

One Stage Transanal Full Thickness Pull-Through for Hirschsprung's Disease: A Report on Six Years Operation Experience with Iraqi Infants and Children

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ABSTRACT

Background: In the past two decades, the traditional surgical techniques for Hirschsprung's disease (HD) have evolved from numerous stages with stoma formation to a more modern one-step surgery with low invasiveness as well as low patient morbidity. The transanal full-thickness rectal resection procedures and case studies are presented in this study. All surgeries were performed at the Al-Kafeel Super Speciality Hospital in Kerbala, Iraq.

Methods: This retrospective study was done in a single unit, from March 2016 to December 2022. Prior to surgery, contrast edema was performed on 100 patients who met the inclusion criteria, such as uncomplicated and short-segment. All patients who met the inclusion criteria were included in this study's clinical, surgical, and follow-up phases; individuals with colostomies, ileostomies, or any other abdominal interference were not included.

Results: The average age of the 100 patients at the time of surgery was 4 months (120 days). At the end of the neonatal phase (first month), three patients received surgery. Ninety patients had their transition zone identified using a contrast enema study, surgical dissection, and rectosigmoid excision. Two patients needed red blood cell transfusions, and the average amount of blood loss was 25 mL. After 24 hours, the average full feeding time occurred. The average hospital-stay before and after surgery, was 3 days. NSAIDs were used to provide analgesia to all patients. Early postoperative complications included stricture (4%), enterocolitis (5%), perianal excoriation (20%), and spasm of the anal sphincter muscle (10%). The patients were monitored for roughly 36 months; 90% of them had age-appropriate normal bowel function, 6% had minor issues, and 4% had serious issues. For anastomotic leak in two patients, severe stricture in two patients who did not respond to anal dilatation in two patients and residual aganglionosis in one patient, a second operation was necessary.

Conclusion: One stage Transanal full-thickness pull-through can be a safe treatment if executed correctly and it promises a successful outcome, especially for patients with short ganglionic segments suffering from HD. Early feeding, early release, no transabdominal investigation, decreased morbidity, and happiness for the family and the baby in the future are all made possible by this procedure.

Keywords: Hirschsprung's Disease; Single stage surgery; Transanal pull through.

INTRODUCTION

Hirschsprung's disease (HD) is histologically characterized by aganglionosis, or the absence of ganglion cells (between 70 to 80%), primarily in the rectal wall of the large bowel (colon). This causes a dynamic intestinal obstruction that causes abdominal distension and delayed or failed meconium passage in the first 48 hours of life, and in severe or long segments (total aganglionosis), a cecal or small bowel perforation may occur [1]. With an incidence rate of 1 in 5000 live births, HD is the most prevalent reason for intestinal blockage in neonates [2]. The usual course of treatment is a colostomy followed by a transabdomino-perineal pullthrough. Functional blockage can be successfully treated without a colostomy during the newborn period using laxatives or a rectal tube and saline irrigation [3].

Ileostomy or proximal de-functioning colostomy followed by a pull-through technique, and finally the closure of the colostomy were the main components of traditional surgical treatment for HD [4]. The colon's aganglionic portion is removed, and the regular innervated bowel is then pushed down and anastomosed to the area above

the dentate line using this approach, which could be completed in two or more steps [5]. The Swenson, Duhamel, and Soave (or endorectal pullthrough) procedures are the most often used pull-through procedures, despite the fact that numerous different kinds have been identified. Given that numerous studies have proven that a one-stage pullthrough is both safe and effective, a growing number of pediatric surgeons have switched from routinely using a colostomy to this method during the past few decades [6-8].

Since Dr. Ovar Swenson's original illustration of the etiology of HD, it has been more than 70 years. Children who had HD typically underwent an initial colostomy and suffered from persistent enterocolitis, constipation, thrive failure, abdominal distension, and dependency on assisted defecation [9, 10]. Following that, a pull-through procedure was performed, and the colostomy was then closed [4]. Numerous changes to the typical one-stage operations have been made as a result of the popularity of minimal-access surgical techniques in recent years. In contrast to traditional open operations, Georgeson proposed a laparoscopic approach which has been utilized by numerous surgeons and is associated with less pain and a shorter hospital stay [11]. Following this, several studies proposed a transanal-only method, which seems to have the same benefits without requiring any intraabdominal dissection [12, 13].

