

An experience with 53 patients of transanal endorectal pull-through for Hirschsprung's disease

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Background : *Transanal endorectal pull-through (TEPT) has been widely used in the treatment of Hirschsprung's disease (HD). We have been using this technique as described by Mondragon and Ortega - Salgado in our hospital since January 2004.*

Objective : *To evaluate the results of TEPT in the management of HD.*

Design : *Retrospective study.*

Setting : *Queen Sirikit National Institute of Child Health.*

Method : *Fifty - three patients underwent TEPT for HD from January 2004 to March 2011. Patients' hospital records were retrospectively analysed. The collected data included associated anomalies, age at diagnosis, enterocolitis before TEPT, weight at pull-through, site of transition zone, initial colostomy, primary TEPT without colostomy, age at pull-through, average length of resected bowel, operating time, blood loss, postoperative complications, daily bowel movements and continence. Follow-up period ranged from 2 weeks to 84 months.*

Results : *The mean operating time was 210 minutes. Average length of resected bowel was 13 cm. Average blood loss during operation was 24 cc. Transition zone was noted at rectosigmoid region in 36 patients (68%). Age at operation under six months was documented in 36 patients. Seven patients had anastomotic stricture managed by anal dilatation and additional anooplasty in 1 case. Twenty-one patients suffered from attacks of enterocolitis postoperatively at least one time. Frequent bowel movements gradually improved with time. Normal bowel function was documented in 46 of 53 patients (87%).*

Conclusions : *A one-stage pull-through for HD can be successfully performed using a transanal approach without intraperitoneal dissection. It takes less time, and has less bleeding, earlier recovery, less complication and no visible scar. Careful long-term follow up is required to assess daily bowel movements and continence.*

Keywords *Hirschsprung's disease, transanal endorectal pull-through, Soave operation.*

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วีระ บุรณะกิจเจริญ, วราภรณ์ มหรรธาตล. ประสบการณ์การผ่าตัดดึงลำไส้ใหญ่ที่มีกลุ่มเซลล์ประสาทมาต่อที่ไส้ตรงส่วนปลายผ่านทางทวารหนักในผู้ป่วย Hirschsprung's disease 53 ราย. จุฬาลงกรณ์เวชสาร 2554 พ.ย. - ธ.ค.; 55(6): 611 - 20

- ภูมิหลัง** : การผ่าตัดดึงลำไส้ใหญ่ที่มีกลุ่มเซลล์ประสาทมาต่อที่ไส้ตรงส่วนปลายผ่านทางทวารหนัก มีการใช้กันอย่างกว้างขวางในการรักษาผู้ป่วย Hirschsprung's disease ผู้รายงานใช้เทคนิคการผ่าตัดของ Mondragon และ Ortega-Salgado ตั้งแต่เดือนมกราคม 2547 จนถึงปัจจุบัน
- วัตถุประสงค์** : เพื่อศึกษาผลการรักษาโดยใช้เทคนิคการผ่าตัดดังกล่าว
- รูปแบบการวิจัย** : เป็นการศึกษาย้อนหลัง
- สถานที่ทำการศึกษา** : สถาบันสุขภาพเด็กแห่งชาติมหาราชินี
- วิธีทำการศึกษา** : ทำการศึกษาผู้ป่วย Hirschsprung's disease 53 รายที่ได้รับการผ่าตัดโดยวิธีดังกล่าวข้างต้น ตั้งแต่เดือนมกราคม 2547 ถึง เดือนมีนาคม 2554 โดยการนำเอกสารผู้ป่วยในมาทำการวิเคราะห์ย้อนหลัง ซึ่งประกอบด้วยข้อมูลดังนี้ ความผิดปกติที่พบร่วม ช่วงอายุที่ได้รับการวินิจฉัย จำนวนผู้ป่วยที่มีการอักเสบของลำไส้ก่อนการผ่าตัด นำหนักผู้ป่วยที่ทำผ่าตัด ตำแหน่งของลำไส้ที่เริ่มมีกลุ่มเซลล์ประสาท การมีลำไส้เปิดทางหน้าท้องก่อนการทำผ่าตัด จำนวนผู้ป่วยที่ทำผ่าตัดโดยปราศจากการมีลำไส้เปิดทางหน้าท้อง ช่วงอายุของผู้ป่วยที่ทำผ่าตัด ความยาวของลำไส้ที่ตัดทิ้ง ระยะเวลาที่ทำผ่าตัด ปริมาณเลือดที่เสียระหว่างผ่าตัด ภาวะแทรกซ้อนหลังผ่าตัด จำนวนครั้งที่ถ่ายอุจจาระในแต่ละวัน และความสามารถในการกลืนอุจจาระ มีการติดตามผลการผ่าตัดตั้งแต่ช่วง 2 สัปดาห์ ถึง 84 เดือน
- ผลการศึกษา** : ระยะเวลาทำผ่าตัดเฉลี่ย 210 นาที ความยาวเฉลี่ยของลำไส้ที่ตัดทิ้ง 13 ซม. สูญเสียเลือดในระหว่างผ่าตัดเฉลี่ย 24 ซีซี พบ transition zone บริเวณ rectosigmoid 36 ราย (คิดเป็น 68 เปอร์เซ็นต์) ผู้ป่วยอายุน้อยกว่า 6 เดือนได้รับการผ่าตัด 36 ราย ผู้ป่วย 7 ราย มีปัญหาลำไส้ตีบบริเวณที่ต่อลำไส้ รักษาด้วยการขยายลำไส้บริเวณที่ตีบ ยกเว้น 1 ราย ต้องผ่าตัดแต่งทวารหนัก ผู้ป่วย 21 ราย เกิดลำไส้อักเสบหลังผ่าตัดอย่างน้อย 1 ครั้ง จำนวนครั้งการถ่ายอุจจาระในแต่ละวันจะค่อย ๆ ดีขึ้นตามระยะเวลาหลังผ่าตัด ผู้ป่วยถ่ายอุจจาระปกติ 46 ราย (คิดเป็น 87 เปอร์เซ็นต์)

