

Long-term outcome of children after single-stage transanal endorectal pull-through for Hirschsprung's disease

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Background: Single-stage transanal endorectal pull-through (TEPT) is a new technique for surgery of Hirschsprung's disease (HD). TEPT can be assisted by laparoscopy (laparoscopic assisted transanal pull-through, LATP) or with non-additional procedure (total transanal endorectal pull-through, TTEP). This study was undertaken to evaluate the long-term outcome of these approaches in children with HD.

Methods: We retrospectively studied 131 patients (112 males and 19 females) aged 7 days to 14 years who underwent single-stage TEPT from October 2003 to July 2008. The medical records were reviewed for pre-, intra- and immediate post-operative complications. The data on stool pattern and complications were collected during the follow-up. Outcome was measured by continence evaluation score.

Results: No patients had intraoperative complications, but 5 had minor immediate postoperative complications. Late postoperative complications in 12 patients included enterocolitis (4 patients, one with severe enterocolitis died 7 months after operation), soiling (6) and constipation (2). There was a significantly higher frequency of stool in patients aged more than 36 months and those with a resected colon more than 30 cm ($P<0.05$). LATP showed significantly higher frequency of stool and soiling ($P<0.05$). Of the 54 patients who were older than 3 years at the time of follow-up, continence score was normal in 10, good in 39, fair in 3, and poor in 2. Seventy-seven patients achieved good bowel control in 12.8 ± 8.1 months after operation, 93.5% of whom within

24 months. Stool function was not improved in patients more than 30 months old after operation.

Conclusions: The long-term outcome of single stage TEPT was excellent. There were few postoperative complications, and stool pattern improved gradually to an excellent level within 24 months. Internal plication can be a good option for reducing the dilated proximal colon.

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Key words: Hirschsprung's disease; internal plication; laparoscopic assisted transanal pull-through; postoperative outcome; total transanal endorectal pull through; transanal endorectal pull-through

Introduction

Hirschsprung's disease (HD) is a congenital disease caused by aganglionosis of the rectum and colon. The principle of treatment is removal of the aganglionic segment and anastomosis. Since the first successful surgery by Swenson and Bill in 1946,^[1] a variety of procedures have been established. Laparoscopic assisted transanal pull-through (LATP) was the minimally invasive procedure described by Georgeson.^[2] Recently, the most popular procedure is the single-stage total transanal endorectal pull through (TTEP) which is first described by De la Torre-Mondragón and Ortega in 1998.^[3] In transanal endorectal pull-through (TEPT), the procedure is completed without diverting colostomy. TEPT can be assisted by laparoscopy as in LATP or without any additional procedure as in TTEP. This technique has various advantages over other traditional ones like Swenson, Duhamel and Soave procedure, and avoids the complications of laparotomy. Other benefits include better cosmesis, sphincter preservation, short operative time, short hospital stay, and cost-effectiveness.^[3,4] The short-term outcomes of the procedure are promising.^[4,5] TTEP was introduced into our hospital in 2001. This retrospective cohort study was conducted to evaluate the

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long-term outcome of TEPT for HD.

Methods

The study was conducted in the pediatric surgery department of First Hospital Affiliated to Zhengzhou University, China. Altogether 131 patients with HD who underwent TEPT (TTEP or LATP) from 2003 to 2008 were included in the study. Data on clinical history, examination findings, preoperative preparation, operative details, immediate post-operative complications were collected from medical records. All patients with HD were confirmed by clinical features, barium enema and histopathological examination. Patients were followed up at least 6 months after operation. Informed consent was obtained from the parents of the patients at the time of follow-up. The late complications, stool patterns, and stool control were recorded during the follow-up.

