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 and 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 (Protor 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 a, 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 with evaluation of 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 were included in this study; 39 boys and 18 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 1cm 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 cases with long aganglionic colonic segment seen in barium enema and patients with missed follow up for at least three months were 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 1cm 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 (table1).

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 3rd. 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 persistent soiling and perianal skin excoriation.

Seven out of the remaining 10 cases showed a steady improvement over the first year; so satisfied stool pattern confirmed in 54 patients (95%) at the last follow up period.

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%
Persistent soiling and perianal skin excoriation	1	1.8%
Anastomotic narrowing	1	1.8%
Total	***16***	***28%***

Photo1: HD with marked abdominal distension

Photo2:Contrast enema

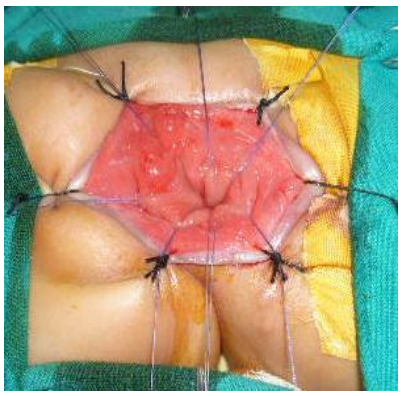

Photo3: Traction sutures

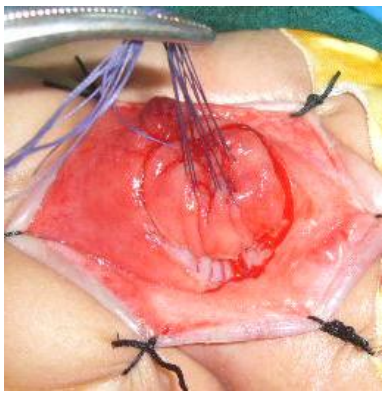

Photo4: Submucosal dissection

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(Liaiwu 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 et al; 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) , 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 ranged from 6 days to14 years.

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 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). These data run in parallel with our results.

Length of postoperative hospital stay 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(Elhslaby et al; 1995 and Wildhaber et al ; 2004). Engum and Grosfeld ;2004 and Vieten and Spicer; 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

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 and Marty et al;1995).Van Leeuwen et al 2002) reported a relatively high incidence of post pull-through enterocolitis in both abdominal and transanal approach (53% and 56% respectively).

Other previous 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 and 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 et al; 1992 and 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 and Gao et al;2001) reported that post TEPT anastomotic stricture occurred in about 4.6%. In our study, it 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 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 showed satisfactory stool pattern.

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.

REFERENCES

-*Amiel J, Lyonnet S. (2001): Hirschsprung disease, associated syndromes, and genetics: a review. J Med Genet. 38:729–39.*

-*Carneiro PMR, Brereton RJ, Drake DP et al. (1992): Enterocolitis in Hirschsprung's disease. Pediatr Surg Int 7:356–360.*

-*Coran AG, Teitelbaum DH.(2000): Recent advances in the management of Hirschsprung's disease. Am J Surg. 180:382–7.*

-*Ekema G, Falchetti D, Torri F et al (2003): Further evidence on totally transanal one-stage pull-through procedure for Hirschsprung's disease. J Pediatr Surg 38:1434–1439.*

-*Elhalaby EA, Coran AG, Blane CE et al. (1995): Enterocolitis associated with Hirschsprung's disease: a clinical–radiological characterization based on 168 patients. J Pediatr Surg. 30:76–83.*

-*Engum S. and Grosfeld J.(2004): Long-term results of treatment of Hirschsprung's disease. Semin pediatr surg. 13:273.*

-*Gao Y, Li G, Zhang X et al.(2001): Primary transanal rectosigmoidectomy for Hirschsprung's disease: preliminary results in the initial 33 cases. J Pediatr Surg 36:1816–1819.*

-*Hackam DJ, Filler RM, Pearl RH.(1998): Enterocolitis after the surgical treatment of Hirschsprung's disease: risk factors and financial impact. J Pediatr Surg 33:830–833.*

-*Harrison MW, Deitz DM, Campbell JR et al (1986): Diagnosis and management of Hirschsprung's disease. A 25-year perspective. Am J Surg. 152:49–56.*

-*Holschneider AM, Puri P.(2000): Hirschsprung's Disease and Allied Disorders. Amsterdam: Harwood Academic Publishers. 2nd ed.149-163.*

-*Ikeda K. and Goto S.(1984): Diagnosis, treatment of Hirschsprung's disease in Japan. An analysis of 1628 patients. Ann Surg. 199:400–405.*

-*Julia V, Castanon M, Tarrado X, Pinzon JC, Morales L.(2004): Transanal endorectal pull-through alone as treatment of Hirschsprung's diseas. Cir Pediatr. 17:85-88.*

