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Single-stage transanal endorectal pull-through procedure for correction of Hirschsprung disease in neonates and nonneonates: A multicenter study☆,☆☆

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ABSTRACT

Purpose: The outcomes of single-stage transanal endorectal pull-through (SSTEPT) for Hirschsprung disease (HSCR) in young patients are favorable; however, reports have shown that diagnosis and surgery at young ages increase the risk for postoperative enterocolitis and slows postoperative recovery. The present study was primarily designed to evaluate the outcomes of SSTEPT in a multi-institutional cohort of neonates and nonneonates with HSCR.

Methods: Between August 2005 and May 2012, a total of 650 children with HSCR were divided into the following two groups: group A (neonatal group, operative age < 28 days [n = 186]); and group B (nonneonatal group, operative age > 28 days [n = 464]). The short-term outcomes were postoperative enterocolitis, perianal excoriation, and anastomotic stricture and leakage rates. The midterm outcomes were incomplete continence and constipation rates based on multi-institutional chart review. Statistical analyses were performed using chi-square (χ^2) tests.

Results: Follow-up was completed in 112 neonates and 303 nonneonates. Short-term outcomes indicated a higher incidence of perianal excoriation (27.6% vs. 6.6%, $\chi^2 = 33.70$, $p < 0.05$), anastomotic strictures (14.3% vs. 6.0%, $\chi^2 = 27.18$, $p < 0.05$), anastomotic leakage (8.0% vs. 1.7%, $\chi^2 = 8.36$, $p < 0.05$), and postoperative enterocolitis (40.2% vs. 10.2%, $\chi^2 = 49.05$, $p < 0.05$) in group A compared to group B. Midterm outcomes indicated a higher incidence of incomplete continence (35.7% vs. 14.9%, $\chi^2 = 21.85$, $p < 0.05$) in group A compared to group B.

Conclusion: Performing single-stage transanal endorectal pull-through in the nonneonatal period may be more appropriate than the neonatal period. There were higher rates of perianal excoriation, anastomotic strictures and leakage, postoperative enterocolitis, and incomplete continence postoperatively in neonates than nonneonates.

Hirschsprung disease (HSCR) is a developmental disorder of the enteric nervous system that is characterized by the absence of ganglion cells in the myenteric and submucosal plexuses of the distal intestine, which results in a peristalsis in the affected bowel and functional intestinal obstruction [1,2]. Short or rectosigmoid HSCR is a type of HSCR that is characterized by a spastic segment extending to the rectum or sigmoid colon. Definitive treatment involves a surgical pull-through procedure resecting the aganglionic portion of the colon. There are three approaches to early treatment, as follows: single-stage transanal endorectal pull-through (SSTEPT) during the neonatal period; multi-stage transanal endorectal pull-through (MSTP), which is characterized by a leveling colostomy during the neonatal period, followed by delayed coloanal reconstruction later in infancy; and anal dilatation or a cleansing enema during the neonatal period followed by SSTEPT during the nonneonatal period. Over time, SSTEPT has been the most commonly performed procedure for HSCR, and has been confirmed to be preferred by comparing SSTEPT and MSTP [3].

The SSTEPT outcomes for HSCR are not always as good as the surgeon may perceive. Postoperative incomplete incontinence, constipation, and enterocolitis should not be ignored [4–8]. Previous studies have attempted to correlate preoperative features to surgical outcome

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in patients with HSCR, such as gender, length of aganglionosis, age at the time of surgery, preoperative enterocolitis, associated anomalies, and genetic background [9–12], but none have yielded definitive conclusions. The proximal intestine with normal ganglia is dilated daily because of persistent spasm of the distal intestine. After infancy, most children with HSCR require staged procedures owing to dilated bowel; postoperative complications are uncommon [13]. Zakaria [14] suggested that SSTEPT is performed with some difficulty in older children; yet, the follow-up results were statistically poorer when compared with patients who had undergone surgery at a younger age. Although more and more surgeons agree with early diagnosis and surgery for HSCR [15], the results are contradictory based on the observations reported to date. Morbidities (perianal erosion or excoriation, enterocolitis, and soiling) were higher in neonates or young infants less than 1 months of age [16–18]. Haricharan et al. [19] reported that children diagnosed with HSCR at younger ages are at greater risk for postoperative enterocolitis. Huang et al. [20] reported that SSTEPT in neonates required significantly shorter operative times and postoperative hospital stays, but longer postoperative recovery periods than nonneonates. Thus, what is the appropriate timing to undergo surgery for HSCR? This study was primarily intended to assess whether or not performing SSTEPT in neonates is superior to nonneonates.

