that one-stage neonatal repair of Hirschsprung's disease was associated with less cost and demand of resources without jeopardizing functional outcome.

Westler and Rintala<sup>11</sup> did a study in 40 patients who had undergone transanal ERPT for Hirschsprung's disease between January 2000 and February 2003. Here, they divided patients into three groups: Patients with neonatal primary pull-through (group I, 15 patients), patients operated on beyond the neonatal period (group II, 11 patients), and patients with a previous colostomy (group III, 14 patients). All colostomies except one were taken down and pulled through concomitantly with the transanal procedure.

They found no difference in median hospital stay (group I, 5 days; group II, 4 days; group III, 5 days) and median time to full oral feedings (group I, 4 days; group II, 2 days; group III, 3 days) between groups. Two patients (group III) had immediate postoperative prolapse of the

pulled-through colon that was reduced without further sequels; one (group III) had infection of the stoma closure wound. Perianal skin rash was more often in neonatal patients (group I, 10 of 15; group II: 4 of 11; group III: 8 of 14). Anastomotic dilatation regimen was required more often in neonatal cases (group I, 6 of 15; group II, 1 of 11; group III, 2 of 14). Enterocolitis requiring hospital care occurred in two patients (group I), and five further patients (group II, 1; group III, 4) were treated as outpatients for symptoms, suggesting mild enterocolitis or bacterial overgrowth. They concluded that transanal ERPT in neonatal patients was as feasible and safe as in older children. Temporary postoperative skin rash occurred more frequently in neonatal patients, and postoperative dilatations were required more often than in older children.

In another article, Minford et al<sup>12</sup> compared the outcome of Duhamel's operation and transanal ERPT. In their study, 70% were neonates (Duhamel, 24 of 34; transanal endorectal coloanal anastomosis, 26 of 37). They assessed the functional outcome. Functional outcome was similar in the two groups. They found that ERPT and Duhamel procedures had similar medium-term functional outcomes. The ERPT had a high incidence of postoperative enterocolitis and transient stricture formation but was suitable for single-stage neonatal treatment of Hirschsprung's disease.

Lu et al<sup>13</sup> also found a high rate of postoperative enterocolitis in neonates undergoing transanal pull-through.

Teeraratkul<sup>14</sup> in an article described the limitation of the procedure of transanal pull-through. Retroperitoneal fixation of the descending colon could not be dissected by the transanal route, especially if it needed mobilization of the splenic flexure. The length of bowel that could be dissected varied from 9 to 25 cm.

Langer et al<sup>15</sup> compared transanal Soave with the open approach to see whether it offers any advantage and whether routine laparoscopic visualization is necessary. He studied 37 children. They had children with open Soave 13, transanal Soave with laparotomy 9, and transanal Soave with selective laparotomy or minilaparotomy 15.

In two patients with transanal Soave, they had to do laparoscopy for a long segment in one and small umbilical incision for mobilization of the splenic flexure in another.

There were no differences in operating time, and intraoperative complications, such as enterocolitis, stricture, or cuff narrowing, but hospital stay was longer in open Soave and there were four reoperations in open Soave, adhesion obstruction, twisted pull-through, and recurrent aganglionosis being the causes. They concluded that transanal pull-through had shorter hospital stay and low incidence of intra-abdominal adhesions. Laparoscopic visualization was needed for children who are at high

risk for long segment disease. Transanal approach was also supported by Dela Torri with shorter hospital stays and fewer complications.

Schofield and Ram<sup>16</sup> compared between open Duhamel's (OD) and laparoscopy-assisted Duhamel's (LD) procedure.

