



Retrospective analysis of laparoscopically managed pediatric patients with Hirschsprung disease

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Abstract

Purpose Hirschsprung disease is characterized by aganglionic bowel segment and our goal is to resect it and pull down the normoganglionic bowel. The described techniques such as Swenson, Duhamel, and Soave have seen modification recently through the advent of laparoscopy. Our purpose is to assess the outcome of laparoscopy for management of patients with Hirschsprung disease.

Methods Twenty eight patients who underwent laparoscopic assisted pull through for Hirschsprung disease were included. Parameters studied included age at surgery, gender, type of pull through, location of transition zone, duration of surgery, post-operative complications, length of hospital stay (LoHS), and complications faced on follow up.

Results Twenty-eight patients who underwent laparoscopy-assisted pull through were included in this study. Median age at presentation was 24 months of age. 26 patients underwent Swenson, 1 underwent Soave, and 1 underwent Duhamel procedure. None required conversion to open. Three patients successfully underwent primary pull through (Swenson). Median duration of surgery was 4 h. Median LoHS was 6 days. Three patients faced complications during period of hospital stay. Every patient was followed up 3 weeks post-operatively and started on regular Hegar dilator program. Median follow up duration was 24 months. One patient developed enterocolitis, 2 developed constipation, and 1 had night time soiling on follow up. The patient with soiling demonstrated complete resolution of symptoms following bowel management for 3–4 months.

Conclusion Laparoscopic assisted pull through is a safe and feasible option in pediatric patients with a considerably low risk of complications.

Keywords Hirschsprung · Laparoscopy · Pediatric · Pull through

Introduction

Hirschsprung disease (HD) is characterized by the presence of an aganglionic bowel segment. This aganglionic segment results in stasis of intestinal contents due to the inability of the segment to contract. The main goal of the surgical management of Hirschsprung disease is to resect the aganglionic segment and pull down the normo-ganglionic bowel segment [1]. There are several described techniques in literature. The commonly utilized techniques around the world include Swenson, Duhamel, and Soave. Laparoscopy is increasingly being used for the definitive management of HD [2]. The

aim of our study is to assess the outcome of the laparoscopically managed pediatric patients with Hirschsprung disease at our center.

Methodology

The study design we employed was a retrospective single institute review. The study was conducted at the Department of Pediatric Surgery, All India Institute of Medical Sciences, Jodhpur. The study population included 28 patients of Hirschsprung disease who underwent surgery with the definitive pull through procedure performed laparoscopically. All patients undergoing exclusive transanal pull through, and laparotomy for pull through were excluded from the study. Data was collected from our center from June 2016 to March 2022.

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The collected data included age at the time of pull through surgery, gender, comorbidities, location of transition zone, the type of pull through procedure, duration of surgery, post-operative complications, length of hospital stay (LoHS), complications on follow up, and total number of stages of surgery. As a protocol, any patient with history and examination findings suggestive of HD is evaluated with a contrast enema performed under fluoroscopic guidance. The anteroposterior and lateral films during injection of contrast are used to identify the position of the transition zone (Fig. 1). The contrast study helps in the assessment of bowel caliber hence influencing our decision to create a colostomy or perform a primary pull through procedure. The diagnosis was confirmed by rectal biopsy before subjecting the patients to the definitive pull-through procedure. Twenty five out of 28 patients underwent a staged procedure while only three patients underwent primary pull-through.

Four patients presented to us with a colostomy done from an outside center. Twenty-one patients underwent colostomy at our center. Three out of 25 patients underwent levelling colostomy while remaining 22 patients were subjected to right transverse colostomy with multiple seromuscular colonic biopsies. Stoma biopsy was sent for histopathological analysis in every patient to confirm presence of ganglion cells.

All patients included in our study underwent laparoscopic assisted pull through procedure. Preoperatively, we advised distal stoma irrigation with preoperative parenteral antibiotic. Patients without stoma received total gut



Fig. 1 Contrast enema (lateral film) depicting transition zone and dilated proximal bowel



Fig. 2 Port positions for laparoscopic pull through surgery

irrigation with Peglec solution for 24 h and preoperative parenteral antibiotic. Out of 28, 26 underwent Swenson pull through. In lithotomy position, a supraumbilical camera port was inserted with right lower and left upper quadrant instrumentation ports (Fig. 2). After inspection and retrieval of frozen sections from multiple levels (when required) of bowel, we proceeded to mobilize the bowel. Mesocolon was divided to enable adequate mobilization after which the colonic loop was pulled down. After assessing if the loop was mobile enough to be brought down without tension we moved to the anorectal dissection. Rectum was circumferentially dissected and a coloanal anastomosis was done with the excision of the aganglionic segment. Laparoscopy was utilized to inspect our pulled down segment and confirm absence of torsion (Fig. 3).

