J Gastrointest Surg (2011) 15:2246–2250
DOI 10.1007/s11605-011-1662-z

ORIGINAL ARTICLE

One-Stage Transanal Endorectal Pull-through for Treatment of Hirschsprung's Disease in Adolescents and Adults

Samir Ahmad Ammar · Ibrahim Ali Ibrahim

Received: 31 May 2011 / Accepted: 9 August 2011 / Published online: 10 September 2011

Abstract
Background One-stage pull-through operation has become increasingly popular for treatment of Hirschsprung's disease. The one-stage transanal pull-through was introduced in the late 1990s and has rapidly replaced traditional procedures in infants and young children in many surgical centers.
Objective The aim of this study is to determine feasibility and safety of transanal primary repair in adolescent and adults.
Methods Fifteen patients who underwent transanal endorectal pull-through were prospectively studied. All patients presented by chronic refractory constipation with the age ranged from 11 to 22 years. The patients were followed up for a mean of 18 months. Anal continence and postoperative complication were evaluated.
Results Incomplete continence in the form of soiling occurred in four patients (26.6%) and improved gradually with conservative management. No patients suffered from complete incontinence. Anastomotic strictures occurred in two patients and were successfully treated with regular dilatations. One patient had continued outlet obstruction and revision was considered for him. One patient complicated with low perianal fistula which needed fistulectomy. There was no impotence in adults.
Conclusion These findings indicate that one-stage transanal endorectal pull-through operation in adolescent and adults is feasible and safe.

Keywords Hirschsprung's disease · Constipation · Megacolon · Adult · Transanal pull-through

Introduction

Hirschsprung's disease (HD) is a congenital aganglionosis of the submucosal and myenteric neural plexuses principally affecting the rectosigmoid or rectal segments of varying length. Most cases manifest during the neonatal period,[1] but in rare instances, the disease is initially diagnosed in older children and adult patients.[2,3]

Swenson first described definitive surgical management of infants and children with HD in the late 1940s.[4] Because these children often presented with severe malnutrition or enterocolitis, a preliminary colostomy was usually done, followed by a pull-through procedure many months later. Earlier recognition and diagnosis of the disease led a number of surgeons in the 1980s to report series of single-stage pull-through procedures in small infants, using each of the three common operations (Swenson, Duhamel, and Soave). Since then, one-stage operations have become increasingly popular because of its safety and cost-effectiveness.[5] The one-stage transanal endorectal pull-through operation (TEPT) was introduced in the late 1990s and has rapidly replaced traditional procedures in infants and young children in many surgical centers around the world.[6–10] However, no studies address TEPT for treatment of HD in adolescent and adults.

S. A. Ammar
Surgery Department, Assiut University Hospitals,
Assiut, Egypt

S. A. Ammar (✉) · I. A. Ibrahim
General Surgery Department, Assiut University Hospital,
El Gamaa Street,
Assiut, Egypt
e-mail: samirahmed70@hotmail.com

The purpose of this study is to evaluate feasibility and safety of TEPT for treatment of HD in adolescent and adults.

Materials and Methods

The study protocol was approved by the ethics committee of our institution and all patients or their parents gave written informed consent. Data were prospectively collected on 15 consecutive patients (11 males and 4 females), all of whom underwent TEPT for HD in the Surgery Department, Assiut University Hospital between January 2004 and May 2010. TEPT was the standard operative technique for HD at Assiut University Hospital during the period of performance of this study. Patients with HD above 10 years old were included in the study. Patients having enterocolitis, acute obstruction not responding to conservative measures, and patients with bad general condition were excluded from this study. For the excluded patients, the initial surgical intervention consisted of formation of a leveling stoma in the ganglionic bowel and the definitive transabdominal pull-through procedure was performed electively at a later stage.

All the diagnoses were made based on clinical symptoms and barium enema showing the classic rectosigmoid transition zone. All patients reported long-standing refractory constipation as the predominant symptom. The diagnoses were confirmed by rectal biopsy showing absence of ganglion cells.

Preoperative chemical preparation was done using a third generation cephalosporin and metronidazole started 12 h before surgery. Mechanical preparation starts 2 days preoperatively using rectal wash two to three times daily. The results were expressed as the mean±SD or percentage.