Endorectal pull-through is the most popular transanal technique, leaving a lengthy muscular cuff that is typically split longitudinally either anteriorly or posteriorly [12]. It is generally recognized that the lengthy muscular cuff left behind causes enterocolitis and obstructive symptoms. A few investigations in the literature have demonstrated that if the transanal resection of the aganglionic segment is carried out in the way specified by Swenson, that is, by dissecting the entire thickness of the rectum the issue of the remaining rectal cuff can be avoided [13, 14].

This report demonstrates the experience with transanal, full thickness Swenson-like dissection for rectosigmoid HD over a 6-year period. It does this by dissecting the anorectal malformation's full thickness rectal wall while utilizing the transanal method [15, 16]. The Swenson transabdominal approach became less popular due to wide dissection made by some of the surgeons which cause many complications [17]. However, this method feels secure, reproducible, and retains sexual, fecal, and urine functions, when executed with an elegant technique using the appropriate plane. The aganglionic gut can be completely removed using this operation without leaving a muscular cuff or a pouch, making it the preferable option [18].

METHODS

The study prospectively recorded paediatric patients who had one-stage transanal full thickness pull-through procedure for the repair of HD between March 2016 and December 2022 at the Al-Kafeel Super Speciality Hospital in Kerbala, Iraq.

Gestational age at birth, sex, birth weight, age at diagnosis, level of aganglionosis, clinical presentation, preoperative management, surgical technique, intraoperative complications, time to full oral feeding, late postoperative complications, and follow-up data, such as complications, ongoing oral gastrointestinal medication needs, and bowel motility, were among the data collected.

Each patient received essentially the same surgical method. The patients required time to flush the excrement out of their colons. The patients were offered a low-fibre diet two days before to the procedures. For two days before to the procedure, erythromycin and neomycin (30 to 50 mg/kg/d) were administered to suppress colonic bacteria. The night before surgery, gentamycin and metronidazole or cefoxitin were administered, and they continued for 5 days after the surgeries. At the time of the surgeries, frozen section biopsies were done. Before we anastomosed the colon to the distal anorectal mucosa, it had to have ganglion cells.

The one-stage transanal endorectal pull-through procedure used were identical to previously described methods by Langer et al. [19] and Albenese et al. [20]. To encourage hemostasis and make mucosal dissection simple, 0.25% lidocaine and 1:200,000 epinephrine were administered around the submucosal area. Prone and lithotomy positions, representing 60% and 40%, respectively, were used. Using 5/0 and 4/0 Vicryl suture, fix stay sutures circularly (circumference of 1.5 cm) above the dentate line. Using unipolar electrodiathermy, dissection began around 0.5 cm above the suture stay line. The colon was dissected by circumferentially cutting the rectal wall to its full thickness before utilizing diathermy and ligatures to effectively achieve good hemostasis in the colon's proximal regions. Identification of the transition zone was done before a safe margin resection (dissection about 10 cm above the transition zone) was performed (Figure 1-6).

An experienced histopathologist at the facility performed a frozen section biopsy on each patient from the safe zone above the transition zone, which revealed an appropriate amount of ganglion cells (Figure 7). Without strain or redundant effort, a coloanal anastomosis was performed from the location of the frozen segment. The results of the extensive histological examination of the permanently removed segments, which revealed aganglionosis in the distal constricted segment and an appropriate amount of ganglion cells, confirmed the diagnosis of HD.

RESULTS

Out of the 100 patients included in the study, 78 were males and 22 were females. The age at diagnosis was ≤ 60 days in 85 children (85%), 2 month to 3 years in 15 (15%). Clinical symptoms included delayed meconium passage in 85 (85%) cases, abdominal distension in 95 (95%) cases, and enterocolitis in 32 (32%); some children presented with more than one symptom. All 100 patients underwent a contrast enema study; in 95 (95%) of the cases, the radiologic transition zone (rTZ) was in the rectosigmoid region, and in only 5 (5%), they failed to show a transition zone. Table 1 summarizes the features of the patients and their clinical presentation.