First follow up was scheduled 3 weeks post-operatively. Bowel function was assessed using the Krickenberg classification in every patient over 3 years of age [3]. For the purpose of the study, any patient who was incontinent was unable to sense passage of stool in underwear. Soiling was graded into occasional soiling which is grade 1 (1–2 times per week), daily soiling which was grade 2, and constant soiling with social problems which was grade 3. Constipation was graded into grade 1 which was manageable by dietary modification, grade 2 was manageable by laxative, and grade 3 which was resistant to laxative and diet. Enterocolitis was defined as episode of increased frequency of loose stool associated with fever or vomiting or abdominal distension. Guidelines for Strengthening the Reporting of Observational Studies in Epidemiology (STROBE) were applied. [4]

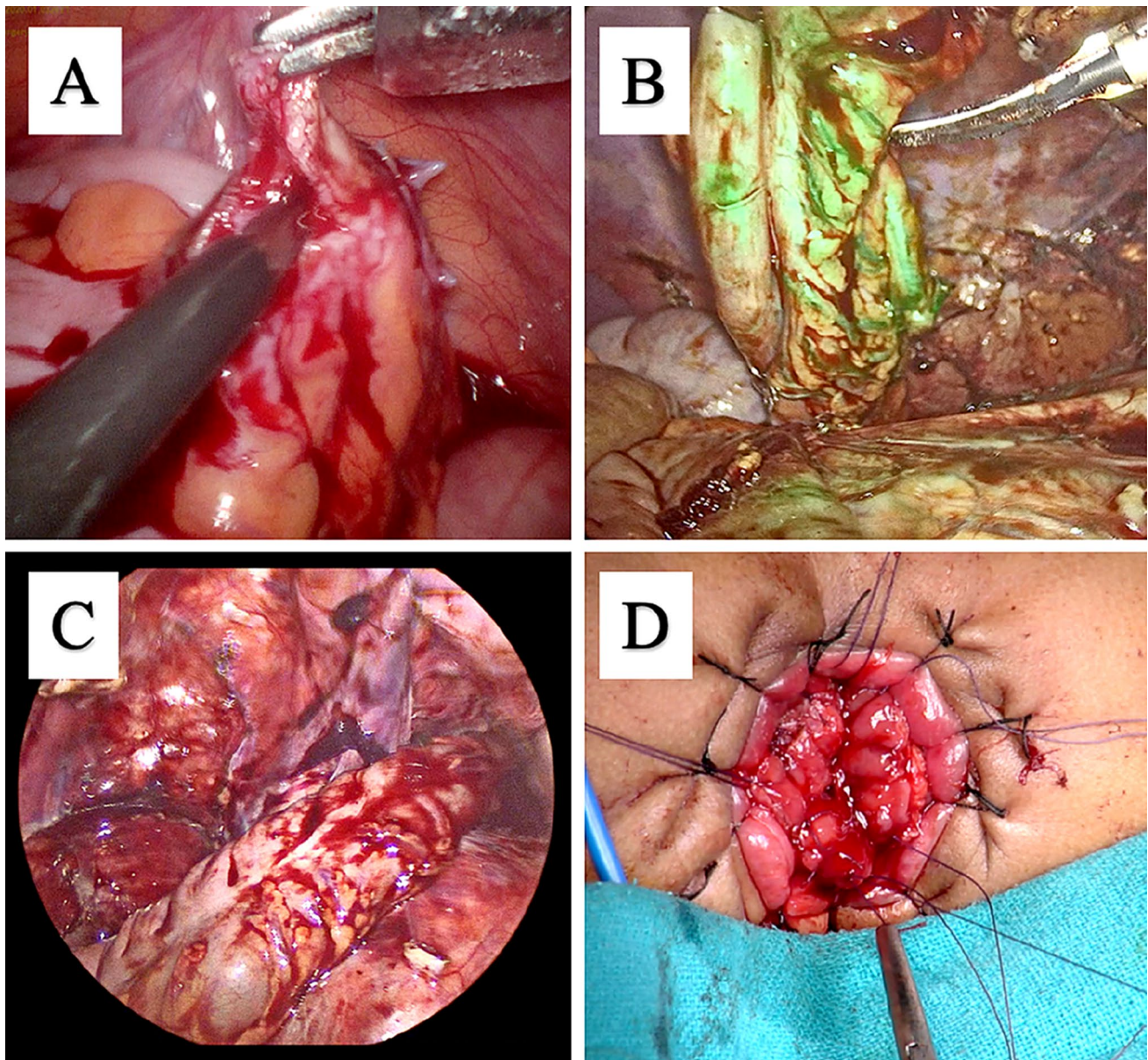


Fig. 3 **A** Seromuscular biopsy taken from multiple levels of bowel. **B** Vascularity confirmed using indocyanine green fluorescence. **C** Colonic loop mobilized and brought down without tension. **D** Coloanal anastomosis done after excision of the aganglionic segment

Results

The study included 24 male patients and four female patients. One patient had a large atrial septal defect. None of the other patients had any significant comorbidity. Twenty-five out of twenty-eight patients underwent staged procedure while only three patients underwent primary pull-through (Table 1). Median age at surgery was 24 months with a range of 3 months to 16 years.

Four patients presented to us with a colostomy done from an outside center. Twenty one patients underwent colostomy at our center. Median age of presentation at the time of colostomy was 26 months. Reasons for diversion colostomy included delayed age of presentation with hugely dilated colon necessitating a stoma for sufficient bowel emptying, poor general condition of patient (presented with shock due to sepsis), severe undernourishment, and enterocolitis. Most of the patients presented to us with significant abdominal distension after years of

Table 1 Clinical data of patients

Number of patients (<i>n</i>)	28
Male: Female	24:4
Comorbidities	1 (large atrial septal defect)
Median age at presentation (months)	24 months (range of 3–192 months)
Preoperative colostomy (<i>n</i>)	25
Position of transition zone on barium enema	13—sigmoid colon 12—rectosigmoid 3—not apparent

