



Feasibility and efficacy of home rectal irrigation in neonates and early infancy with Hirschsprung disease

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Abstract

Purpose A single-stage pull-through (SSPT) is the most commonly performed procedure for Hirschsprung disease (HSCR) and has been shown to be better than multi-stage procedures. However, performing a SSPT in the neonatal period or early in infancy is a risk factor for an inaccurate diagnosis, post-operative enterocolitis, and a protracted post-operative recovery. The present study was primarily designed to evaluate the feasibility and efficacy of home rectal irrigation in the neonatal period and early in infancy, followed by a delayed and planned SSPT in a prospective cohort with HSCR.

Methods Between January 2014 and December 2016, a total of 147 neonates diagnosed with HSCR were enrolled in the study. Six patients were excluded as a result of ganglion cells found in second rectal biopsies after the neonatal period. One hundred twenty-two patients successfully underwent 2–4 months of home rectal irrigation during the neonatal period, followed by a SSPT procedure after the neonatal period (group A, $n = 122$). Nineteen patients were not candidates for home rectal irrigation, and thus, colostomies were performed during the neonatal period followed by multi-stage procedures after the neonatal period (group B, $n = 19$). One hundred twenty-two healthy children, age- and gender-matched to group A were enrolled as the healthy control group for assessment of nutrition status (group C, $n = 122$). The birth weight, gender ratio, aganglionic segment, age, and Hirschsprung-associated enterocolitis (HAEC) score at the time of HSCR diagnosis were measured to evaluate the feasibility of home rectal irrigation in neonates and early in infancy. The nutritional indices, including weight, body length, serum albumin, serum prealbumin, serum retinol-binding protein, and incidence of HAEC after 2–4 successful home rectal irrigation, were used to assess the efficacy of home rectal irrigation. Anastomotic strictures or leakage, perianal excoriation, frequency of defecation, and morbidity of post-operative HAEC were recorded to evaluate the beneficial effects to pull through (PT), which were facilitated by home rectal irrigation.

Results Higher HAEC scores and older age at the time of diagnosis of HSCR were associated with group B, compared to group A (4.34 ± 1.25 vs. 11.0 ± 2.56 [$t = 18.20$, $p < 0.05$] and 2.8 ± 1.46 days vs. 12.1 ± 5.3 days [$t = 16.10$, $p < 0.05$], respectively). The ratio of rectosigmoid HSCR to non-rectosigmoid HSCR was higher in group A than group B (104/18 vs. 4/15 [$\chi^2 = 34.29$, $p < 0.05$]). There were no differences in birth weight, weight at the time of diagnosis of HSCR, and gender ratio between groups A and B. There were no differences in birth weight, birth length, post-home rectal irrigation age, post-home rectal irrigation weight, post-home rectal irrigation length, and post-home rectal irrigation serum albumin between groups A and C (3.47 ± 0.42 kg vs. 3.48 ± 0.40 kg [$t = 0.10$, $p > 0.05$], 50.02 ± 0.49 cm vs. 50.05 ± 0.46 cm [$t = 0.61$, $p > 0.05$], 98.59 ± 13.34 days vs. 97.83 ± 13.58 days [$t = 0.44$, $p > 0.05$], 6.77 ± 0.66 kg vs. 6.97 ± 0.87 kg [$t = 1.95$, $p > 0.05$], 61.55 ± 2.14 cm vs. 61.70 ± 2.07 cm [$t = 0.59$, $p > 0.05$], and 41.78 ± 2.42 g/L vs. 41.85 ± 2.37 g/L [$t = 0.22$, $p > 0.05$], respectively). The rate of HAEC in the period of home rectal irrigation in group A was low; however, the post-home rectal irrigation serum prealbumin level and retinol-binding protein were significantly lower in group A than group C (0.15 ± 0.04 g/L vs. 0.17 ± 0.05 g/L [$t = 3.50$, $p < 0.05$] and 22.51 ± 7.53 g/L vs. 30.57 ± 9.26 g/L [$t = 7.46$, $p < 0.05$], respectively). There were

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no anastomotic strictures or leakage after definitive PT performed in group A. The frequency of defecation ranged from 2–6 times per day, 10 patients had perianal excoriation 3 months after PT, and 11 patients had post-operative HAEC during 6 months of follow-up after PT.