Preoperative bowel preparation included digital rectal stimulations (90%), bowel irrigations (64%), or a combination of both (52%) for all 100 children. Breast milk (36%), elemental meals (17%), or normal formula (47%) were used to maintain nutrition.

The operation time ranged from 40 to 240 minutes, with an average of 120 minutes. The average intraoperative blood loss during surgery was calculated to be 50 mL, and two patients needed blood transfusions.

Before beginning the anal dissection, the pathologic transition zone was found in 97 instances. The transition zone was located in the splenic flexure in 3 cases; this was confirmed by laparoscopic assistance. In 81% of the children, the pathologic transition zone was restricted to the rectosigmoid; in the remaining 24%, it was located in the more proximal colon. In 6 cases, there was a difference between the radiologic and pathologic transition zones; the pathologic transition zone was closer to the patient. A minor infraumbilical incision was required to mobilize the colon and perform the coloanal anastomosis precisely in these cases. None of the patients needed to have a colostomy formed.

Two cases of intraoperative complications were identified; one involved the anastomosed segment of the colon prolapsing through the anus, and the other involved bowel and laparotomy wound dehiscence. Both cases were taken back to the operating room, where the first patient had the redundant segment excised and the coloanal re-anastomosis performed. For the second patient, tension suturing was used to reconstruct the abdominal wall, and the stitches were removed two weeks later. In both of my experiences, the postoperative period went without a hitch.

The average time required postoperatively for oral feeding was 6 to 72 hours. Hegar anal dilators were used to start the rectal dilatation regimen with all patients. Following the procedure, dilating began and lasted for 16 days to 4 months. The average follow-up period for 88 kids was 18.4 ± 8.4 months. 85 (85%) of these children's parents stated that their children had normal bowel function, 21 had ongoing minor issues like loose stools, mild-to-moderate constipation, or perianal excoriation, and 2 had ongoing substantial issues like persistent severe constipation in one kid and severe vomiting in another child. Eleven patients were using regular medications for their gastrointestinal issues, such as antibiotics, enemas, or stool softeners.

Additional surgery was required for three patients, two of whom experienced obstructive symptoms (severe stricture at the anastomosis site). Colostomy management followed by a second pull-through was used in both cases. The other child needed a colostomy diverting from the transverse colon as well as a redo transanal pull-through helped by a minor infra umbilical mid-line incision. This child also had residual aganglionic segment. The fourth patient underwent adhesiolysis due to intestinal blockage.

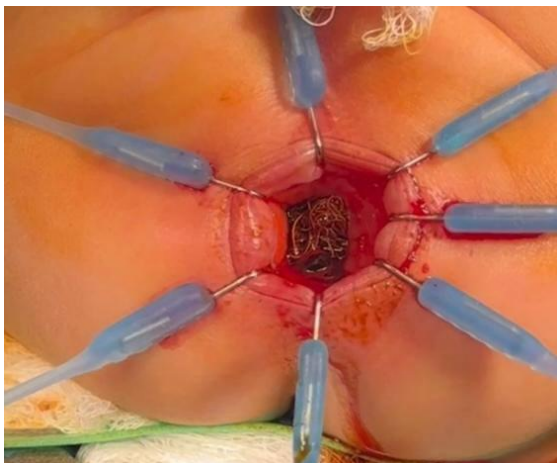


Fig.1. Location of lone star retractor at the anocutaneous junction taking the dentate line within.

