# One Stage Transanal Endorectal Pull-through for Treatment of Hirschsprung's Disease: 5 years experience

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**ABSTRACT**

**Background/purpose:** Approximately 1 in 5000 newborns is born with Hirschsprung's disease (HD). The surgical management of HD has progressed from multi-stage procedure to primary transanal operation. This study aims to evaluate the safety, feasibility and the outcome of one stage transanal endorectal pull-through technique (TEPT) for treatment of HD.

**Patients and methods:** From January 2005 to January 2010, fifty seven children with rectosigmoid HD were included in this study. All underwent one stage TEPT procedure. Operative data, postoperative complications and outcome, all were reported.

**Results:** Of 57 patients, 39 were males and 18 were females. All were diagnosed as HD via their clinical features, contrast enema study and full thickness rectal biopsy. The age at the time of diagnosis ranged from 2 days to 30 month, the age at time of surgery ranged from 3 months to 30 month and their weight ranged from 4Kg to 14 Kg. the mean operative time was 80±15 minutes and hospital stay ranged from 2.5 to 5 days. Postoperative complications included perianal excoriation, dermatitis, recurrent enterocolitis and soiling Follow up periods ranged from 4 months to 24 month.

**Conclusion**: One stage TEPT procedure is a safe, feasible with satisfactory long term outcome.

**Key words:** Hirschsprung's disease, one stage transanal endorectal pull-through technique.

## Introduction:

Hirschsprung's disease occurs in one out of 5,000 births (Amiel. and Lyonnet ; 2001). The disease is caused by failure of ganglion cells to migrate cephalocaudally through the neural crest during period from $4^{th}$. to $12^{th}$ week of gestation (Parisi. and Kapur ;2000) .

HD most commonly involves the rectosigmoid reigon of the colon, but can affect the entire colon. Rarely, the small and large intestines are involved (Stewart and von Allmen ;2003).

The symptoms vary with the age of the patient and the extent of the disease. Symptoms range from neonatal intestinal obstruction to chronic progressive constipation in older children (Holschneider. and Puri;2000/ Coran and Teitelbaum ;2000).

A transitional zone is the point where the normal bowel becomes aganglionic and it may be visualized on contrast enema study; however, the aganglionic colon may extend beyond this point in about 10% of patients. The diagnosis of HD should be confirmed with rectal biopsy (Proctor; et al;2003).

Traditionally, surgical therapy for HD has consisted of a proximal defunctioning colostomy, followed months later by a definitive reconstructive "pullthrough" procedure and lastly closure of colostomy. In the past several decades, increasing numbers of pediatric surgeons have abandoned the routine use of a colostomy in favor of a 1-stage pullthrough, with multiple studies suggesting that this approach is safe and efficacious (Langer. et al; 1996, Teitelbaum et al; 2000 and Pierro. et al; 1997).

This report describes our 5 years experience with a 1-stage transanal pull-through operation in 57 patients with rectosigmoid HD and to evaluate their surgical outcomes.

## Patients and methods:

This study was carried out in pediatric surgery unit at Sohag University Hospitals from January 2005 to January 2010. Fifty seven patients included in this study; 39 were boys and 18 were girls. All were diagnosed as HD via their clinical features, contrast enema study and full thickness rectal biopsy. All patients had an aganglionic segment confined to the rectosigmoid area which was confirmed by the preoperative barium enema and postoperative histopathological examination.

**Technique: (Photo 1-6)**

HD suspected patients were proved by rectal biopsy at 1 cm above the dentate line (All of the rectal biopsies are full thickness because of the lack of facility and experience with submucosal biopsy in our centre). The patient with long segment aganglionic colon seen in barium enema was excluded. All patients underwent saline colonic irrigation and full bowel preoperative preparation at the day before operation. Wide spectrum antibiotic was used with the induction of anesthesia. Foley urinary catheter was routinely used. After general anesthesia, colonic irrigation with warm saline and povidone-iodine, the patients were placed in prone position.

The mucosa was incised circumferentially 1 cm above the dentate line and a submucosal dissection was carried till reaching the peritoneal reflection proximally and the muscular rectal cuff was divided before performing the anastomosis. The colon pulled down, the affected bowel was resected and colo-anal anastomosis was performed by interrupted 4/0 absorbable suture. No drain was used.

Three weeks after surgery, gentle anal calibration was done and dilatation was performed when required with a Hegar dilator for 3–6 months. Clinical outcome was assessed by outpatient interviews and questionnaires. The following data were collected: sex, gestational age at birth, age at diagnosis, clinical presentation, operative technique, intraoperative complications, time to full oral feeding, early (within 30 days) and late postoperative complications and follow-up data, including complications, and effectiveness of potty training(state of continence).