Conclusion Home rectal irrigation in neonates and early in infancy, followed by a delayed and planned SSPT is feasible and effective in patients with HSCR, and could be beneficial to definitive PT. However, for patients with an extended aganglionic segment, older age, or high HAEC score at the time of diagnosis of HSCR, rectal irrigation maybe not suitable.

Trial registration This was a prospective comparative study designed to evaluate the effects of home rectal irrigation for facilitating and enhancing recovery after PT, and was registered at Clinical Trials.gov as NCT02776176.

Keywords Hirschsprung disease · Home rectal irrigation · Neonate · Early infancy

Introduction

Hirschsprung disease (HSCR) is characterized by the absence of ganglion cells (aganglionosis) in the myenteric and submucosal plexuses of the distal gut, thus, resulting in persistent spasm in the affected bowel and a functional intestinal obstruction [1, 2]. HSCR is classified as rectosigmoid HSCR, long-segment HSCR, and total colonic HSCR based on the aganglionic segment, and definitive treatment is a surgical pull-through resecting the aganglionic portion of the colon [3, 4]. Over time, single-stage pull-through (SSPT) has become the most common procedure for HSCR [3–8]; however, the best time to perform SSPT for a patient with HSCR has not been established. There are many studies that have shown that a SSPT performed after the neonatal period compared to during the neonatal period for correction of HSCR has many limitations, such as lower diagnostic accuracy and higher rates of perianal excoriation, anastomotic strictures or leakage, post-operative enterocolitis, and post-operative incomplete continence [9–11]. Moreover, an online questionnaire distributed to the European Paediatric Surgeons' Association (EUPSA) members showed that pull through (PT) is performed at the time of diagnosis by 33% and delayed by 67% (4 months or > 5 kg) [12]. Another survey to evaluate the management of Hirschsprung's disease in the UK and Ireland indicated that most surgeons aim to perform definitive surgery at < 3 months of age [13]. Thus, home rectal irrigation during the neonatal period and early in infancy followed by a delayed and planned SSPT procedure may also be an option for correction of HSCR; however, the feasibility and efficacy of home rectal irrigation during the neonatal period and early in infancy have not been reported. This study was primarily intended to assess the feasibility and efficacy of home rectal irrigation during the neonatal period in patients with HSCR.

Methods

Study design

A prospective study was conducted to evaluate the feasibility and efficacy of home rectal irrigation during the neonatal period and early in infancy for patients with HSCR. The parents learned the technique of home rectal irrigation in the hospital and after mastering the technique, home rectal irrigation was performed. Definitive surgery is suggested to be performed at < 3 months of age; however, not all parents comply with the suggestion. The ultimate duration of home rectal irrigation ranged from 2–4 months. The status of the patients before and after 2–4 months of home rectal irrigation was evaluated.

Inclusion criteria and patients

Low birth weight, prematurity, non-HSCR gastrointestinal anomalies, and mechanical ventilation have been reported as important risk factors for colostomies during the neonatal period instead of home rectal irrigation during the neonatal period [7, 14]. The inclusion criteria were as follows: patients diagnosed with HSCR during the neonatal period; and rectosigmoid, long-segment, and total colonic HSCR. The exclusion criteria were as follows: low birth weight; non-HSCR gastrointestinal anomalies; mechanical ventilation; and early preterm birth.

Between January 2014 and December 2016, a total of 147 children diagnosed with HSCR during the neonatal period treated at the Children's Hospital of Nanjing Medical University met the inclusion criteria and were enrolled in the study. All of the patients were diagnosed with HSCR based on barium enema and pathologic evaluation of suction rectal biopsy specimens during the neonatal period. Home rectal irrigation was performed in the neonates diagnosed with HSCR to wash out retained stool; however, six patients were excluded because ganglion cells were found in second rectal biopsies after the neonatal period. One hundred twenty-two patients successfully underwent

home rectal irrigation during the neonatal period and early in infancy, followed by the SSPT procedure (group A, $n = 122$). Nineteen patients were not candidates for home rectal irrigation (massive bloody stools in eight patients, recurrent abdominal distension in 10 patients, and intestinal perforation in one patient), and thus, colostomies were performed during the neonatal period followed by a multi-stage PT procedure after the neonatal period (group B, $n = 19$). One hundred twenty-two healthy children, age- and gender-matched to group A were enrolled as the healthy control group for assessment of nutrition status (group C, $n = 122$). In China, babies must be examined for growth and development from 42 days of life to 3 years of age; blood testing is part of the normal health examination. We obtained and used data with consent of the parents of the children. The study was authorized and supervised by the Institutional Ethics Committee of the Children's Hospital of Nanjing Medical University.