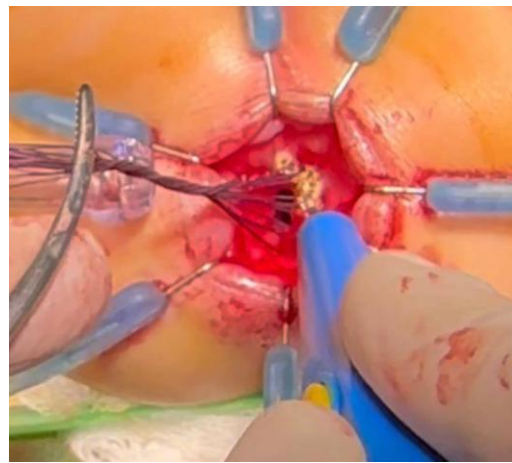


Fig.2. Starting full thickness dissection and separation of rectal wall.

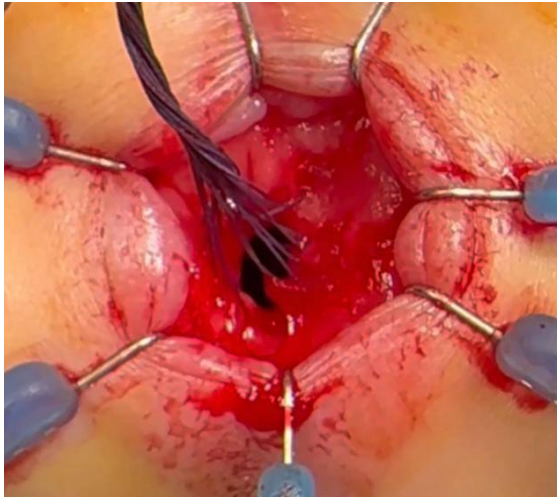


Fig.3. Circumferential rectal mucosal staying sutures (4/0 vicryl), 1.5 cm above dentate line.

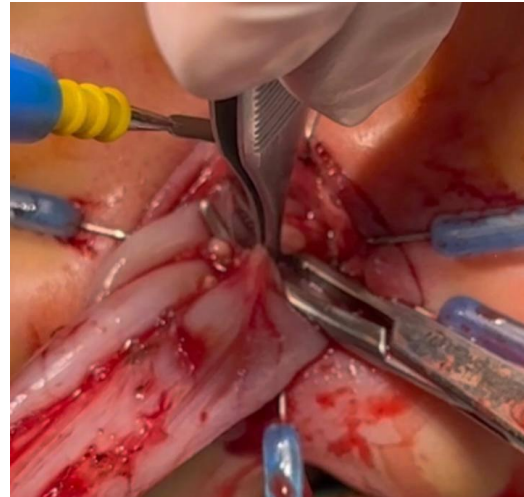


Fig. 4. Isolation of feeding mesenteric vessels.



Fig.5. Taking full thickness colonic biopsy for frozen section at 8 cm above transition zone.

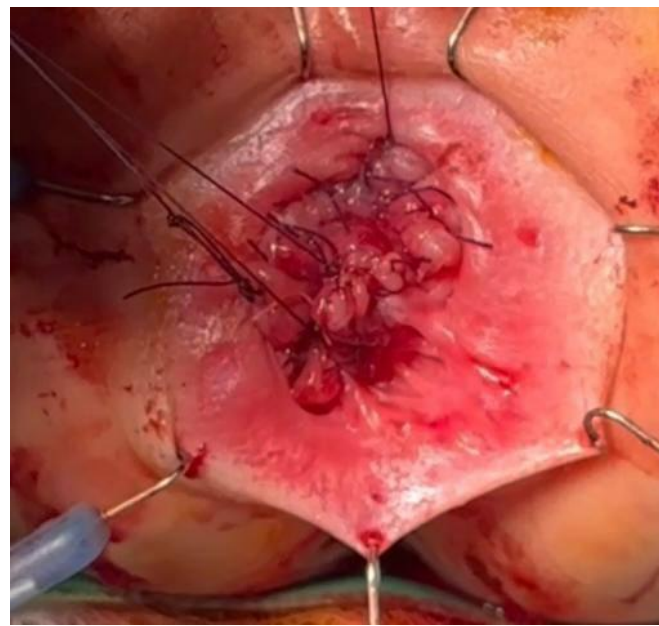


Fig.6. Coloanal anastomosis with tension free, good blood supply and preservation of anal canal.