## Results:

This study included 57 children. There were 39 (68.4%) boys and 18 (31.6%) girls. A male to female ratio of 2.2:1 was observed. None of them had significant associated abnormalities. At birth 51 (89.5%) of them were full-term.

The mean age at the time of diagnosis was 3.7 month (ranged from 2 days to 30 month); the mean age at time of surgery was 4.9 month (ranged from 3 months to 30 month) and weight ranged from 4Kg to 14 Kg with the mean 6.2 Kg.

Clinical presentation varied from neonatal intestinal obstruction to chronic progressive constipation in older children and some of them had more than one presenting symptom (table 1).

Difficult bowel movement and progressive abdominal distention presented in 44 cases (77.2%), picture of neonatal bowel obstruction occurred in 17 cases (29.8%), history of failure to pass meconium in the first 24 hours of life was found in 51 cases (89.5%) and HD associated enterocolitis (HAEC)-related diarrhea presented in 11 cases (19.3%).

No major anesthetic complications were reported. The mean operating time was 80 ± 15 minutes (range from 75 to150 minutes). One patient required blood transfusion during surgery. The mean length of aganglionic resected segment was 15 cm (range from10 to 45 cm).
Mean time to full oral feeding was 32 hours (range from 24 to 72 hours), and mean length of postoperative hospital stay was 3.2 days (range from 2.5 to 5 days).
During the first 30 days after operation, early postoperative complications occurred in 5 patients (8.8%), which included enterocolitis in 1 patient (1.8%) and transient perianal excoriation occurred in 4 cases (7%), during the first month postoperatively, due to increased frequency of defecation and soiling (5 to 10 times every day) that was improved with time (table 2).
Follow up was carried out in outpatient clinic visits and ranged from 4 months to 24 month (mean 10 month).
Late postoperative complications (after 1 month) occurred in 16 patients (28%) and included HAEC in 4 patients (7%) that was successfully managed by medical treatment in the form of 3$^{rd}$. generation cephalosporin, metronidazol and colonic irrigation with saline, perianal excoriation persisted in 1 patient (1.8%), and anastomotic narrowing developed in 1 case (1.8%) that was successfully managed with serial dilations at the outpatient clinic (table 3). No patients developed postoperative anastomotic leak and there was no postoperative mortality.
The stooling patterns and state of continence were considered satisfactory in 47 patients (82%), with the mean stool times were 1 to 3 per day. The remaining 10 children (18%) developed abnormal stooling pattern in the form of; 2 patients (3.5%) with stool frequency (5 to 8 times per day), postoperative soiling was found in 2 children (3.5%), constipation was

observed in 5 cases (8.8%) and there was 1 patient (1.8%) with stool incontinence.

Four patients with soiling and increased frequency showed a steady improvement over the first year. Complete anorectal continence was confirmed in 54 patients (95%) at ten month follow up.

**Table 1: Clinical presentation**

| ***Clinical finding*** | ***Number*** | ***Percentage*** |
|---|---|---|
| Failure to pass meconium in the first 24 hours of life | 51 | 89.5% |
| Chronic progressive constipation | 44 | 77.2% |
| Progressive abdominal distension | 44 | 77.2% |
| Neonatal intestinal obstruction | 17 | 29.8% |
| HAEC | 11 | 19.3% |

**Table 2: Early postoperative complications within the first month:**

| ***Complication*** | ***Number*** | ***Percentage*** |
|---|---|---|
| Perianal excoriation | 4 | 7% |
| HAEC | 1 | 1.8% |
| ***Total*** | 5 | 8.8% |

**Table 3: Late postoperative complications after first month:**

| ***Complication*** | ***Number*** | ***Percentage*** |
|---|---|---|
| Constipation | 5 | 8.8% |
| Soiling | 2 | 3.5% |
| Increased stool frequency (5-8 time/day) | 2 | 3.5% |
| HAEC | 4 | 7% |
| Persistent perianal excoriation | 1 | 1.8% |
| Stool incontinence | 1 | 1.8% |
| Anastomotic narrowing | 1 | 1.8% |
| ***Total*** | ***16*** | ***28%*** |