The study flow chart is shown in Fig. 1.

Home rectal irrigation technique

Rectal irrigation is a common technique for bowel preparation before PT in patients with HSCR, and can effectively wash out retained stool in the colon. The details of the technique include placing a soft tube into the distal colon and repeatedly injecting 0.9% NaCl for irrigation, and 100–150 mL of 0.9% NaCl as needed. In the present study, the parents of the patients were given instructions on how to perform rectal irrigation by professional physicians and nurses; the duration of training usually lasted approximately 1 week. Effective rectal irrigation was defined as no abdominal distention and hematochezia in patients with normal oral feeding. After the technique of rectal irrigation was mastered, all of the parents performed rectal irrigation 1–2 times every day at home after hospital discharge. Communication between each parent and physicians or nurses was established after hospital discharge, and questions were answered and advice was offered, as needed.

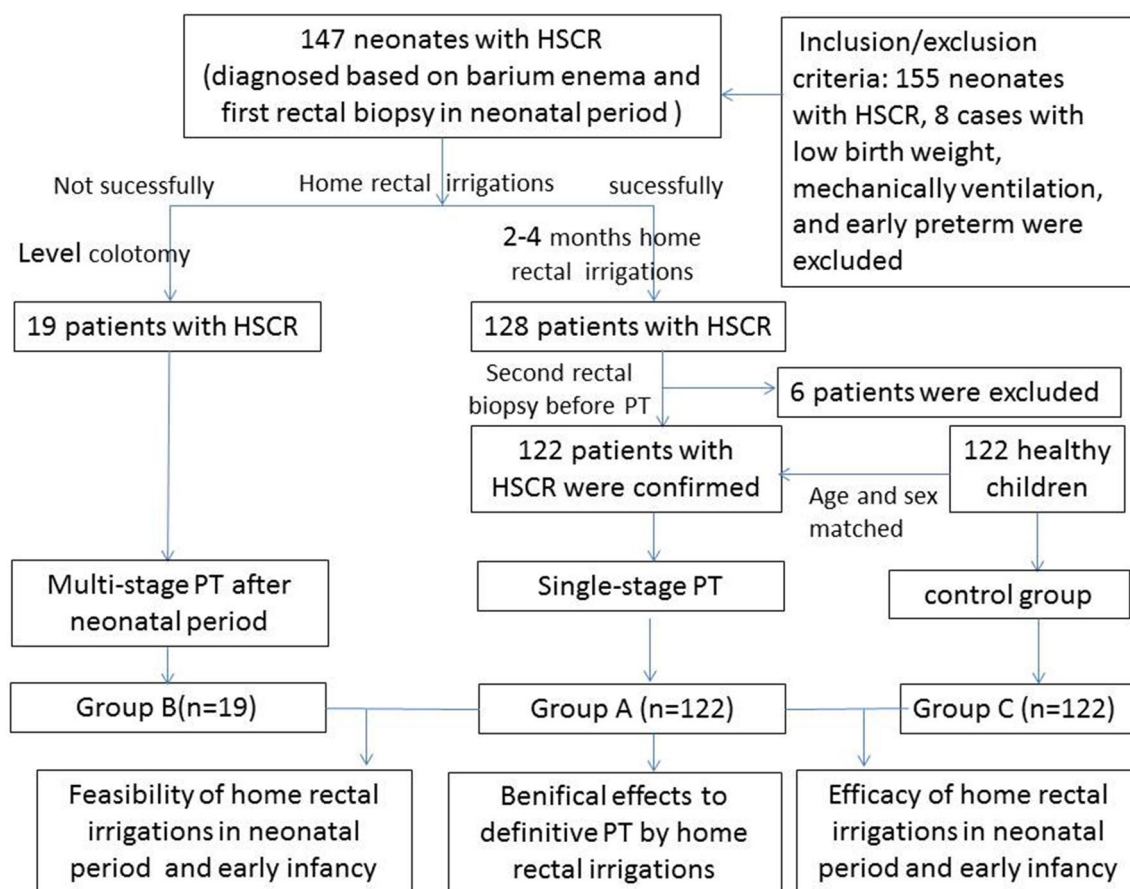


Fig. 1 The flow chart of participants entered into the study

Assessment of physical status at the time of HSCR diagnosis during the neonatal period