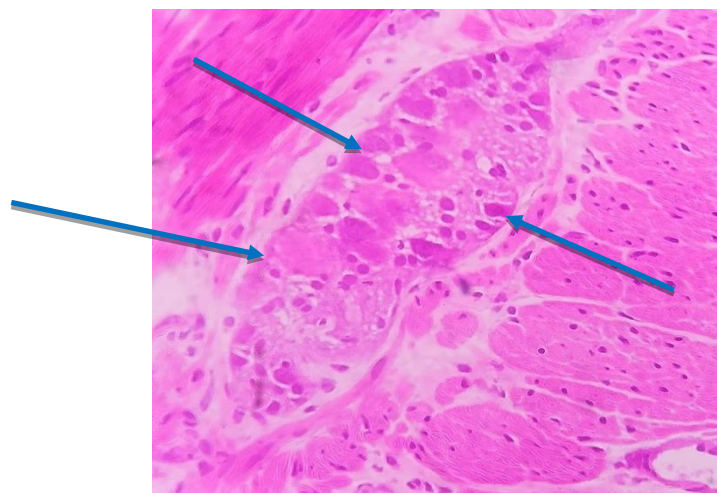


Figure 7: Histopathological analysis of frozen section biopsy above transition zone

Each patient underwent a frozen section biopsy from the safe zone above the transition zone. Blue arrows indicate that a sufficient number of ganglion cells are present. Frozen section for colonic tissue with adequate ganglion cells in Myenteric nerve plexuses (arrow heads), H&E stain, X400.

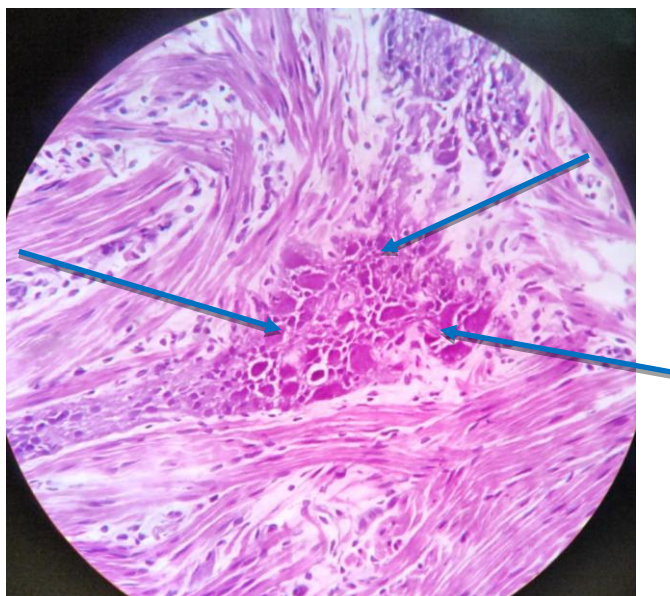


Figure 8. Adequate no. of ganglion cells in myenteric nerve plexuses among muscularis bundles, H&E stain. Blue arrows indicate that adequate no. of ganglion cells in myenteric nerve plexuses. H&E stain, X400.

DISCUSSION

HD is treated by resecting the aganglionic portion of the rectum and colon, removing normally innervated bowel, and anastomosing this bowel at the anorectal area, all while maintaining the sphincter muscle [21]. Previously, individuals with a late manifestation of HD underwent a one-stage transabdominoperineal pull-through operation. Recently, however, it was discovered that the one-stage method was technically possible and had low morbidity in neonates [22]. Transabdominal exploration in conjunction with the perineum route was previously used to carry out every typical surgical technique mentioned by Duhamel, Soave-Boley, and Swenson. Later, a successful minimally invasive HD treatment method via laparoscopic dissection was revealed [23].

Recently, one-stage transanal endorectal pull-through surgery was performed during the neonatal period by Zhang et al., using Albanese et al.'s novel method [20, 24]. Additionally, it can take away the chance of pelvic nerve damage. Aesthetic results were superior to those achieved with the transabdominal technique, and postoperative pain was barely noticeable [24].