From 11 articles, 456 patients were included (253 OD, 203 LD), with no significant difference in age at surgery and length of follow-up ( $p > 0.05$ ). The open group had a significantly greater incidence of soiling/incontinence (11 vs 4%;  $p = 0.02$ ) and further surgery (25 vs 14%;  $p = 0.005$ ), longer hospital stay (9.79 vs 7.3 days;  $p < 0.00001$ ), and time to oral feed (4.05 vs 3.27 days;  $p < 0.00001$ ). Operative time was significantly longer in the laparoscopic group (3.83 vs 4.09 hours;  $p = 0.004$ ). There was no significant difference in incidence of enterocolitis (15 vs 10%;  $p = 0.14$ ) and constipation (23 vs 30%;  $p = 0.12$ ). They have compared the quality of life and Fecal Continence Index in children with Duhamel's operation and transanal pull-through and normal children. They have found that both quality of life and Fecal Continence Index were lower than normal children in both groups and transanal pull-through had still a lower score.

## DISCUSSION

Open laparotomy and transanal pull-through procedures whether Duhamel's or Swenson's or Soave are associated with immediate and delayed complications and morbidity. This includes wound infections, intra-abdominal and pelvic abscesses, wound dehiscence, anastomotic leak, stricture at the anastomotic line, intestinal adhesions and intestinal obstructions, constipation, incontinence problems, and perineal excoriations in addition to the enterocolitis. Colostomy in addition has its own complications like prolapse, herniation, stenosis, abdominal wall excoriation in addition to the stoma management problems.

But the transanal pull-through procedures were associated with fewer complications even though various authors have reported increased episodes of enterocolitis. So the endorectal dissection became the dominant minimal access procedure, which could be done easily in the neonates without entering the peritoneal cavity. The peritoneal dissection is avoided and hence, its early and late complications.

Number of days of hospital stay was reduced, but the level of resection was an arbitrary choice of visualized transitional zone. The resected ends could be sent for biopsy, but necessitated opening up of the abdomen if the level was higher up. Also if the transition zone was above the usual rectosigmoid junction, peritoneal dissection especially at the splenic flexure needed laparotomy or a minilaparotomy at the umbilicus. Some studies

noted an increased number of episodes of enterocolitis postoperatively, and perineal excoriation also was noted as a major problem that gradually resolved.

These difficulties were overcome by the laparoscopy-assisted transanal pull-through approach. The laparoscopy-assisted transanal ERPT allows early biopsies to determine the extent of aganglionic and dysfunctional bowel before dissection of the rectum and mesocolon begins. Frozen section biopsies are taken from multiple sites before the dissection begins. Dissection of the peritoneal attachments from the left colon and isolation of the marginal artery can be done laparoscopically. Total colonic aganglionosis is managed by a laparoscopy-assisted Duhamel procedure.

The International Pediatric Endosurgery Group prepared the guidelines for laparoscopic approach in 2004 and summarized that the implementation of laparoscopy allowed the surgeon to safely use the concept of pull-through while eliminating the major source of morbidity, which consists of colostomy and its consequences, postoperative immediate and late complications like wound dehiscence, wound infection, and intestinal adhesion obstruction. Laparoscopic technique avoids internal and external scarring. There is rapid recovery and less perianal excoriation. It has also been mentioned about the excessive stretching and damage of the anal sphincter during rectal dissection especially in the neonates, which gradually recover in time, and this can be avoided by a laparoscopic rectal dissection.

But in neonates with classical rectosigmoid transition zone, endorectal dissection has become the dominant minimal access procedure because of ease and reliability. Creation of pneumoperitoneum can be avoided and Surgery can be done by any surgeon without laparoscopic skills makes it universal. Also, there is no abdominal scarring and the postoperative complications are minimized.

## SUMMARY

Laparoscopy has improved the outcome and management of Hirschsprung's disease to a large extent. The duration of postoperative recovery and hospital stay has decreased. Also it avoids the immediate and late complications of a laparotomy and peritoneal dissection like wound dehiscence and postoperative adhesion obstruction.

The most commonly seen Hirschsprung's disease with transition zone in the rectosigmoid is done in the neonatal period as one-stage transanal ERPT procedure.

In all other children, it must be one-stage laparoscopy-assisted transanal pull-through procedure. This will deal with the issue of getting a biopsy report of the level of transition zone and the normal ganglionic segment. It allows peritoneal dissection and isolation of marginal artery

under vision. Adequate length of colon can be dissected. Total colonic aganglionosis must be treated with laparoscopically assisted transanal pull-through procedure.

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