Surgical Technique

The patient was anesthetized and placed in the supine lithotomy position. Caudal block was given to all patients. Complete relaxation using neuromuscular blocking agents excludes any interference due to reflex contraction of striated pelvic floor muscles and permits the level of the force necessary for the dilatation to be reduced. Complete relaxation and gradual dilatation prevent injury of the striated muscles. Anal retraction was performed using four traction sutures placed at the four corners of the anus (Fig. 1). The mucosa was incised circumferentially 1.5 cm above the dentate line, and a submucosal dissection was carried out proximally (Fig. 2) until above the peritoneal reflection. The submucosal dissection was carried out using combination of sharp and blunt dissection. To promote hemostasis and facilitate dissection 1:200,000 epinephrine was injected into the submucosa above the dentate line. The muscle of rectum was then incised circumferentially

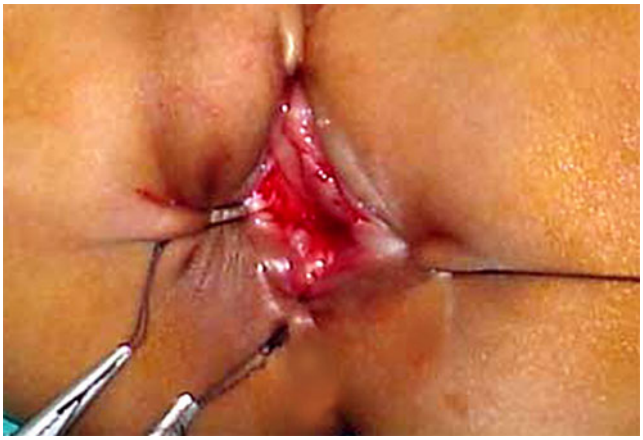

Fig. 1 Operative view showing four traction sutures placed at the four corners of the anus

allowing exposure of the full thickness of the colon, and the dissection was carried proximally along the outer wall of the rectal muscle. The vessels were ligated and divided just as they enter the bowel wall to avoid injury of the pelvic nerves and vessels and to avoid injuries of nearby structures such as vagina or prostate. The proximal mobilization and dissection of the colon were continued until the caliber was nearly normal. The bowel was then transected and the posterior wall of the muscular cuff was split. Coloanal anastomosis (Fig. 3) was done using braided absorbable suture material. A good bite of the colon was anastomosed to the underlying muscles of the rectum including a small bite of the distal mucosa. The anastomosis should be done above the dentate line so that the transitional epithelium is not damaged. This is important to prevent loss of