The basic transanal pull-through techniques, with numerous modifications, have been adopted as the industry standard of care worldwide. One stage transanal procedures have a number of benefits, including avoiding multiple laparotomies and preliminary colostomies and the complications they can cause, as well as shorter hospital stays, less blood loss, no damage to the pelvic structure, no external scars, and improved cosmetic results without sacrificing functional results [25]. Most importantly, because it eliminates the need to care for an infant with a colostomy, there is a high level of parental acceptance of the treatment. The volume of the replacement sigmoid and rectum is nevertheless constrained by the aganglionic rectal muscle cuff in the transanal Soave's technique. Cuff strictures, blockage, enterocolitis, and incontinence are additional effects of this residual rectal cuff [26]. Numerous studies have suggested separating the muscle cuff posteriorly up to the sphincter to avoid these difficulties and prevent restriction of the lumen via the gut. During gut pull-through, the cuff can be rolled down and may create a ring-like constriction right before the colo-anal anastomosis [27]. Numerous studies performed shorter mucosectomy with a shorter muscle cuff about 1-2 cm above the dentate line to avoid this issue [28, 29]. Some studies describe a transanal Swenson-like surgery without preserving any remnant muscle cuff, fully avoiding the submucosal dissection [27, 30].

Transanal complete thickness pull-through in this study revealed that transanal rectal dissection was simple in infants but challenging in older children. Older patients' thickened mesentery, inflammatory mucosa, chronically dilated colons, and prior rectal biopsies were among the issues. One older child's posterior myomectomy resulted in adhesions, challenging mucosal dissection, and recto-sigmoid colon mobilization.

Some of the patients who had postoperative enterocolitis may have had chronic intestinal inflammation and blockage. Rectal tube decompression and antibiotics were used to treat it. If the patient's condition did not improve with conservative therapy, a colostomy must be performed.

Given that enterocolitis is one of the most dreaded postoperative complications that might happen after surgery for HD. Due to its severity, its administration employed an aggressive strategy. Although there is no definitive definition of enterocolitis, treatment began when any postoperative patients experienced abdominal distention, fever, stomach pain, and/or foul-smelling loose stools. This was followed by rectal irrigations and metronidazole. If necessary, parents conduct rectal dilatation. Following the acute episode, weaning is done for a period of 3 to 4 weeks before returning to irrigation and metronidazole. This strong strategy, in my opinion, prevents many hospital admissions. Thus, the overall rate of enterocolitis in our study is close to 20%, which is comparable to data after a Swenson treatment [17] and the 18–20% stated rate after Duhamel or Soave surgery [6, 8].

While the one step transanal fullthickness pullthrough largely follows the same technique as HD operations, it does so without requiring a laparotomy or intraabdominal rectum mobilization. Because of this, it ought not to be linked, theoretically, to a higher likelihood of complications or failure. There shouldn't be any intra-abdominal dissection, this could speed up feeding and discharge times and maybe reduce the risk of intraabdominal adhesion formation. Additionally, there shouldn't be any significant abdominal incisions, and this should improve the cosmetic outcome. Minimal discomfort, quick feeding and elimination times, and an abdomen with few scars or none at all are further benefits. This was observed whether or not a laparoscopic or periumbilical incision was made. The transanal method, which does not require intraabdominal rectum mobilization and may be carried out by any experienced pediatric surgeon, even one who is not proficient in laparoscopic surgery, may be superior than the laparoscopic pullthrough described by Georgeson [11, 31].

In conclusion, the transanal Swenson fullthickness operation, according to data of this study, offers a better treatment option for children with Hirschsprung disease and can be performed by any skilled pediatric surgeon. Older children may get early postoperative enterocolitis, which requires immediate treatment. A rectal tube inserted into the anus to relieve pressure on the dilated pull-through colon could be used to treat the partial coloanal anastomosis blockage seen in the older children.

Further studies documenting the long term results of this approach, particularly with respect to continence and stool frequency, will be needed as these children grow and develop.

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Conflict of Interest: None

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