Surgical procedures

The patients were subjected to retrograde enema for preoperative bowel preparation. They underwent anorectal dissection through the transanal route as described by De la Torre-Mondragón and Ortega-Salgado in 1998.^[3] The dissection of whole rectum, at least up to peritoneal reflections was done in all patients. The colon was resected several centimeters proximal to the transition zone. Coloanal anastomosis was made transanally without proximal colostomy. Whether TTEP or LATP was performed depended on the barium enema results. If barium enema showed straight rectosigmoid and markedly dilated proximal colon (>10 cm), it was difficult to complete the procedure via the anus. Thus in the LATP group of the present study, the sigmoid mesorectum up to the desired length was dissected laparoscopically. In 4 patients, the planned TTEP was converted to LATP because of failure to mobilize the colon. But no patients received an open procedure.

Continence evaluation score

The outcome was measured by continence evaluation scoring system.^[6] The scoring system includes stool frequency, stool consistency, soiling, urgency period and requirement of diapers. The total score was 10; outcome was graded as normal (score 10), good (score 6-9), fair (score 3-5) and poor (score 0-2).

Statistical analysis

The data were analyzed using SPSS 11.5 for windows. Demographics, preoperative data, operative details, and early post-operative complications were expressed as means, standard deviation or median and ranges. The

operative outcome was compared by age of the patients, gender, type of the procedure, level of transitional zone, duration of the follow-up, and age at follow-up. Categorical variables were analyzed with the Chi-square test or Fisher's exact test where appropriate. Unpaired *t* test was used for continuous variable. The correlation test was used for two continuous variables. *P* value less than 0.05 was considered significant.

Results

Demographics

TTEP was performed in 98 patients (74.8%) and LATP in 33 (25.2%). Internal plication for reduction of the lumen of the colon was performed in 15 patients (8 in TTEP and 7 in LATP). Out of 131 patients, 112 (85.5%) were male and 19 (14.5%) were female with a male to female ratio of 5.9:1. The median age of the patients was 10 months with a range of 7 days-14 years. The mean weight of the patients was 10.7±6.8 kg ranging from 3 to 29 kg at the time of surgery. The mean diameter of dilated proximal colon was 5.9±1.7 cm (Table 1).

The level of the transition zone was observed in the the rectum in 4 patients, in rectosigmoid junction in 26, in the sigmoid colon in 83, long segment in 17 and total colonic aganglionosis in 1. The length of removed colon was 26.4±12.6 cm on average excluding total colonic aganglionosis. Gender, age and weight of the patients were similar in the TTEP and LATP groups. LATP was performed in the patients with long segment disease and markedly dilated proximal colon (*P*<0.05).

Postoperative complications

There were no intraoperative complications in the 131 patients. Five patients had immediate postoperative complications: perianal excoriation (3), early intestinal obstruction (1) and enterocolitis (1).

Follow-up

The average duration of the follow-up was 29.4±16.2 months ranging from 6 to 56 months. The average age of the patients at the time of follow-up was 51.6±36.1 (range: 6-171) months. Twelve patients had postoperative complications including enterocolitis (4 patients, one died of severe enterocolitis 7 months after operation), soiling (6) and constipation (2).

Younger age at operation (<3 years), TTEP and length of the removed colon (<30 cm) were associated with less bowel frequency (less than 3/day) (*P*<0.05). LATP, long segment disease, and removal of colon more than 30 cm were associated with soiling (*P*<0.05) (Table 2). All the patients who were treated with internal plication had no complications.

Table 1. Characteristics of patients

| | TEPT (n=131) | TTEP (n=98) | LATP (n=33) | P value |
|-------------------------------------|-----------------|----------------|----------------|---------|
| Gender | | | | 0.410 |
| Male | 112 | 85 | 27 | |
| Female | 19 | 13 | 6 | |
| Age at TEPT | | | | |
| Mean (mon) | 15±10.2 | 13±8 | 17±18.2 | 0.084 |
| Infants, n | 73 | 65 | 8 | |
| 1-3 years, n | 23 | 19 | 4 | |
| >3 years, n | 35 | 13 | 22 | |
| Weight at TEPT | | | | |
| Mean (kg) | 10.9±7.0 | 10.3±5.0 | 12.8±11.0 | 0.077 |
| Barium enema | | | | |
| Mean diameter of dilated colon (cm) | 5.9±1.7 | 5.5±1.2 | 7.6±2.0 | 0.001* |
| Level of aganglionosis | | | | 0.001* |
| Rectosigmoid region | 113 | 92 | 21 | |
| Long segment | 18 | 6 | 12 | |
| Mean length of colon resected (cm) | 26.4±12.6 | 20.3±8.6 | 31.4±15.7 | 0.001* |