None of the patients in groups A and B passed meconium in the first 24 h after birth. The birth weight, gender ratio, aganglionic segment, age, and Hirschsprung-associated enterocolitis (HAEC) score were assessed at the time of HSCR diagnosis in groups A and B to evaluate the feasibility of home rectal irrigation during the neonatal period. The HAEC score system was approved in 2009 [15], and consists of historical data, physical examination findings, and radiologic and laboratory testing results. Sixteen variables are evaluated to determine the HAEC score. Each variable had a corresponding score and at the end of the assessment, the scores were added to give a total HAEC score. Traditionally, scores > 10 indicate HAEC, while the most recent research suggests that scores > 4 indicate HAEC [16, 17]. In the current study, we also defined HAEC as scores > 4.

Assessment of physical status after 2–4 months of successful home rectal irrigation in group A and the healthy control group (group C)

A period of 2–4 months (range 60–118 days; mean 94.87 ± 13.02 days) of successful home rectal irrigation was performed before SSPT in group A. The HAEC rate, weight, body length, serum albumin level, serum prealbumin level, and serum retinol-binding protein were recorded in group A after 2–4 months of successful home rectal irrigation and in group C to evaluate the efficacy of home rectal irrigation.

Assessing beneficial effects of definitive PT which facilitated home rectal irrigation in group A

Every patient was followed for at least 6 months after PT. The operative time of PT, the rate of anastomotic strictures or leakage, perianal excoriation, frequency of defecation, and morbidity of post-operative HAEC were recorded to evaluate the beneficial effects which facilitated home rectal irrigation. Perianal excoriation and anastomotic stricture complications were assessed approximately 3 months

post-operatively (range 3–4 months; mean 3.3 months), and anastomotic leakage was assessed at the time of occurrence, but there were no anastomotic strictures and leakage in group A after PT. Post-operative enterocolitis was assessed approximately 6 months post-operatively (range 6–7 months; mean 6.5 months). Post-operative enterocolitis was also be defined as a score > 4 according to the 2009 Hirschsprung-associated enterocolitis (HAEC) scoring system.

Statistical analysis

Data are presented as percentages and the mean $\pm$ standard deviation. The level of statistical significance was set at 0.05. Statistical analysis was performed using Chi-square (χ^2) tests and *t* tests (*t*) or comparison of differences between groups. SPSS software (version 18.0) was used for statistical analyses.

Results

Results of physical status in groups A and B at the time of diagnosis of HSCR during the neonatal period

Higher HAEC scores and older age at the time of diagnosis of HSCR were associated with group B compared to group A (4.34 ± 1.25 vs. 11.0 ± 2.56 [$t = 18.20$, $p < 0.05$], and 2.8 ± 1.46 days vs. 12.1 ± 5.3 days [$t = 16.10$, $p < 0.05$], respectively). The ratio of rectosigmoid HSCR to non-rectosigmoid HSCR was higher in group A than group B ($104/18$ vs. $4/15$ [$\chi^2 = 34.29$, $p < 0.05$]). There were no differences in birth weight, weight at the time of diagnosis of HSCR, and gender ratio between the two groups (Table 1).

Results of physical status in groups A and C after 2–4 months of successful home rectal irrigation

There were no differences in birth weight, birth body length, post-home rectal irrigation age, post-home rectal irrigation

Table 1 Assessment of physical status at the time of HSCR diagnosis in the neonatal period

Group	Group A ($n = 122$)	Group B ($n = 19$)	t/χ^2	p
Birth weight (kg): <i>M</i> (SD)	3.47 ± 0.42	3.48 ± 0.39	0.15	> 0.05
Sex (male/female): <i>n/n</i>	96/26	14/5	0.04	> 0.05
Age at diagnosis of HSCR (days): <i>M</i> (SD)	2.8 ± 1.46	12.1 ± 5.3	16.10	< 0.05
Weight at diagnosis of HSCR (kg): <i>M</i> (SD)	3.47 ± 0.39	3.40 ± 0.27	0.67	> 0.05
HAEC score at diagnosis of HSCR: <i>M</i> (SD)	4.34 ± 1.25	11.0 ± 2.56	18.20	< 0.05
Aganglionic segment (rectosigmoid/non-rectosigmoid)	104/18	4/15	34.29	< 0.05

