Original Article

Outcome of Transanal Endorectal Pull-Through in Patients with Hirschsprung's Disease

Tunde Talib Sholadoye, Oluseyi Oyebode Ogunsua, Yakubu Alfa[1], Philip Mari Mshelbwala[2], Emmanuel Adoyi Ameh[3]

Department of Surgery, Division of Paediatric Surgery, Ahmadu Bello University and Ahmadu Bello University Teaching Hospital, [1]Department of Surgery, Division of Paediatric Surgery, Ahmadu Bello University Teaching Hospital, Shika-Zaria, [2]Department of Surgery, University of Abuja and University of Abuja Teaching Hospital, [3]Department of Surgery, Division of Paediatric Surgery, National Hospital, Abuja, Nigeria

Abstract

Background: Transanal endorectal pull-through (TERPT) has become one of the preferred treatments for Hirschsprung's disease (HD) in our setting. This report aims to evaluate the current outcome of TERPT in the setting. **Materials and Methods:** A retrospective review of 71 children who had TERPT for histologically-confirmed HD in 11 years (2006–2017) in Nigeria. **Results:** There were 48 boys and 23 girls; aged 3 days–12 years at initial presentation (median = 10 months). Three (4.2%) patients had associated anomalies (duodenal atresia; anorectal malformation and sensorineural deafness with hypopigmented skin patches each). Age at TERPT was 2 months to 15 years (median = 3 years), with surgery waiting time of 1 month-14.9 years (median = 18 months). Sixty-six (93.0%) patients had rectosigmoid, four (5.6%) patients had long segment and one (1.4%) had total colonic disease. Five (7.0%) patients with large megacolon and one (1.4%) with the total colonic disease had assisted abdominal resection of the colon at TERPT. Seventeen (23.9%) patients had post-operative complications, including post-operative enterocolitis 8 (11.3%); anastomotic dehiscence 3 (4.2%); retained aganglionic segment 2 (2.8%); anastomotic stenosis 2 (2.8%), resulting in prolonged hospital stay ($P = 0.0001$; range = 1–30 days; median = 5 days). The mortality rate was 4.2% (3) from malignant hyperthermia in one patient and in 2 patients, the cause of mortality was unclear. Patients were followed up for 3–6 years (median = 3.5 years). Bowel movement stabilised to 2–4 times daily by 6 weeks after surgery. **Conclusion:** TERPT is a safe treatment for HD in this setting with good short-term outcomes. Longer follow-up is necessary to further evaluate the long-term bowel movement outcomes.

Keywords: Enterocolitis, Hirschsprung's disease, pull-through, transanal

Introduction

Transanal pull-through for Hirschsprung's disease (HD) is becoming the preferred treatment option worldwide. This technique has been practiced in Nigeria for over 15 years but less so in sub-Saharan Africa.[1] An important benefit of transanal pull-through is the absence or minimal abdominal manipulations, especially when combined with laparoscopy or laparotomy. Laparoscopy as an effective tool for colonic biopsy, identification of transitional zone, and mobilisation of the colon is well established.[2] Transanal endorectal pull-through (TERPT) is our preferred treatment of choice for HD and it is well accepted by the parents of our patients. There is a need to review the outcome of TERPT to identify its relevance in current practice. This is a report of the current short-term outcome of TERPT for HD in this setting.

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Materials and Methods

This is a retrospective review of 71 children who had TERPT as treatment of histologically confirmed HD from January 2006 to December 2017, at the Ahmadu Bello University teaching Hospital, Zaria. Patients who had other forms of surgical treatment of HD and histological diagnosis of hypoganglionosis were excluded. All patients had barium enema done and the diagnosis was histologically confirmed by rectal biopsy. The level of aganglionosis was determined by barium enema and on-table assessment in earlier years, while initial open multiple

Address for correspondence: Dr. Tunde Talib Sholadoye, Department of Surgery, Division of Paediatric Surgery, Ahmadu Bello University and Ahmadu Bello University Teaching Hospital, Shika-Zaria, Nigeria.
E-mail: hamedtalib2000@yahoo.co.uk

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colonic biopsies were performed in later years (2011–2013) to confirm the histologic level of aganglionosis before definitive surgery due to inaccuracy of visual and radiological assessment of level of ganglionic colon. Patients routinely had saline colonic washout for bowel evacuation initially at the hospital then parents were taught how to do it at home while waiting for TERPT. Initial colostomy was done for patients who had preoperative enterocolitis not responsive to nonoperative measures (resting the gastrointestinal (GIT), saline colonic irrigation) and in megacolon not responsive to colonic decompression by saline washouts.