*: $P < 0.05$, comparing between TTEP and LATP. TEPT: transanal endorectal pull-through; TTEP: total transanal endorectal pull-through; LATP: laparoscopic assisted transanal pull-through.

Table 3. Continence evaluation score of children above 3 years at the time of follow-up

| Parameters | Score | No. of patients (n=54) |
|-------------------------------------|-------|------------------------|
| Frequency of stool | | |
| Normal (1-2 per day) | 2 | 45 |
| Often (3-5 per day) | 1 | 7 |
| ≥6 per day | 0 | 2 |
| Stool consistency | | |
| Formed | 2 | 32 |
| Loose | 1 | 22 |
| Liquid | 0 | 0 |
| Soiling | | |
| Clean (never) | 2 | 48 |
| Occasional (1-6 per week/if liquid) | 1 | 4 |
| Permanent (every day) | 0 | 2 |
| Urgency period | | |
| Normal (in minutes) | 2 | 17 |
| Short (in seconds) | 1 | 34 |
| None | 0 | 3 |
| Requiring diaper | | |
| Never | 2 | 47 |
| Occasionally (<1 per week) | 1 | 5 |
| Continuously | 0 | 2 |

Table 4. Duration for the control of stool (n=77)

| Duration | No. of patients | % | Cumulative % |
|-----------------------------|-----------------|------|--------------|
| Immediately after operation | 2 | 2.6 | 2.6 |
| Within 6 mon | 24 | 31.2 | 33.8 |
| 6-12 mon | 24 | 31.2 | 65.0 |
| 13-18 mon | 5 | 6.5 | 71.5 |
| 19-24 mon | 17 | 22.0 | 93.5 |
| 25-30 mon | 5 | 6.5 | 100.0 |

Table 2. Outcome of followed up patients against different parameters (n=83*)

| | Frequency <3/day | Formed to loose stool | Soiling | Constipation |
|----------------------------|---------------------|--------------------------|---------|--------------|
| Age | | | | |
| <3 y (n=67) | 50 | 65 | 4 | 2 |
| >3 y (n=16) | 16 | 16 | 2 | 0 |
| P value | 0.033† | 1.0 | 0.326 | 1.0 |
| Gender | | | | |
| Male (n=75) | 52 | 73 | 5 | 2 |
| Female (n=8) | 8 | 8 | 1 | 0 |
| P value | 0.09 | 1.0 | 0.4 | 1.0 |
| Type of operation | | | | |
| TTEP (n=63) | 60 | 61 | 2 | 2 |
| LATP (n=20) | 11 | 20 | 4 | 0 |
| P value | 0.001† | 1.0 | 0.027† | 1.0 |
| Level of aganglionosis | | | | |
| Rectosigmoid region (n=76) | 61 | 75 | 2 | 2 |
| Long segment (n=7) | 6 | 7 | 4 | 0 |
| P value | 1.0 | 1.0 | 0.001† | 1.0 |
| Length of colon removed | | | | |
| <30 cm (n=61) | 56 | 60 | 1 | 2 |
| >30 cm (n=22) | 14 | 22 | 5 | 0 |
| P value | 0.004† | 1.0 | 0.004† | 1.0 |

*: died patient excluded. †: $P < 0.05$.