M represents the mean, *SD* represents the standard deviation

weight, post-home rectal irrigation body length, and post-home rectal irrigation serum albumin between the two groups (3.47 ± 0.42 kg vs. 3.48 ± 0.40 kg [$t=0.10$, $p>0.05$], 50.02 ± 0.49 cm vs. 50.05 ± 0.46 cm [$t=0.61$, $p>0.05$], 98.59 ± 13.34 days vs. 97.83 ± 13.58 days [$t=0.44$, $p>0.05$], 6.77 ± 0.66 kg vs. 6.97 ± 0.87 kg [$t=1.95$, $p>0.05$], 61.55 ± 2.14 cm vs. 61.70 ± 2.07 cm [$t=0.59$, $p>0.05$], and 41.78 ± 2.42 g/L vs. 41.85 ± 2.37 g/L [$t=0.22$, $p>0.05$], respectively). The rate of HAEC during the period of home rectal irrigation in group A was low (4.1%); however, the post-home rectal irrigation serum prealbumin and retinol-binding protein levels were significantly lower in group A than group C (0.15 ± 0.04 g/L vs. 0.17 ± 0.05 g/L [$t=3.50$, $p<0.05$] and 22.51 ± 7.53 g/L vs. 30.57 ± 9.26 g/L [$t=7.46$, $p<0.05$], respectively; Table 2).

Outcomes of definitive PT facilitated by home rectal irrigation in group A

The operative time ranged from 140 to 180 min. There was no anastomotic strictures or leakage after definitive PT performed in group A. Ten patients still had perianal excoriations 3 months after PT and 11 patients had post-operative HAEC 6 months after PT. Among the post-operative HAEC patients, seven were defined as Grade I and four patients were defined as Grade II by guidelines for diagnosis and management of HAEC, which were published in 2017 [18]. There was no constipation and the frequency ranged from 2 to 6 times per day 6 months after PT (Table 3).

Table 3 Assessment of recovery of PT after 2–4 months of home rectal irrigation

Group	Group A ($n=122$)
Sex (male/female): n/n	96/26
Age (days): M (SD)	98.59 ± 13.34
Operative weight (kg): M (SD)	6.77 ± 0.66
Operative time (min): range~(M)	140–180 (160.82)
Anastomotic strictures or leakage: n (%)	0
Perianal excoriation: n (%)	10 (8.1%)
Frequency of defecation: range~(M)	2–6 (3.9)
Post-operative HAEC: n (%)	11 (9.02%)

M represents the mean, SD represents the standard deviation

Discussion

Feasibility of home rectal irrigation during the neonatal period for HSCR

Rectal irrigation, a conventional and effective means for pre-operative preparation, can relieve the bowel obstruction caused by an aganglionic segment in patients with HSCR [10]. Home rectal irrigation is effective when a tube is carefully placed into the dilated segment [19, 20]. The subsequent growth and development of patients are satisfactory; however, intestinal perforation is a serious complication [21]. Thus, it is essential to determine which patients with HSCR are suitable for home rectal irrigation during the neonatal period followed by SSPT. Moreover, the feasibility and efficacy of home rectal irrigation during the neonatal period also needs to be determined.

Low birth weight, prematurity, non-HSCR gastrointestinal anomalies, and mechanical ventilations are risk factors

Table 2 Assessment of physical status after 2–4 months of successful rectal irrigation in group A and the control group