The outcome was assessed using a Continence evaluation score.[3,4] The scoring system includes frequency of stool (normal (1–2/day) as 2, often (3–5/day) as 1, and ≥6/day as 0); stool consistency (formed as 2, loose as 1, and liquid as 0); soiling (clean (never) as 2, occasional (1–6/week/if liquid) as 1, and permanent (every day) as 0); urgency period (normal (in minutes) as 2, short (in seconds) as 1, and none as 0); and requiring diaper (never as 2; occasionally (<1/week); and continuously) in patients 3 years and above. 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). Data were analysed using SPSS version 22.0 (IBM Corp. 2013, IBM SPSS Statistics for Windows, Version 22.0. IBM Corp., Armonk, NY.). Chi-squared analysis was done to show the association of categorical data and a $P < 0.05$ was considered statistically significant.

Results

Demographics

There were 48 boys and 23 girls (M: F = 2:1) aged 3 days– 12 years at initial presentation (median = 10 months). Only 22 (31.0%) patients presented in the neonatal period [Table 1].

Twenty-three (32.4%, 3–6 days, median = 5 days) patients had a history of delayed passage of meconium, and 28 (39.4%) had intestinal obstruction in the neonatal period. One patient that presented with vomiting had associated upper GIT obstruction. Twenty-six (36.6%) patients had chronic constipation and 2 (16.9%) failure to thrive. Three (4.2%) patients had other associated congenital anomalies. One patient had type 2 duodenal atresia and midgut malrotation, another had anorectal malformation with recto vestibular fistula, while the last patient had sensorineural deafness, hypopigmented skin patches and subnormal mentation.

Initial colonic decompression

Sixty-five (91.5%) patients had daily normal saline colonic washouts (done by parents at home after being taught how to do it in the hospital) as required to have daily adequate colonic decompression. In 2 patients with preoperative Hirschsprung's-associated enterocolitis, initial colonic washouts were ineffective, necessitating colostomy and resection of markedly dilated segment of the colon. In another 12-year-old with severe megacolon, colonic washouts could not achieve adequate decompression, necessitating resection of megacolon and a colostomy. Six (8.5%) patients had daily digital anal stimulation (done by parents) to achieve decompression.

Table 1: Distribution of age at presentation of patients

Age range (months)	*n* (%)
≤1	22 (31)
≤12	28 (39.4)
12-48	15 (21.1)
49-108	4 (5.6)
≥109	2 (2.8)
Total	71 (100)

Table 2: Age at transanal endorectal pull-through in 71 patients

Age range (years)	*n* (%)
≤1	10 (14.1)
>1-5	46 (64.8)
>5-9	10 (14.1)
>9-13	4 (5.6)
>13-15	1 (1.4)
Total	71 (100)

Table 3: Post-operative complications following transanal endorectal pull-through (*n*=71)

Complication	Frequency (%)
Post-operative enterocolitis*	8 (11.3)
Anastomotic dehiscence	3 (4.2)
Anastomotic stenosis with faecal soilage[+]	3 (4.2)
Mortality	3 (4.2)
Incomplete resection	2 (2.8)
Malignant hyperthermia	1 (1.4)

*One of these patients developed acquired rectovaginal fistula at 17 months after surgery, [+]One of these patients was after redo surgery for anastomotic dehiscence and another was following enterocolitis after the initial surgery

Diagnosis

All patients had barium enema suggestive of HD. 91.5% (65) of patients had a partial-thickness rectal biopsy and due to inconclusive histology, a full-thickness biopsy was done in 8.5% (6) of patients to confirm the diagnosis of HD. Thirty eight percentage (27) of patients had multiple colonic biopsies though a mini-laparotomy or at colostomy to delineate the level of aganglionosis, while in the remaining 44 patients, level of aganglionosis was determined by a combination of barium enema and on-table examination rather than frozen section which was unavailable at the time. Sixty-six (93.0%) patients had rectosigmoid aganglionosis, 4 (5.6%) patients had long-segment aganglionosis and one patient (1.4%) had total colonic aganglionosis respectively

Transanal endorectal pull-through

The age at TERPT was 2 months to 15 years (median = 3 years), and only 10 (14.1%) patients had pull-through within the 1st year

of life [Table 2]. Overall, the waiting time for pull-through ranging from 1 month to 14.9 years (median = 18 months). Delay in surgery was attributed to several factors including delayed presentation to the hospital [Table 1]; delay in confirmation of diagnosis, and financial constraints as 51 (71.8%) patients' parent paid out of pocket, 9 (12.7%) patients' parent used the national health insurance scheme and only 11 (15.5%) patients' parent were able to pay for fast track surgery. Furthermore, the additional surgery of multiple colonic biopsies (27, 38%) to confirm the level of aganglionosis due to unavailability of frozen section further prolong the waiting time even in patients that had premium payment for the surgery.