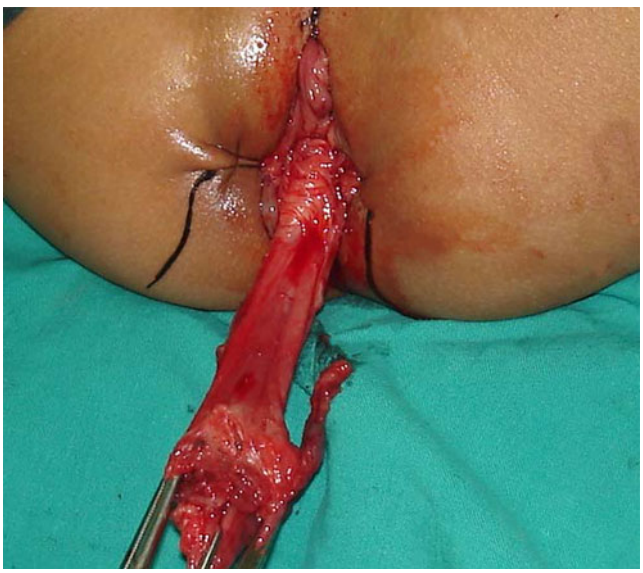

Fig. 2 Operative view showing submucosal dissection of the rectum

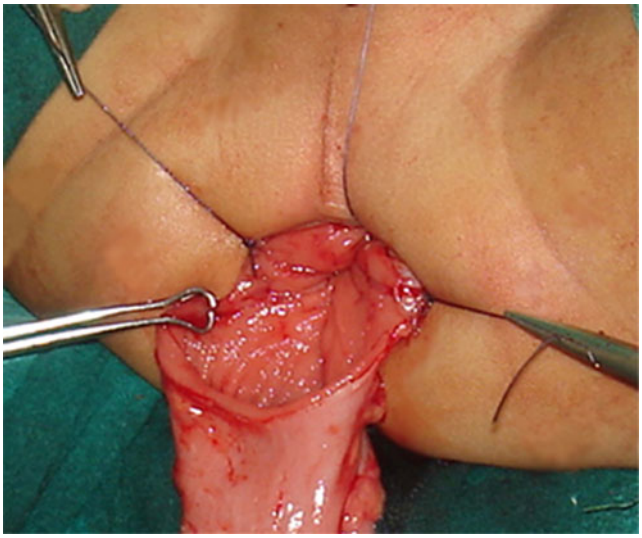

Fig. 3 Operative view of coloanal anastomosis

sensation, which may predispose to long-term problems with anal continence. A drain was placed paracolic and extracted after 24 h. Antibiotics were discontinued after 48 h, and feeding was begun when bowel function returned. The first per rectal examination of the patients was done 15 days postoperatively and followed by dilatation on regular basis in any patients with suspicion of anastomotic narrowing.

Results

Age at operation was 12.6±1.9 years (range 11–22). Operating time was 121±19.9 min (range 90–150). Bowel movements returned to normal within 24 h in all patients. Progression of oral feeding was uneventful. Postoperative hospital stay was 2.7±0.95 days (range 2–5). Two patients had postoperative bleeding (350–400 cc) and both of them were treated conservatively with blood transfusion without the need for re-exploration. The mean follow-up time was 18 months (range 6 months to 4 years). There was no urinary complication and no impotence in adults. None of the patients suffered from postoperative enterocolitis. The most frequent early complications after TEPT were frequent defecation (up to ten times per day), perineal dermatitis, and skin rash in four patients (26.6%). The rash was probably caused by frequent bowel movements. Frequent defecation and skin rash usually lasted for 6–10 weeks and improved gradually with conservative management. Anastomotic strictures occurred in two patients (13.3%) and were successfully treated with regular dilatations. One patient complicated with low perianal fistula which needed fistulectomy. There was no exposure of anal mucosa in any of the patients.

Functional Outcome

Continence was considered complete when the patient spontaneously evacuated soft stools, and there were no diurnal or nocturnal fecal soiling. When the patient had voluntary evacuations and few episodes of fecal soiling, he was considered partially continent. Data on fecal continence and bowel control of the patients were based on short-term follow-up. The frequency of stool in all patients 4 months after surgery was one to three bowel movements per day. None of the patients suffered from complete fecal incontinence. Partial incontinence in the form of soiling occurred in four patients (26.6%) and improved gradually within 10–16 weeks. One patient had continued outlet obstruction with failure of conservative management and revision was considered for him

Discussion

HD in adolescents and adults is a rare and frequently misdiagnosed cause of long-standing refractory constipation. All patients in this study reported long-standing refractory constipation as the predominant symptom. In these cases, the disease goes undiagnosed early because the proximal innervated colon can be hypertrophied and, thus, compensates for the distal obstructed, aganglionic rectum. In addition, these patients often try to relieve the constipation by taking cathartics and using enemas. Eventually, the dilated colon is no longer able to propel the feces distally. The term adult HD has been arbitrarily applied by some investigators to cases in which the patient is older than 10 years when the diagnosis is established,[11–13] whereas others have defined adult HD as cases in which the diagnosis was made after the age of 18 or 19 years.[14]

One-stage operation is safe, cost-effective, and avoids the morbidity of stomas. Specific stoma complications, including prolapse, stenosis, and wound infection, are prone to occur.[5,15] The requirement for multiple admissions and operations places a significant burden on both the family and the health-care system. Thus, the desire to avoid stoma creation and to reduce the duration of treatment prompted surgeons to adopt a strategy of primary one-stage repair once a definitive diagnosis of HD was established.[5,15] A preliminary colostomy is still needed in some conditions as presentation by complication as enterocolitis, acute on top of chronic intestinal obstruction not respond to conservative methods of treatment, and patients in bad general condition who cannot withstand major surgery.