1. Methods

1.1. Study design

A retrospective multi-institutional cohort study was conducted to evaluate SSTEPT in neonates and nonneonates with short or rectosigmoid Hirschsprung disease (SRSHSCR). Postoperative enterocolitis, perianal excoriation, anastomotic stricture or leakage, constipation, and continence after SSTEPT were assessed.

1.2. Patients

Between August 2005 and May 2012, a total of 650 children with SRSHSCR (497 boys and 153 girls; age range, 4 days–6 months; mean age, 109.6 days) underwent SSTEPT without laparotomy or laparoscopy at Nanjing Children's Hospital, Zhengzhou Children's Hospital, and Hebei Children's Hospital. The children were divided into the following two groups: group A (neonatal group, operative age < 28 days [n = 186]) and group B (nonneonatal group, operative age > 28 days [n = 464]; Table 1). All of the patients with HSCR were diagnosed preoperatively based on barium enema, anorectal manometry, and biopsy specimen pathology, or a combination of at least two of these procedures preoperatively and confirmed by postoperative pathologic examination. Children with Down syndrome, or other comorbidities that can impair functional outcomes and normal global development were excluded.

1.3. Surgical technique

Anal dilatation was performed or cleansing enemas were administered in group B patients 1–5 months preoperatively and no perforations were detected. Transanal endorectal pull-through was performed in all neonates and nonneonates. After induction of general anesthesia, patients were placed in the lithotomy position and a Foley catheter was placed to decompress the bladder during surgery. The mucosal incision was made 5 mm proximal to the dental line. A dilute epinephrine solution was injected in the submucosa. Multiple interrupted 4/0 sutures were placed in the proximal cut edge of the mucosal cuff, and traction was applied while the endorectal submucosal dissection was carried out proximally. Dissection of full thickness was initiated from the level of the peritoneal reflection. Frozen section biopsies were used to detect the level of ganglionosis in all cases. The rectal muscular sheath was split longitudinally from the posterior aspect and resected in a V-shape. The rectal muscular sheath adjacent to the internal sphincter was used in the postoperative histopathologic specimen to confirm the diagnosis of HSCR. One-third of the internal sphincter was divided as in a partial sphincter myotomy. The proximal colon was anastomosed just above the dental line using absorbable interrupted 4/0 sutures.

1.4. Assessment of short-term outcomes

The short-term outcomes included postoperative enterocolitis, perianal excoriation, and anastomotic stricture and leakage rates. Perianal excoriation and anastomotic stricture complications were assessed approximately 3 months postoperatively (range, 3–4 months; mean, 3.4 months), and anastomotic leakage was assessed when it happened, but all anastomotic leakages happened less than 1 week postoperatively (range, 4–7 days; mean, 5.7 days). Postoperative enterocolitis was assessed <1 year postoperatively (range, 10–12 months; mean, 11.4 months). Postoperative enterocolitis was defined according to the 2009 Hirschsprung's-associated enterocolitis (HAEC) scoring system, which consisted of history, physical examination, radiologic, and laboratory findings. Sixteen variables were evaluated for the HAEC score. Each variable had a corresponding score and at the end of the interview, the scores were added to give a total HAEC score. Scores >10 were consistent with HAEC [21]. The demographic and perioperative data for the study population are shown in Table 2.

1.5. Assessment of midterm outcomes

Midterm outcomes (incontinence and constipation rates) were assessed approximately 36 months after SSTEPT by the pediatric incontinence or constipation score system (PICSS; range, 33–36 months; mean, 35.6 months; Table 3) [22,23]. According to the PICSS, three variables were shown to be decisive for the incontinence score only, five variables were decisive for the constipation score only, and five variables were equally involved in incontinence and constipation. The age-specific mean scores and 95% confidence intervals (CIs) for incontinence and constipation were calculated by testing the 13 variables on healthy children. Based on the PICSS, children who scored below the age-specific lower limit of the 95% CI were considered to have incomplete continence or constipation. The incomplete continence and constipation rates were compared between the neonatal and nonneonatal groups. Furthermore, the mean PICSS score was compared for the difference in degree of incontinence or constipation between the two groups. Significance was set at a $p < 0.05$.

The study was approved by the Ethics Committees of Nanjing Children's Hospital. The diagnosis, type of operation, and outcomes were confirmed based on medical records from each participating

Table 1
Patient details.

	Pediatric surgery department in hospital 1 (n = 381, male 300, female 81)	Pediatric surgery department in hospital 2 (n = 258, male 191, female 67)	Pediatric surgery department in hospital 3 (n = 11, male = 6, female = 5)
Group A Neonates (≦28 days) N = 186	103	78	5
Group B Nonneonates (>28 days) N = 464	278	180	6