At TERPT, the initial colostomy was taken down in the three patients who had a colostomy, before the commencement of the transanal approach. The standard technique of TERPT was used.[5,6] In patients with stoma takedown, two others with large megacolon, and one with total colonic aganglionosis, abdominal dissection was included to facilitate TERPT.

No patient had laparoscopic-assisted TERPT due to the unavailability of appropriate equipment.

Three patients who had resection of large megacolon had an intraoperative blood transfusion to replace blood loss, while 13 (18.3%) patients were transfused after surgery due to anaemia.

Oral feeding was commenced in 62 (87.3%) patients within 24 h after surgery but delayed in 5 (7.0%) patients who had postoperative enterocolitis (two of which initially commenced feeding but was stopped due to nontolerance), one that had delayed bowel function, and oral feeding was not commenced in the 3 (4.2%) patients that died.

Outcome

Seventeen (23.9%) patients had post-operative complications [Table 3] which were associated with prolonged hospital stay ($P = 0.0001$). Of the 17 patients, 15 had the TERPT after age 1 year of life. Eight patients (11.3%) developed enterocolitis after surgery and were managed non-operatively with the resolution of symptoms. Three (4.2%) patients had anastomotic dehiscence and required faecal diversion by colostomy and Swenson transanal and Duhamel pull-through (one patient had Duhamel done at another hospital) at a later date. In patients with incomplete resection of aganglionic segment (2, 2.8%), bowel management program was commenced. Three (4.2%) patients had anastomotic stenosis which resolved with dilatation.

The hospital stay was one–30 days (median 5 days). The longer hospital stay was in patients who had post-operative complications. Eighty-seven percentage of patients without complications were discharged within 1 week after surgery, while only 29.4% of those with complications were discharged within same period. Redo pull-through was done in another 2 patients (one that had residual aganglionic segment and another patient who had enterocolitis with rectovaginal fistula 17 months after surgery).

Three patients died, giving a mortality rate of 4.2%; one patient had clinically diagnosed malignant hyperthermia, while the circumstances that led to death in the other two patients were unclear. The parents declined an autopsy.

Patients were followed up for 3–6 years (median = 3.5 years). Initial stooling frequency after surgery was 6–15 times daily (median 8 times) but stabilised to 2–4 times daily within 4–10 weeks (median = 6 weeks) after surgery. By 3 months, majority of the patients (67, 94.4%) had 1–2 daily bowel movement. Sixty-six patients had continence evaluation and were scored 10-0. The continence evaluation as reported by the parents are normal bowel function in 12 (18.2%) patients, good bowel function in 51 (77.3%) patients, fair bowel function in one (1.5%) patient and poor bowel function in two (3.0%) patients.

Discussion

Transanal endorectal technique with rectal mucosectomy, resection of the aganglionic colonic segment and pull-through of normal ganglionic colon, followed by primary coloanal anastomosis done without laparotomy or laparoscopy was described over two decades ago.[5] Evidence has shown that the transanal approach is superior to the abdominal approach in operative time, hospital stay, cosmesis, lack of complications associated with laparotomy and risk of incontinence and constipation following surgery.[7-9] Therefore, most paediatric surgeons developed the necessary skills to offer varying types of transanal approach.

In Nigeria, paediatric surgeons offer varying techniques based on skills, training, available resources and patient factors such as length of aganglionosis, age at presentation and presence of colostomy. In one report, it was observed that only about 35% of paediatric surgeons indicated that they performed TERPT for their patients with HD in Nigeria and our centre is one of the pioneers for TERPT in this region.[1]

Presentation

HD is one of the top ten most common causes of intestinal obstruction among neonates in sub-Saharan Africa.[10] This is underestimated due to the late presentation of the disease in this setting and delayed presentation remains a major challenge. While the age of presentation is observed to be decreasing,[10] only about a third of patients in this study presented within the 1st month of life compared to reports from high-income countries and parts of Asia where neonatal presentation is more common representing about 60%–90% of their patients.[6,11]

TERPT is an effective and acceptable treatment for most patients with HD, particularly in patients with the short-segment disease.[9] TERPT is not widely performed in sub-Saharan African, perhaps due to delayed presentation. Where transanal pull-through is performed, the Soave type is more common than the Swenson technique in Nigeria.[1]

TERPT has been the surgical choice for the treatment of HD in our centre for about two decades now. Long-segment disease requires laparotomy or laparoscopically assisted approach. In this setting, however, the use of laparoscopy is still limited by availability and resources. To facilitate TERPT in longer segment disease, we utilised periumbilical laparotomy assist. Patients with preexisting colostomy had stoma takedown, transanal resection of the aganglionic segment and pull-through of the proximal stoma. This is possible due to retention of dilated colon during the creation of the stoma. We observed previously using the very dilated colon for stoma creates problems with massive stoma prolapse and difficulties with stoma care for the families. In addition, the calibre of the colon remains relatively large for a long time even up to the time of definitive surgery.