In the early 1990s, Georgeson et al.[16] described a minimal access approach consisting of a laparoscopic biopsy to identify the transition zone, laparoscopic mobilization of the rectum below the peritoneal reflection, and a short endorectal

mucosal dissection from below. The anastomosis was done from below after prolapsing and excising the rectum. Subsequently, laparoscopic approaches have been described for the Duhamel and Swenson operations with good short-term results reported.[17,18]

In adults, transanal technique has been used successfully in the treatment of rectal malignancies, ulcerative colitis, familial polyposis coli, and colorectal vascular malformation.[19–23] The one-stage TEPT for the treatment of HD in infants and children was introduced in the late 1990s[6] and presents several advantages compared to classical pull-through techniques; it is a one-stage approach that can be conducted even during the neonatal period, previous colostomy is unnecessary, it is technically simple, no intraperitoneal adherence or scarring is observed, and it is associated with good fecal continence.[6–10]

Most of older children with congenital megacolon have short aganglionic segment.[24] All patients in the present study had short aganglionic segment and no patients required laparotomy. This is in contrast to congenital megacolon in infants and children by Tannuri et al.[25] who reported that three cases (8.5%) in TEPT group, age ranged from 10 days to 6 years, required laparotomies because the normoganglionic colon could not be reached or clearly identified.

All studies involving patients who underwent TEPT report the occurrence of coloanal anastomosis stricture although the incidence rate varies. The reported incidence of stricture ranged from 4.8% by Elhalaby et al.[8] to 43% by Stensrud et al.[26] Anastomotic stricture may be the result of ischemia of the lowest part of the mobilized colon and can be successfully managed by anal dilatations.[8,25,26] The incidence of anastomotic stricture in the present study was 13.3%. The strictures were easily treated and completely resolved with serial anal dilatations. There was no routine prophylactic dilatation because of the trauma, anxiety, and pain caused to the patient and to the parents.

During TEPT procedure, the use of four traction sutures and not using retractors is to avoid injury to the internal sphincter.[25] A general anesthesia in addition to regional sacral anesthesia induces satisfactory relaxation of both the internal and external sphincters. This makes it possible to perform free dissection of the rectal mucosa without using retractors. Attention to these fine details is important so as to avoid long-term continence issues from sphincter injury during the operation.[24,25] None of the patients in this study suffered from complete fecal incontinence.

Partial incontinence in the form of soiling occurred in four patients (26.6%). Patients with partial incontinence showed a steady improvement in their continence status. One multicenter study with a median follow-up of 12 months revealed that complete anorectal continence was achieved in 83.3% of patients who underwent TEPT older than 3 years.[8] In another study done by Tannuri et al.[25] complete continence was achieved in 70.8% of patients who underwent TEPT pull-through.

Conclusion

Hirschsprung's disease should be suspected in the context of refractory chronic constipation. One-stage TEPT in older children and adults is feasible and safe.