Group	Group A ($n=122$)	Control group/ group C ($n=122$)	t/χ^2	p
Birth weight (kg): M (SD)	3.47 ± 0.42	3.48 ± 0.40	0.10	>0.05
Birth body length (cm): M (SD)	50.02 ± 0.49	50.05 ± 0.46	0.61	>0.05
Sex (male/female): n/n	96/26	96/26	0	>0.05
Duration of home rectal irrigation (days): M (SD)	94.87 ± 13.02	—	—	—
Age (days): M (SD)	98.59 ± 13.34	97.83 ± 13.58	0.44	>0.05
Weight (kg): M (SD)	6.77 ± 0.66	6.97 ± 0.87	1.95	>0.05
Body length (cm): M (SD)	61.55 ± 2.14	61.70 ± 2.07	0.59	>0.05
Serum albumin (g/L): M (SD)	41.78 ± 2.42	41.85 ± 2.37	0.22	>0.05
Serum prealbumin (g/L): M (SD)	0.15 ± 0.04	0.17 ± 0.05	3.50	<0.05
Serum retinol binding protein: M (SD)	22.51 ± 7.53	30.57 ± 9.26	7.46	<0.05
HAEC incidence: n (%)	5 (4.1%)	—	—	—

M represents the mean, SD represents the standard deviation

for home rectal irrigation [7, 14] and such patients were excluded from the current study. Are there any other risk factors associated with home rectal irrigation failure during the neonatal period for HSCR. The HAEC score indicates the severity of HAEC [16–18]; a higher HAEC score is associated with a poorer prognosis. In the current study, the patients in group B who failed home rectal irrigation had higher HAEC scores than those in group A, which indicated that the severity of HAEC may be a risk factor for home rectal irrigation failure. The neonates who had high HAEC scores may not be suitable for home rectal irrigation. Older age at the time of HSCR diagnosis, especially > 1 week, leads to delayed diagnosis and treatment of HSCR, which is a risk factor for HAEC [22, 23]. In the current study, the patients in group B who failed home rectal irrigation were older at the time of HSCR diagnosis and had higher HAEC scores, which indicated that older age at the time of HSCR diagnosis is also a risk factor for failing to undergo home rectal irrigation.

Previous studies have reported that an extended length of aganglionic segment may be an important risk factor for HAEC [24–26]; however, there is an opposing opinion [23]. In the current study, 19 neonates who failed to undergo home rectal irrigation had a higher rate of non-rectosigmoid HSCR with the aganglionic segment extending proximal to the sigmoid colon, which indicated that an extended length of aganglionic segment may be a risk factor for failing to undergo home rectal irrigation that cannot be ignored. To relieve the bowel obstruction caused by an aganglionic segment, effective home rectal irrigation should be performed by placing a tube into the dilated segment. In fact, an extended length of aganglionic segment proximal to the sigmoid colon may increase the difficulty of placing the tube in the dilated colon for home rectal irrigation and decrease the effect of home rectal irrigation, which may result in HAEC. Furthermore, overzealous attempts at tube placement may lead to intestinal perforation [21]. Thus, delayed age at diagnosis, a high HAEC score, and an extended aganglionic segment might be significant parameters to evaluate the feasibility of home rectal irrigation during the neonatal period.

Efficacy of home rectal irrigation during the neonatal period for HSCR

The subsequent growth and development of patients should be as normal as healthy children undergoing home rectal irrigation, however, the duration should not be indefinite. It has been reported that the growth and development are satisfactory for chronic constipation after performing effective home rectal irrigations [20, 21]. To evaluate the efficacy of home rectal irrigation to HSCR, the nutritional status regarding the growth and development were assessed after 2–4 months of home rectal irrigation. Weight, body

length, and serum albumin level are important parameters for evaluating nutritional status [27–29] and are indicative of neonates and infants who are well, or poorly developed or nourished [30]. In this study, there were no differences in the post-home rectal irrigation weight, body length, and serum albumin level between group A and the healthy control group, which suggested that home rectal irrigation during the neonatal period in patients with HSCR is effective and does not impair general growth and development. The serum prealbumin and retinol-binding protein levels, however, are also important indices for evaluating nutritional status [31, 32], and can indicate the immediate nutritional status, which may be attributed to a short half-life. In the current study, the post-home rectal irrigation serum prealbumin and retinal binding protein levels in group A were significantly lower than the healthy control group, which was interesting and indicated that the nutritional status may begin to deteriorate and might accelerate as home rectal irrigation continues. Thus, home rectal irrigation should not be continued indefinitely, and based on the results of the current study, we suggest that the duration of home rectal irrigation should be 2–4 months.