TERPT is safe in children,[6] even though most of our patients presented late with a median of 3 years at the surgery.

The outcome of TERPT is comparable to other methods in terms of commencement of diet, complications, hospital stay and outcome,[11] but was found to be significantly better than the two-stage Duhamel and other abdominal approaches in operative time and length of hospital stay following surgery.[9,11,12] In one report comparing transanal Swenson with transanal Soave (27 patients for each group), there was no significant differences in operation time, intraoperative complications, early outcomes and intermediate outcomes.[13] However, another report[14] observed that transanal Swenson was better than Soave pull through in terms of lower volume of blood loss, shorter operation time and lower complication rate but the sample size was small and the difference did not reach statistically significance.

Key advantages of transanal endorectal pull-through

One-stage TERPT is our standard practice and an initial colostomy is usually reserved for cases of megacolon or patients with enterocolitis unresolved with non-operative measures. The transanal approach is adequate except in marked megacolon or when a longer segment colon is involved.[6] To reduce the risk of post-operative obstructive symptoms, constipation and enterocolitis, a short muscular cuff with incision of the cuff posteriorly was found to be adequate though other surgeons do a V-shaped partial resection in the posterior wall of the muscular cuff.[15] In the present report, blood transfusion was required in patients who had long colonic resections or marked megacolon as in other reports.[6,9,15]

TERPT allows for early commencement of oral intake after surgery, and this was achieved within 24 h in the majority of the patients in the present report. Other studies reported oral intake ranging from 2 h to 9 days.[6,9,11] Delayed oral intake is more so in patients who had an abdominal assisted procedure or post-operative complications such as enterocolitis in this report.

Median hospital stay following TERPT in our patients was 5 days as in other reports.[6,9,14,15] This was much shorter when the surgery was done in the neonatal period,[6] but more prolong with the abdominal approach,[9] and when post-operative complication occur.

Complications

In this study, delayed surgical treatment and longer segment HD are associated with a higher incidence of complication. This was more so among patients older than 1 years at surgery as observed in an earlier study on TERPT.[16] One report observed that there is a lower complication rate with good to excellent continence score rate in patients who had surgery by 42 months of life compared to those older.[17] Another report observed better outcomes in patients less than 3 years at surgery with short-segment disease and about 50% of patients with long-segment disease had complications.[4] It may well be that the risk of complications after surgery is less if TERPT for HD is undertaken at a younger age. This could be due to reduced number of episodes of enterocolitis with reduced fibrosis from inflammation making dissection during surgery minimal and better healing.

The rate of post-operative complication was lower in this study but comparable to other studies on TERPT.[6,15] The most common complication was post-operative enterocolitis (11.3%), and this may occur as early as 24 h after surgery. Some reports observed the incidence of Hirschsprung's associated enterocolitis to be 5%–50% depending on the definition,[18] while the majority had a lower rate between 6% and 8.8%.[6,15] This varying definition and assessment makes it possible to miss mild preoperative enterocolitis that may present early after surgery. Several factors may predispose to enterocolitis such as delayed surgery,[16] retained proximal hypoganglionosis[19] and recurrent preoperative enterocolitis. Retained hypoganglionosis could predict occurrence and severity of postoperative enterocolitis.[19]

The occurrence of incomplete resection could be avoided if there were facilities for intraoperative frozen section to determine the ganglion status of bowel segment before anastomosis, but even where this is available, residual aganglionosis or hypoganglionosis could still occur.[6,15,16] Resecting part of proximal ganglion-containing dilated part of the bowel would minimise the risk of retained aganglionic segment and risk constipation after surgery.

Mortality following TERPT is uncommon,[6,9] but could occur when there are severe associated anomalies such as major cardiac anomalies,[6] or malignant hyperthermia as seen in the present report.

When symptoms of constipation persist after surgery, a repeat rectal biopsy is necessary,[20] to identify residual aganglionosis and redo-surgery is mandatory.[6,11,20] In this report, redo pull-through was transanal Swenson's to avoid the associated peri-anastomotic fibrosis/stricture. Similar approaches have been used by other surgeons where open approach or Duhamel pull-through was done.[6]

Monitoring of symptoms after surgery for years is essential to determine a true representative of the outcome of TERPT.[4,6,14]

Unresolved bowel function symptoms usually improved in most patients after surgery with appropriate intervention. This is expected within 30 months after which little or no improvement is seen.[4] Therefore, follow-up beyond 3 years in the present report is required.

Conclusion

TERPT is a safe treatment for HD associated with low morbidity and mortality, and offer patients a scar-less procedure. Long-term follow-up is necessary to adequately assess the outcome of surgery and intervene appropriately when necessary.

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Conflicts of interest

There are no conflicts of interest.

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