neglect of chronic constipation. They were also noted to be malnourished and hence not optimal candidates for a primary surgery. Three patients presented to us with features of Hirschsprung disease associated enterocolitis requiring an emergency diversion colostomy. Three out of 25 patients with stoma underwent levelling colostomy while remaining patients were subjected to right transverse colostomy with multiple seromuscular colonic biopsies. Right transverse colostomy was done without a routine fresh frozen specimen for ganglion cells. The frozen section biopsy facility is not available at emergency hours in our institute. Hence, the levelling colostomy is not feasible in those patients who require colostomy beyond the routine hours. In such cases, if the preoperative contrast enema is suggestive of short segment HD, we prefer to do the right transverse colostomy. This is based on the fact that the pathological, gross, and radiological transition zone may not always correlate. Poorly sited colostomy at transition zone based on intra-operative or contrast enema findings may prove disastrous in these late presenting severely undernourished patients. Right transverse colostomy in patients with radiological and gross recto-sigmoid transition zone ensures that the site of colostomy is far away from the transition zone. Multiple biopsies proximal to the gross transition zone helps us to identify the exact site of normal ganglionic bowel before the definitive pull-through. The flip side is that it mandates three-stage procedure. However, the protecting stoma in these patients negates the need of pre-operative whole gut preparation and post-operative total parenteral nutrition at the time of definitive pull-through. Thus, right transverse colostomy was essentially done in patients where the contrast enema or intraoperative findings were suggestive of short segment HD. These patients with right transverse colostomy underwent 3 stage procedure while three patients with levelling colostomy underwent two-stage procedure.

Thirteen patients had the transition zone on contrast enema located at sigmoid colon, in 12 patients it was noticed at the rectosigmoid, and in three of the patients the transition zone was not apparent (Table 1).

Out of 28 patients, 26 patients underwent Swenson, 1 underwent Duhamel, and 1 underwent Soave pull through. The type of procedure was at the discretion of the operating

surgeon. Three patients successfully underwent a primary pull through (single stage). None of our patients required conversion to open surgery. Median duration of surgery (patient in and out time including the waiting period for frozen section report) was 4 h with a range of 3 to 6 h. Median length of hospital stay was 6 days ranging from 3 to 24 days. One patient developed severe sepsis requiring a 21 course of intravenous antibiotics which led to a hospital stay of 24 days in total (Table 2).