In the 54 children older than 3 years at the time of follow-up, 10 had normal function (score 10), 39 good (score 6-9), 3 fair (score 3-5), and 2 poor (score 0-2) (Table 3). Of 8 patients with long segment disease, 1 had normal, 3 had fair, 2 had fair and 2 poor function.

During the follow-up, 77 patients achieved good bowel control in 12.8±8.1 months (within one month to 38 months) after operation; 72 (93.5%) patients achieved good bowel control within 24 months after operation. Stool function was not improved in patients more than 30 months after operation (Table 4).

There was no significant correlation between the continence score and duration of follow-up after operation and age at follow-up ($r = +0.177$, $P = 0.095$ and $r = +0.164$, $P = 0.122$).

Discussion

Currently, TTEP is the most popular procedure for HD. It has been one decade since De la Torre-Mondragón and Ortega-Salgado^[3] first described this procedure in 1998. Early reports of the single stage TEPT have shown that the procedure is safe.^[4,7-10]

Age is not a criterion for choosing the procedure. Successful operations have been reported in patients of 7 days^[4] to 14 year.^[11] The early and late outcome of the procedure in neonates and young infants are similar to that in older children.^[5] One of the major concerns of pull-through in early infancy is that the delicate structure such as sphincter may be injured. In

the endorectal pull-through, the muscular coat of the rectum remains intact, thus the structure in front of the rectum and presacral nerve plexus is not at risk of operative damage.^[12] In our series, the youngest patient was 7 days with long segment involvement. Enema failed to relieve constipation, but the stool control was good at 29 months after the operation.

LATP was performed only if there are markedly dilated proximal colon, short mesentery (as evidenced by relatively straight look of the rectosigmoid in barium enema) or long segment. In TTEP, dissection was made by anal route almost up to the hepatic flexure of the colon. LATP was needed for total type. It was reported that there were no differences in outcomes after additional laparotomy or laparoscopy.^[10,13,14] In our study, soiling and constipation were observed more often in LATP because they were used for long segment disease.

Plication technique in 15 patients avoids removal of longer segment of normal colon. To reduce the lumen of the dilated colon, wedge resection of part of colon wall and anastomosis can be done. It is also feasible for hugely dilated colon more than 10 cm.

The most important measures of outcomes for HD procedure are continence and constipation.^[5,15-17] By the transanal approach the sphincter needs to be stretched. Anastomosis is made above the dentate line but still there is some injury to mucosa below the dentate line. These factors may adversely affect continence. The temporary loss of continence and intestinal function has been reported.^[11] The rate of constipation ranges from 3% to 28%^[18-22] and that of incontinence from 0 to 16% in various series.^[9,11,18] Zhang et al^[5] reported 5 cases of constipation and 3 of incontinence in a series of 58 patients after transanal one stage pull-through. Teitelbaum et al^[18] found constipation in 28% of 78 children. Elhalaby et al^[11] reported a complete continence in 35 out of 42 patients after one stage TEPT and 6 patients had constipation (14.3%). We found constipation in 2 patients (2.2%) and incontinence in 6 (6.6%). Multidisciplinary behavioral treatment is effective for incontinence and constipation after correction of anorectal malformation.^[23]

Teitelbaum et al^[18] reported a rapid decrease of stool frequency in the first 6 months and a slower decrease after 1-2 years after primary endorectal pull-through. Stool control was achieved at 34 months on average.^[6] Incontinence and other bowel functions improved with the patients' age irrespective of the procedure.^[5,24-27] We found that stool control was achieved at 12 months (median) and 93% of patients within 24 months.

Enterocolitis is a common operative complication. The rate of enterocolitis varies from 42% to 1.4%.^[18,28] Recent reports about single stage TEPT have shown a

lower rate of enterocolitis.^[5,28] Only 3 (3.3%) cases of enterocolitis were found in our series during the follow-up.

In conclusion, the long-term outcome of single stage TEPT is excellent. There are few postoperative complications, and stool pattern improves gradually to an excellent level within 24 months after surgery.

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