References

1. Roy CC, A. Silverman A, Alagille D. Congenital aganglionic megacolon (Hirschsprung's disease). In: Roy CC, Silverman A and Alagille D, ed, Pediatric clinical gastroenterology, 4th ed, Mosby, St. Louis, 1995 pp. 503–515.
2. Miyamoto M, Egami K, Maeda S, Ohkawa K; Tanaka N; Uchida E; Tajiri T. Hirschsprung's disease in adults: report of a case and review of the literature, J Nippon Med Sch. 2005; 72:113–120
3. Chen F, Winston III J, Jain S, Frankel W. Hirschsprung's disease in a young adult: report of a case and review of the literature. Ann Diagn Pathol 2006; 10: 347–351
4. Swenson O, How the cause and cure of Hirschsprung's disease were discovered. J Pediatr Surg. 1999; 34:1580–1581.
5. Somme S, Langer JC. Primary vs staged pull-through for the treatment of Hirschsprung disease. Semin Pediatr Surg. 2004;13:249–255
6. De la Torre-Mondragon L, Ortega-Salgado JA. Transanal endorectal pull-through for Hirschsprung's disease. J Pediatr Surg. 1998;33:1283–1286.
7. Langer JC, Minkes RK, Mazziotti MV Skinner MA, Winthrop A L. Transanal one-stage Soave procedure for infants with Hirschsprung's disease. J Pediatr Surg. 1999;34:148–151
8. Elhalaby EA, Hashish A, Elbarbary MM, Soliman HA, Wishahy MK, Elkholy A, Abdelhay S, Elbehery M, Halawa N, Gobran T, Shehata S, Elkhouly N, Hamza AF. Transanal one-stage endorectal pull-through for Hirschsprung's disease: a multicenterstudy. J Pediatr Surg. 2004;39:345–51.
9. Wester T, Rintala RJ. Early outcome of transanal endorectal pullthrough with a short muscle cuff during the neonatal period. J Pediatr Surg 2004;39:157–60.
10. Zhang SC, Bai YZ, Wang W, Wang WL. Clinical outcome in children after transanal 1-stage endorectal pull-through operation for Hirschsprung disease. J Pediatr Surg. 2005;40:1307–11.
11. Fairgrieve J. Hirschsprung's disease in the adult. Br J Surg. 1963;50: 506–514.
12. Barnes PR, Lennard-Jones JE, Hawley PR Todd IP. Hirschsprung's disease and idiopathic megacolon in adults and adolescents. Gut. 1986;27:534–541.
13. Kim CY, Park JG, Park KW, Park KJ, Cho MH, KIM WK. Adult Hirschsprung's disease. Int J Colorectal Dis. 1995; 10:156–160
14. Anuras S, Hade JE, Soffer E. Natural history of adult Hirschsprung's disease. J Clin Gastroenterol. 1984; 6: 205–210
15. Langer JC, Fitzgerald PG, Winthrop AL, Srinathan SK, Foglia RP, Skinner MA, Ternberg JL, Lau GY. One stage versus two stage soave pullthrough for Hirschspmng's disease m the first year of life. J Pediatr Surg 1996;31:333–36.
16. Georgeson KE, Cohen RD, Hebra A et al., Primary laparoscopic-assisted endorectal colon pull-through for Hirschsprung's disease: a new gold standard. Ann Surg. 1999; 229: 678–683

17. Travassos DV, Bax NM, Van der Zee DC. Duhamel procedure: a comparative retrospective study between an open and a laparoscopic technique. Surg Endosc. 2007; 21: 2163–2165.
18. Curran TJ, Raffensperger JG. Laparoscopic Swenson pull-through: a comparison with the open procedure. J Pediatr Surg. 1996; 31: 1155–1156
19. Maeda K, Maruta M, Sato H. Hanai T, Masumori K, Matumoto M, Koide Y, Matuoka H, Katuno H. Outcomes of novel transanal operation for selected tumors in the rectum. J Am Coll Surg. 2004; 199: 353–360
20. Zacharakis E, Freilich S, Rekhraj S, Athanasiou T, Paraskeva P, Ziprin P, Darzi A . Transanal endoscopic microsurgery for rectal tumors: the St. Mary's experience. Am J Surg. 2007; 194: 694-69
21. Fishman S J, Shamberger RC, Fox V L, Burrows P E. Endorectal pull-through abates gastrointestinal hemorrhage from colorectal venous malformations. J Pediatr Surg. 2000; 35: 982–984
22. Dolgin SE, Shlasko E, Gorfine S, Benkov K, Leleiko N. Restorative proctocolectomy in children with ulcerative colitis utilizing rectal mucosectomy with or without diverting ileostomy. J Pediatr Surg. 1999; 34: 837–840
23. Kartheuser AH, Parc R, Penna CP, Tiret E, Frileux P, Hannoun L, Nordlinger B, Loygue J. Ileal pouch-anal anastomosis as the first choice operation in patients with familial adenomatous polyposis: A ten-year experience. Surg. 1996; 119: 615–623
24. De La Torre L, Langer JC. Transanal endorectal pull-through for Hirschsprung disease: technique, controversies, pearls, pitfalls, and an organized approach to the management of postoperative obstructive symptoms. Semin Pediatr Surg. 2010; 19: 96–106
25. Tannuri A, Tannuri U, Romão R. Transanal endorectal pull-through in children with Hirschsprung's disease—technical refinements and comparison of results with the Duhamel procedure. J Pediatr Surg. 2009; 44: 767–772
26. Stensrud KJ, Emblem R, Bjørnland K. Functional outcome after operation for Hirschsprung disease-transanal vs transabdominal approach. J Pediatr Surg. 2010; 45(8):1640–4