- สรุป** : การผ่าตัดรักษา Hirschsprung's disease โดยการดึงลำไส้ที่มีกลุ่มเซลล์ประสาทมาต่อในชั้นตอนเดียว สามารถทำได้โดยผ่านทางทวารหนัก โดยปราศจากการเลาะเนื้อเยื่อในช่องท้อง เทคนิคดังกล่าวใช้เวลาผ่าตัดน้อย เสียเลือดน้อย ผู้ป่วยฟื้นตัวเร็ว เกิดภาวะแทรกซ้อนน้อย และไม่เห็นแผลเป็น การติดตามผลในระยะยาวเป็นสิ่งจำเป็นเพื่อประเมินความถี่ในการถ่ายอุจจาระในแต่ละวัน และความสามารถในการกลั้นอุจจาระ
- คำสำคัญ** : โรค Hirschsprung's, transanal endorectal pull-through, Soave operation.

The surgical treatment of Hirschsprung's disease (HD) has developed new technique during the last two decades. The traditional multistage surgery has progressed from transabdominal exploration combined with the perineal approach or laparoscopically assisted one-stage repair in many cases during the neonatal period to a one-stage of totally transanal procedure.⁽¹⁻⁶⁾ The role of one-stage surgery for HD is well established and the results are comparable or better to those following two - or - three - stage operations.⁽⁷⁻⁹⁾ However, the totally transanal repair of HD is still being developed with various results.

Most surgeons using the transanal approach for HD perform an extensive endorectal dissection from the dentate line up to the peritoneal cavity following the principles of a classic soave operation.⁽¹⁻⁶⁾ The aim of this study is to present the preliminary experience with this operative approach.

Methods

This is a retrospective study. Hospital records of 53 (44 males, 9 females) patients who underwent

transanal pull-through for HD between January 2004 and March 2011 were reviewed. The patients' ages ranged from 1 day to 5 ⁶/₁₂ years. The data collected from the hospital records included associated anomalies, age at diagnosis, enterocolitis before TEPT, weight at pull-through, site of transition zone, initial colostomy, age at pull-through, the length of resected bowel, operation time, blood loss, postoperative complications, daily bowel movements and continence. HD was diagnosed only by barium enema. The site of ganglionic bowel for coloanal anastomosis was confirmed by intraoperative biopsy results analyzed by frozen section. The definite diagnosis of HD of the resected colon is later confirmed from permanent section.

The authors have used preoperative preparation and operative technique followed the same principles described by De la Torre-Mondragón and Ortega-Salgado except rectal mucosectomy.⁽³⁾ We have used a circumferential incision 0.5 -1 cm above the dentate line. A submucosal plane was dissected by bipolar cautery and was extended for 4 - 6 cm.



Figure 1. Submucosal plane was developed and extended

Oral feeding begins within the first operative 24 hours to 7 days. Antibiotic prophylaxis is continued until discharge. The patients are discharged when they have full enteral feedings.

The median follow-up time was 8 months (range 2 weeks - 84 months). The patients visited the outpatient clinic every 2 weeks for 1 month; then every 1 month for 3 months; and then every 3 to 6 months.

Results

Patient Demographics (Table 1)

The associated anomalies were detected in 6 of the 53 patients. Two patients had Down's syndrome with heart defects (ventricular septal defects). Three patients had heart diseases (1 atrial septal defect and 2 ventricular septal defects). One patient had imperforate anus with neurogenic bladder. In 16 infants (30%) the diagnosis was established during the neonatal period. Sixteen patients had preoperative enterocolitis. The mean weight at operation was 6.13 kilograms. In 46 children (87%), the site of transition zone was confined not over than the rectosigmoid region. Three patients had initial sigmoid colostomy above the transition zone because of severe enterocolitis. Fifty children (94%) underwent primary TEPT without colostomy. In this group, 36 patients underwent TEPT under the age of 6 months. Average length of the resected bowel was 13 cm (range 8 - 22 cm). The average operative time was 210 minutes (range 157 - 237 minutes) and average blood loss was 24 cc.