-*Langer JC, Durrant AC, Torre L., Teitelbaum DH. et al (2003): One-Stage Transanal Soave Pullthrough for Hirschsprung Disease:A Multicenter Experience With 141 Children. Ann Surg. Vol. 238(4): 569–576, Oct.*

-*Langer JC, Fitzgerald PG, Winthrop AL, et al. (1996): One vs two stage Soave pull-through for Hirschsprung's disease in the first year of life. J Pediatr Surg. 31:33-37, 1996.*

-*Liai-wu, Zhang Wen-tong, LI Fu-hai, CUI Xin-hai, Duan Xiang-sheng (2006): A new modification of transanal Soave pull-through procedure for Hirschsprung's disease. Chin Med J. 119(1):37-42.*

-*Marty TL, Seo T, Matlak ME, Sullivan JJ, Black RE, Johnson DG.(1995): Gastrointestinal function after correction of Hirschsprung's disease: Long-term follow-up in 135 patients. J Pediatr Surg. 30:655-658.*

-*Parisi MA, Kapur RP. (2000): Genetics of Hirschsprung disease. Curr Opin Pediatr. 12:610–7.*

-*Pierro A, Fasoli L, Kiely EM, et al.(1997): Staged pull-through for rectosigmoid Hirschsprung's disease is not safer than primary pull-through. J Pediatr Surg. 32:505-509, 1997.*

-*Pratapa A, Gupta DK, Shaky VC.(2007): Analysis of problems, complications, avoidance and management with transanal*

pullthrough for Hirschsprung disease. J Pediatr Surg 42:1869–1876.

-***P**rem Puri.(2010): Systematic review and meta-analysis of enterocolitis after one-stage transanal pull-through procedure for Hirschsprung's disease. Pediatr.Surg.Int. 26:1101-1105.*

-***P**roctor ML, Traubici J, Langer JC, Gibbs DL, Ein SH, Daneman A, et al.(2003): Correlation between radiographic transition zone and level of aganglionosis in Hirschsprung's disease: implications for surgical approach. J Pediatr Surg. 38:775–8.*

-***R**ouzrokh M, Khaleghnejad A, Mohejerzadeh L, Heydari A, Molaei H. (2010): What is the most common complication after one-stage transanal pull-through in infants with Hirschsprung's disease? Pediatr Surg Int. 26:967–970.*

-***S**arioglu A, Tanyel FC, Buyukpamukcu et al.(1997): Clinical risk factors for Hirschsprung's associated enterocolitis II: postoperative enterocolitis. Turk J Pediatr 39:91–98.*

-***S**karsgard ED, Superina RA, Shandling B.(1996): Initial experience with one-stage endorectal pull-through procedures for Hirschsprung's disease. Pediatr Surg Int. 11:480-482.*

-***S**tewart DR, Von Allmen D. (2003): The genetics of Hirschsprung disease. Gastroenterol Clin North Am. 32:819–37.*

-***S**urana R, Quinn FMJ, Puri P.(1994): Evaluation of risk factors in the development of enterocolitis complicating Hirschsprung's disease. Pediatr Surg Int. 9:234–236.*

-***T**eitelbaum DH, Cilley RE, Sherman NJ, et al.(2000): A decade of experience with the primary pull-through for Hirschsprung disease in the newborn period: a multicenter analysis of outcomes. Ann Surg. 232:372-380.*

-***V**an Leeuwen K, Geiger JD, Barnett JL, Coran AG, Teitelbaum DH. (2002):Stooling and manometric findings after primary pull-throughs in Hirschsprung's disease: Perineal versus abdominal approaches. J Pediatr Surg. 37:1321-1325.*

-***V**ieten D. and Spicer R.(2004): Enterocolitis complicating Hirschsprung's disease. Semin Pediatr Surg. 13:263.*

-***W**ildhaber BE, Pakarinen M, Rintala RJ et al.(2004): Posterior myotomy/myectomy for persistent stooling problems in Hirschsprung's disease. J Pediatr Surg. 39:920–926.*

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 يناير سنة ٢٠٠٥-٢٠١٠ م بهدف تقييم نتائج عملية سحب القولون عن طريق الشرج، وقد تم تشخيص المرضى عن طريق الفحص الاكلينكى 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 تسجيل 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 انه لوحظ زيادة فى عدد مرات التبرز لبعض المرضى وقد تحسن ذلك تدريجيا وكل المرضى بعد العملية تمتعوا بتحكم إرادى ما عدا مريض واحد الذى تحسن بالعلاج التحفظى وبمرور الوقت ، ولم تسجل لدينا أى حالات وفيات أثناء وبعد العملية.

ونستنتج من هذه الدراسة أن 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.