Three patients developed postoperative complications. One patient developed severe sepsis requiring intravenous antibiotic course for 21 days, one patient had postoperative generalized tonic clonic seizure due to dys-electrolytemia which was managed accordingly and one patient developed urinary retention following catheter removal. The ultrasound in this child was suggestive of infective internal echoes in the bladder, and the patient was hence managed for urinary tract infection.

Every patient was followed up after three weeks in outpatient department. Patients undergoing staged Swenson's procedure were initiated on Hegar dilation. The dilatation was initially demonstrated to the parents at outpatient visit, and advised to be done twice daily with the adequate sized dilator for 6–8 weeks. Twelve patients with age more than 3 years were evaluated with Krickenbeck scoring for evaluation of bowel function (Table 3). At the time of follow up, 1 patient (8 year female) presented with night time soiling (grade 1 according to Krickenbeck scoring). This lasted 3–4 months post-operatively and resolved successfully after initiating bowel management program. A 4-year-old and 6-year-old male presented with constipation which was grade 2 according to the Krickenbeck scoring. Both patients were managed successfully using laxatives. As part of bowel management program, patient underwent serial abdominal radiography. Rectal washouts, laxatives, and enemas were administered as per the requirement. The goal was to keep the patient completely clean without soiling for 24 h a day. One patient presented with post-operative enterocolitis. This was a 3-year-old male whose initial presentation to our center was also with enterocolitis for which he required a colostomy. Six months after undergoing colostomy closure, he presented with similar features of abdominal distension, fever and loose foul smelling stools and was managed by

Table 2 Surgical data

Type of pull through procedure (<i>n</i>)	Swenson—26 Duhamel—1 Soave—1
Median duration of surgery (hours)	4 h (Range of 3–6 h)
Median length of hospital stay (days)	6 days (Range of 3–24 days)
Immediate post operative complications (<i>n</i>)	3
Number of stages of surgery (<i>n</i>)	Single stage—3 Two stage—3 Three stage—22

Table 3 Krickenbeck questionnaire for bowel function in patients above 3 years of age

Voluntary bowel movement (<i>n</i>)	
Yes	12 patients
No	0 patient
Soiling (<i>n</i>)	
Grade 1—occasional	1 patient with grade 1 soiling
Grade 2—daily with no social problem	
Grade 3—daily with social problem	
Constipation (<i>n</i>)	
Grade 1—manageable with diet	2 patients with grade 2 constipation
Grade 2—manageable with laxatives	
Grade 3—resistant to diet and laxatives	

rectal washouts and intravenous antibiotics. Median follow up duration was 24 months.

Discussion

The principle of surgical management of Hirschsprung disease is to remove the defunct aganglionic segment and anastomose the normally innervated bowel just above dentate line. Swenson initially described resection of the aganglionic segment followed by primary anastomosis just above the anal sphincter. Two other operations were subsequently described to avoid the potential risk of nerve and vessel damage during Swenson's procedure. The Soave's procedure involved submucosal dissection with pull through with a remnant aganglionic muscular cuff. The Duhamel procedure involved bringing the bowel through an avascular retrorectal space and staple anastomosis to native aganglionic rectum.

No prospective study has so far definitively established the superiority of one procedure over the other. [5]

Laparoscopy in pull through procedure for Hirschsprung disease was first described by Georgeson et al. (1995) and since then it has been widely adopted in practice. The use of laparoscopy enhanced visibility and safer dissection was possible [6]. The initial open methods transitioned to a more minimally invasive approach with time after the advantages were proven. On comparison with another preferred technique that is the transanal pull through, there are several notable advantages of laparoscopy-assisted pull through such as the ease of obtaining multiple seromuscular biopsies, mobilization of bowel up to long distance under vision, serial sampling of any portion of bowel under vision without having to wait for frozen section report, reduced risk of perineal injury from excessive traction, and identification of torsion of pulled down segment [7]. Laparoscopy also enables intraoperative usage of indocyanine green based fluorescence to verify the vascularity of bowel segment [8]. We included the use of indocyanine green in several of our laparoscopic pull through surgeries. Indocyanine green is utilized for its ability to become fluorescent when excited by near-infrared light. It is intravenously injected and a real-time angiography is used to evaluate and confirm the bowel perfusion prior and after colorectal anastomosis [9].