HAEC is a common and sometimes life-threatening complication of HSCR [33]. The pathogenesis of HAEC is multifaceted, involving stool retention, more proximal extension of aganglionosis, genetic variations, and disorders of the intestinal microbiota, mucus barrier, and immune system [34–36]. Less effective home rectal irrigation would result in stool retention and HAEC. In contrast, effective home rectal irrigation can eliminate the retained stool in the colon. It has been reported that rectal washout or home rectal irrigation can reduce the rate of HAEC and lead to an improved outcome [37]. Hence, HAEC may be an important index for evaluating the efficacy of home rectal irrigation during the neonatal period for HSCR. In the current study, HAEC occurred in five patients in group A during the period of home rectal irrigation, which was very low and indicated that home rectal irrigation during the neonatal period for patients with HSCR is effective and could wash out the stool in the colon.

Home rectal irrigation facilitated definitive PT surgery

Avoid misdiagnosis during the neonatal period or early in infancy

The diagnostic methods for HSCR consist of barium enema, anorectal manometry, and biopsy pathologic evaluation, among which rectal biopsy is the most effective method for confirming the diagnosis of HSCR [38, 39]. In the current study, all patients had two rectal biopsies, and six patients who were diagnosed with HSCR by rectal biopsy during the

neonatal period were excluded from HSCR because ganglion cells were demonstrated in the second rectal biopsy after the neonatal period. The reason for misdiagnosis based on rectal biopsy during the neonatal period is often multifactorial; tissue yield and a false-negative interpretation of a single rectal biopsy may play important roles. Due to the risk of causing intestinal perforation in neonates, an adequate amount of tissue is difficult to obtain and may not be obtained on rectal biopsy, thus, a single rectal biopsy may also account for false-negative interpretations. Keyzer-Dekker et al. [40] reported that the sensitivity of a single rectal biopsy is 81% and the specificity is 97%. Thus, diagnosis of HSCR during the neonatal period based on biopsy may not be highly accurate and should be repeated for confirmation after the neonatal period. Therefore, misdiagnoses could be reduced by repeated biopsies after the neonatal period, which was based on performing home rectal irrigation during the neonatal period to wash out retained stool.

Benefit of home rectal irrigation to definitive PT

Pull-through is the most common surgical procedure for correction of HSCR, and usually need laparoscopic assistance [3, 4]. Neonates and young infants always have a lower weight and smaller abdominal space than older infants, which may increase the difficulties with laparoscopic-assisted surgical procedures. Furthermore, decreased abdominal space and tolerance to a pneumoperitoneum [41, 42] increases the risk of injury to adjacent normal tissues when laparoscopic-assisted PT is performed. Home rectal irrigation can delay the timing of PT from the neonatal period or early in infancy to later in infancy, with increased abdominal space due to growth and development. In the current study, the average operative weight was 6.77 kg, and the abdominal space would be sufficient for laparoscopic-assisted PT. Anastomotic strictures and leakage are severe complications after PT, which is frequently caused by infection [2, 11]. Indeed, the absence of anastomotic strictures and leakage after definitive PT may result from robust immunity and tolerance to infection and surgery in older infants. With growth and development, active immunity and tissue tolerance in is better in older infants than neonates or young infants [43] and decreases the risk of infection. Perianal excoriation is usually accompanied by high stool frequency [44]. In the current study, by performing PT in older infants the average stool frequency was 3.9 times per day, which was accompanied with a lower incidence of perianal excoriation than performing PT during the neonatal period, as in a previous study [9]. At the 6-month follow-up evaluation, there was no constipation and a low incidence and mild degree of severity of post-operative HAEC. Seven post-operative grade I HAEC patients were managed by outpatient care and 4 Grade II patients were treated in the

hospital. In our last study, the incidence of post-operative HAEC was high when performing PT during the neonatal period. A delay in PT by including home rectal irrigation in the management scheme from the neonatal period or early in infancy to older infants may be beneficial to decrease the difficulty of a definitive PT procedure.

Conclusions

Home rectal irrigation during the neonatal period or early in infancy followed by a delayed and planned SSPT is feasible and effective in patients with HSCR that could be beneficial to definitive PT. However, for patients with an extended aganglionic segment, older age, or a high HAEC score at the time of diagnosis of HSCR, home rectal irrigation may not be suitable.

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Compliance with ethical standards

Conflict of interest The authors declare no conflicts of interest.

Ethical approval All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki Declaration and its later amendments or comparable ethical standards.

Informed consent Informed consent was obtained from the parents of all participants included in the study.

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