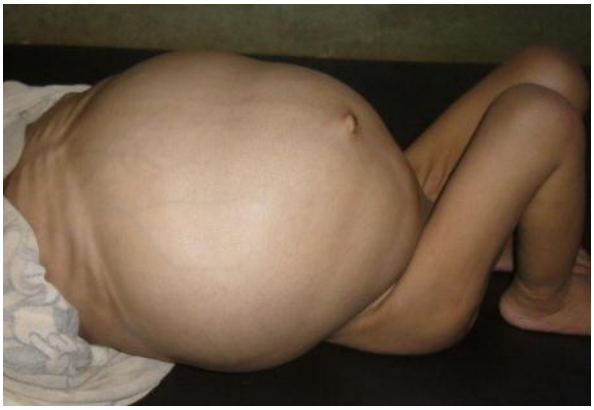

Photo1: HD with marked abdominal distension

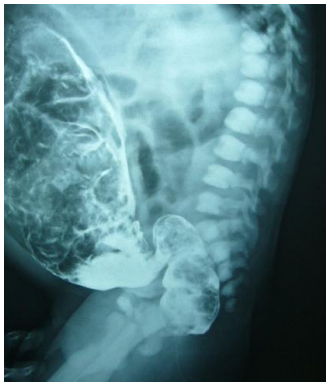

Photo2: Contrast enema

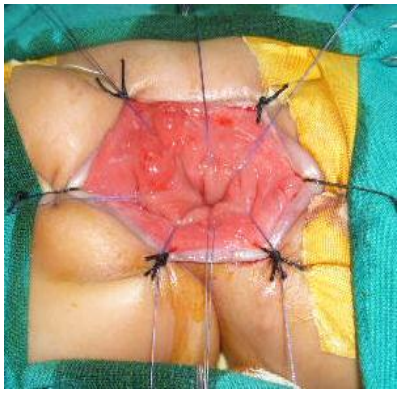

Photo3: Traction sutures

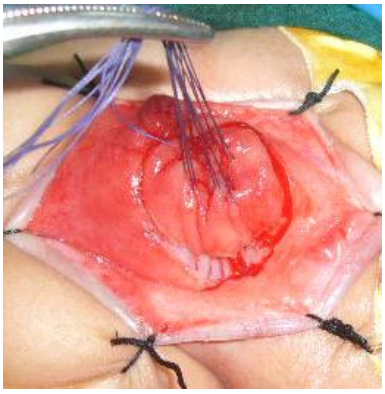

Photo4: Submucosal dissection

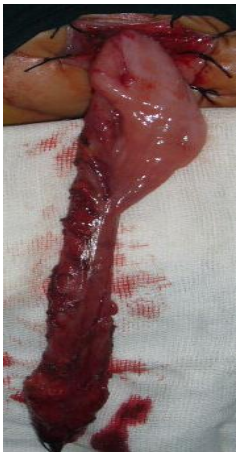

Photo5: Pulled mobilized colon

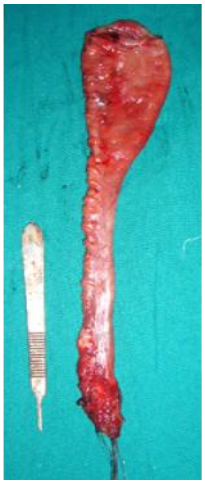

Photo6: Resected aganglionic segment

## Discussion:

Single stage transanal Soave pull-through procedure for HD makes itself superior by a shorter operating time, less bleeding, less morbidity and earlier recovery compared with similar trans-abdominal pull-throughs (Li ai-wu. et al; 2006). Theoretically it is the same operation that has been performed open for decades, but it avoids the need for laparotomy and abdominal mobilization of the rectum with their added risks (Langer. et al; 2003).

Langer in 2003, documented in his study the average gestational age at birth was 39 ± 0.78 weeks. The age at diagnosis was 30 days or less in 59.6% of his cases, 1 month to 1 year in 17%, and greater than 1 year only in 10.6%, with the mean age at diagnosis being 108.5 ± 100.2 days (range, 1 day to 12 years) (Langer. et al; 2003). Which is in agreement with our results.

In our study the age at time of operation ranged from 3 months to 30 months. However, it is considerably variable from the study of Prem Puri 2010, who reported that patient's age at the time of operation was ranged from 6 days to14 years (Prem Puri; 2010).

Approximately 80 % of patients present in the first few months of life with difficult bowel movements, poor feeding, and progressive abdominal distention. Up to 90 % of infants with HD fail to pass meconium in the first 24 hours of life (Holschneider. and Puri;2000) and this is nearly consistent with our results.

Rouzrokh. et al 2010, reported the mean operating time was 105 min. Only one child out of 86 needed blood transfusion during surgery, and the mean length of aganglionic segment was 20 cm (range 5–45) (Rouzrokh. et al; 2010). These data run in parallel with our results.

Length of postoperative hospital stay was ranged between 2 to 21 days (mean 5.3 days) (Prem Puri; 2010). Our results showed the mean length of post-operative hospital stay was 3.2 days (range 2.5- 5 days) which was less than prior reports.

Hirschsprung's disease associated with enterocolitis is the most serious and potentially life-threatening complication of HD, with significant

morbidity and mortality (Elhalaby. et al; 1995/ Wildhaber. et al; 2004). Engum. in 2004 and Vieten. in 2004, stated that HAEC remains the major source of morbidity and mortality in HD and the incidence is approximately 25%, occurring before surgical intervention as well as in the immediate postoperative period and several years after a definitive operation (Engum. and Grosfeld; 2004/ Vieten. and Spicer; 2004).