In general, laparoscopy is associated with less time to initiation of feeds and less chance of development of future adhesions on comparison to open surgery [10]. Kubota et al. reported the outcome of surgery on comparison of laparoscopic and open pull through in 41 patients and stressed that laparoscopy was less invasive and offered better functional outcome in terms of incontinence [11]. An exclusive transanal approach is said to be the least invasive procedure but there are concerns regarding significant anal sphincter damage due to excessive traction. Presence of a more proximal transition zone necessitates colonic mobilization which cannot be done exclusively through perineal approach. Karlsen et al. compared the difference in functional outcome between an exclusively transanal and laparoscopy-assisted surgery in 91 patients. Their study demonstrated no significant difference in long term bowel function between both groups. The complication rate in both groups was around 10% which is in line with previous publications. They concluded that there was no special advantage of an exclusive transanal approach over laparoscopy and that laparoscopy was advantageous in providing appropriate visibility for safe extensive dissection [12].

A meta-analysis by Tomuschat et al. evaluated 16 studies with a total of 820 patients undergoing laparoscopic pull through for Hirschsprung disease. Nearly 1/3rd of the patients continued to have long term bowel problems but there was discrepancy in the scoring used to assess bowel function. A variable follow up duration also evoked an

amount of bias. The advantages of laparoscopy however were concluded with demonstration of increased safety and ease of mobilization [13]. A systematic review and meta-analysis published by Thomson et al. presented evidence from 5 retrospective studies of 405 patients. They compared transanal pull through with laparoscopy-assisted pull through and concluded that there was no significant difference in incidence of post operative incontinence, enterocolitis, and constipation. The only significant finding was shorter operative time with trans anal owing to avoidance of abdominal access [14].

The disadvantages we faced during our study included a wide range of age of presentation of most of our patients. The late presentation meant requirement of an initial decompressive colostomy and also meant that we were unable to offer the option of a primary pull through to most of our patients since they mostly presented at a later age with massively loaded and dilated bowel loops not amenable for single stage surgery. Unavailability of frozen section facility at emergency hours also compelled us to perform right transverse colostomy in most of the patients with radiological and intraoperative findings suggestive of short segment HD. Levelling colostomy could be performed in only three patients.

On comparison with previous studies, none of our patients developed stricture on follow up. This is in contrast to previous literature which show that development of stricture is a known complication of pull through procedure. Oblique, tension free coloanal anastomosis, routine anal calibration following Swensons's procedure, and confirmation of vascularity using ICG in case of any doubt on the viability of mobilized bowel were the probable contributing factors. All patients in our study responded to bowel management program and have successfully remained continent on follow up. Conway et al. suggested that post operative bowel function in operated patients of Hirschsprung's is expected to improve with time [15]. Granstrom et al. evaluated functional outcome after laparoscopic pull through surgery in 35 patients and it was seen that the incidence of constipation postoperatively reduced over time [16].

Our study emphasizes on the need for regular outpatient visits and the need for adherence to procedures such as rectal washouts and Hegar dilation. Timely follow up can help obtaining optimal functional outcome. It is notable that none of our patients developed stricture over the period of follow up. This may be attributed to the mandatory dilation program we advise to every patient at follow up. Every patient has been examined at follow up including per rectal evaluation of caliber. Accordingly the size of the dilator is modified.

A larger study population with longer follow up however, is definitely required to definitively establish the effectiveness of laparoscopy assistance in pull through procedures. Comparison of laparoscopy to open and transanal procedure

in our institute may help us compare and conclude better regarding the advantages. Other factors such as conditions conferring unfavorable outcome like Down's syndrome were not seen in our study. We expect patients of such conditions to be increasingly prone to develop post operative complications compared to those without such comorbidities.

Conclusion

Laparoscopy-assisted pull through for HD is both safe and feasible in pediatric patients. Delayed presentation with dilated colon, poor nutritional status, and general condition may compel us to opt for a staged procedure. Excellent short term results with satisfactory functional outcome can be achieved in most of these patients even in developing countries where delayed presentation is almost a norm.

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Declarations

Conflict of interest The authors declare that they have no conflict of interest.

Human participants or animals This article does not contain any studies with human participants or animals performed by any of the authors.

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