Postoperative Complications (Table 2)

Most of early post operative complications are perianal dermatitis or excoriation. Twenty - one patients (40%) had 1- 9 episodes of postoperative enterocolitis that were treated successfully with short-term gentamycin and metronidazole and regular rectal washouts. Most of them were attacked by only 1 episode of enterocolitis. Two patients required colostomy because of their severe enterocolitis. Nine of these 21 children (43%) had had at least 1 episode of preoperative enterocolitis. One patient had functional intestinal obstruction that required colostomy. Seven patients had anastomotic stricture requiring anal dilatation, except one that required anoplasty. Seventeen patients had perianal dermatitis or excoriation from frequent bowel movements, especially in the first 2 weeks after operation. These patients were managed by wound dressing and the use of barrier creams.

Functional Outcome (Table 3)

Frequent bowel movements, more than 10 times, were seen in the immediate postoperative period in 16 children (30%). This gradually improved over time from a median stool frequency of 4 (range 1 - 15) at 3 months after TEPT to 2 (range 1 - 4) stools per day at 2-years' follow-up.

Forty-six patients had normal bowel function giving the procedure an overall success rate of 87%. Of the remaining 7 patients, 4 had soiling more than once a week but 2 of them were improving with time, 1 had daily soiling and was treated with colostomy, and 2 had constipation .

Table 1. Patient data.

Patient demographics	
No. of patients	53
Male to Female ratio	44:9
Associated anomalies	6 (11%)
- Down's syndrome with heart defects	2
- Heart diseases (ASD, VSD)	3
- Imperforate anus with neurogenic bladder	1
Age at diagnosis (mo)	
< 1	16 (30%)
1 - < 6	25 (47%)
6 - < 12	2
≥ 12	10
Enterocolitis before TEPT	16 (30%)
Weight at pull-through (kg.)	6.13 (3.95 - 11.70)
Site of transition zone	
- Midrectum	10
- Rectosigmoid	36 (68%)
- Low sigmoid	5
- Midsigmoid	2
Initial colostomy	3
Primary TEPT without colostomy	50 (94%)
Age at pull-through (mo)	
< 1	5
1 - < 6	31 (58%)
6 - < 12	4
≥ 12	13
Average length of resected bowel (cm)	13 (8 - 22)
Average operative time (min)	210 (157 - 237)
Average blood loss (cc)	24 (5 - 120)

Table 2. Postoperative complications after TEPT operation.

Complications	Numbers
Postoperative enterocolitis	21
Cuff infection	-
Functional intestinal obstruction requiring colostomy	1
Anastomotic stricture	7
Perianal dermatitis / excoriation	17

Table 3. Functional results after TEPT or Hirschsprung's disease.

Functional outcome	Numbers
Daily bowel movements (median range)	
3 mo postoperatively	4 (1 - 15)
6 mo postoperatively	3 (1 - 7)
12 mo postoperatively	2 (1 - 5)
24 mo postoperatively	2 (1 - 4)
Continence	
Normal bowel habit	46
Soiling more than once a week	4
Daily soiling, requiring stoma	1
Constipation	2

Discussion

Totally transanal endorectal pull-through (TEPT) for HD was first reported by De La Torre-Mondragón and Ortega-Salgado in 1998.⁽³⁾ Since then many reports have been published. They clearly show that this approach is feasible for the treatment of classic rectosigmoid HD.^(1,4,8,10) It also can eliminate the possibility of pelvic nerve injury from pelvic dissection, bleeding, injury to other organs and adhesions formation.^(4,8) TEPT leaves no scars. It has also been indicated that total hospital costs are significant lower when transanal surgery is used.^(11,12)

Patients who had previous enterocolitis before TEPT did not have increased incidence of postoperative enterocolitis. Although most reports suggest that TEPT is suitable for rectosigmoid aganglionosis, we found that aganglionosis confined not over to the midsigmoid region can also be done with TEPT especially in the neonate or infant.⁽¹³⁾

The total operating time for TEPT was influenced by many factors including age (the younger, the less time needed), prior enterocolitis, adherent mucosa, intraoperative bleeding (the older the child, the more the bleeding), the time for frozen

section results and initial colostomy (the dissection required to free the stoma).

The postoperative complications are shown in Table 2. We found only a few patients of enterocolitis; one patient had functional intestinal obstruction, and 17 patients had acute perianal dermatitis or excoriation in early postoperative complications. Most enterocolitis and anastomotic stricture were documented in late postoperative complications. Postoperative enterocolitis developed in 40%. This rate was higher than many series.^(14,15)

Functional results are shown in Table 3. Frequent bowel movements were seen in the immediate postoperative period. Then it gradually improved within a few weeks to a few months. Most patients (87%) have normal bowel habit. Soiling may come from fecal impaction or enterocolitis and constipation may come from remaining of ganglionic dilated and thick-walled bowel.

Conclusion

Today, the totally transanal pull-through operation has a well-established feasibility for the management of classic rectosigmoid HD in all age groups and midsigmoid HD in infants. Further studies and long-term follow-up will be required for better results, especially in the points regarding how high above dentate line should the dissection start, length of the rectal cuff, how much of the ganglionated bowel should be resected, how to decrease rate of incontinence, frequent bowel movements and postoperative enterocolitis, ect.

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