Enterocolitis has been considered one of the main problems in patients with HD both before and after definitive treatment (Julia et al; 2004). The general incidence of postoperative enterocolitis was as high as 12% to 33% (Skarsgard. et al; 1996/Marty. et al;. 1995). Van Leeuwen et al., reported a relatively high incidence of post pull-through enterocolitis in both abdominal and transanal approach (53% and 56% respectively) (Van Leeuwen et al; 2002).

Other reported studies documented that the incidence of post pull-through HAEC ranges from 4.6 to 54% in various series, which may be partially due to the use of different diagnostic criteria (Harrison. et al; 1986/Ikeda. and Goto; 1984/Hackam. et al; 1998). However, the majority of recent reports found the incidence of HAEC to be unrelated to type and timing of definitive surgery (Wildhaber. et al; 2004/Ikeda and Goto; 1984/Surana et al; 1994/Carneiro PMR. et al; 1992./ Sarioglu. et al; 1997).
In this study preoperative HAEC was found in 11 patients (19.3%). During the postoperative period, 5 cases (8.8%) developed HAEC, 1 of them during early postoperative follow up and the other 4 patients later on; all of them improved by medical treatment.

Among the different studies it is noticed that there is wide variation in the incidence of HAEC before and after surgical treatment of HD and this may be due to variation in the length of aganglionic segment, time of the operation and duration of postoperative follow up period.

Current studies (Pratapa. et al; 2007/Ekema. et al; 2003/Gao. et al; 2001) reported that post TEPT anastomotic stricture was occurred in about 4.6%. In our study, it was occurred in 1 patient only (1.8%) that was managed by serial anal dilatation at the outpatient clinic.

Normal bowel function is the primary goal of surgery for HD. Ekema et al,2003 (Ekema. et al; 2003) stated that postoperative soiling and

increased frequency of bowel movements after HD surgery may be due to over stretching of the pulled colon and it is temporary as the bowel movements will become normal in the majority of cases within 1 to 3 months. As regards to our results, 95% of the patients had complete anorectal continence and a continuous improvement in patients with soiling and incontinence (3 patients) was observed.

## Conclusion:

Transanal one stage endorectal pull-through is a feasible and safe procedure in children with rectosigmoid HD with satisfactory clinical outcome. Due to minimal invasive surgery and low complications, it offers many advantages and excellent results.

Most postoperative complications improved with time. However, this is our preliminary experience therefore; we recommend more studies for long-term outcome.

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## الملخص العربى

# عملية سحب القولون عن طريق الشرج بمرحلة واحدة لعلاج مرض الهرشسبرنج: خبرة *خمس سنوات*

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*أجريت هذه الدراسة على سبعة وخمسون طفلا ممن يعانون من مرض* الهرشسبرنج من سنة ٢٠٠٥إلى ٢٠١٠م بهدف تقييم نتائج عملية سحب القولون عن طريق الشرج، وقد تم تشخيص المرضى عن طريق الفحص الاكلينيكى وعمل أشعة بالجاستروجرافين على القولون وكذلك أخذ عينة للتحليل الباثولوجى، وتم إختيار المرضى لعملية سحب القولون عن طريق الشرج بدقة وعناية ،حيث أنه تم إجراؤها فقط *على المرضى الذين يعانون من هذا المرض فى منطقة المستقيم والقولون النازل فقط. وقد تم نسجيل ملاحظات النتائج أثناء وبعد العملية، وتبين أنه لم تحدث مضاعفات أثناء وبعد العملية فيما عدا خمسة مرضى عانوا من التهاب معوى قولونى بعد العملية والذى استجاب للعلاج ، وحدث أيضا ضيق بالشرج لمريض واحد فقط والذى تحسن بالتوسيع الشرجى ،كما أنه لوحظ زيادة فى عدد مرات التبرز لبعض المرضى وقد تحسن ذلك تدريجيا،وكل المرضى بعد العملية تمتعوا بتحكم إرادى للتبرز ما عدا مريض واحد الذى تحسن بالعلاج التحفظى وبمرور الوقت، ولم تسجل لدينا أى حالات وفيات أثناء وبعد العملية.

*ونستنتج من هذه الدراسةأن عملية سحب القولون عن طريق الشرج هى عملية امنة وسهلة ولها مدلول طيب فى النتائج ولا تمثل خطورة على المريض، كما أن المضاعفات الناجمة عنها قليلة، ولا تحتاج الى فتح استكشافى للبطن وما يحمله من مضاعفات أثناء وبعد العملية.*