

Functional Outcome in Children with Hirschsprung's Disease after Modified Duhamel's Procedure: A Longitudinal Study

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ABSTRACT

Introduction: The classical surgical treatment of Hirschsprung's Disease (HD) is performed by resection of the aganglionic segment of the distal intestine and bringing down the ganglionic segment by Modified Duhamel's procedure or other pull-through operations. The assessment of the postoperative functional outcome and continence scoring systems are important considerations during follow-up of these children. In the present study, a scoring system was used to analyse the postoperative functional outcome of HD as there are very few studies, that have objectively assessed the same.

Aim: To assess the functional outcome in children under six years of age, who underwent Modified Duhamel's procedure for HD.

Materials and Methods: This longitudinal study was conducted in the Tertiary Teaching Institution at Government Medical College, Thiruvananthapuram, Kerala, India. The duration of the study was one year and nine months, from February 2017 to November 2018. Forty cases of biopsy proven HD, who underwent Modified Duhamel's procedure were analysed. Postoperative follow-up was done at one, two, and three years after the definitive surgical procedure. Patients were classified as good, fair and

poor outcome groups based on the Taylor-Duthies-Zachary continence scoring system. Statistical analysis was performed using Statistical Package for Social Sciences (SPSS) software version 22.0. Pretest comparison of scores and p-value were analysed by Wilcoxon signed-rank test. A p-value <0.05 was considered as statistically significant.

Results: The mean age of the study participants was 12.7±4.1 months and age ranges from three to 25 months. Median stool frequency per day after one, two, and three years of surgery, was 6.0, 4.0, and 3.0, respectively. The observed difference in median frequency between one and two year, one and three, and two year and three year was statistically significant (p<0.05). Median continence score to assess the anal sphincter control at one, two and three years after surgery were 2.3, 4.0 and 4.5, respectively. The observed difference in scores over successive years, was statistically significant (p<0.05).

Conclusion: The functional outcome in patients, who underwent Duhamel's procedure was good and the postoperative issues like increased stool frequency, and sphincter control improved over successive years.

Keywords: Aganglionosis, Colonic, Congenital, Megacolon

INTRODUCTION

The HD is a developmental disorder of the intrinsic component of the enteric nervous system, that is characterised by the absence of ganglion cells in the myenteric and submucosal plexus of the distal intestine presenting with functional intestinal obstruction at the level of aganglionosis [1,2]. The incidence is approximately one in 5000 live births [3,4]. Histologic evaluation of rectal biopsy remains the gold standard diagnostic test. The classical treatment is surgical resection of the aganglionic segment of the distal intestine and bringing down the ganglionic segment to the anus by Duhamel's (modified) procedure or other pull-through operations [2,3]. The other surgical procedures for HD, include the Soave endorectal pull-through and Swenson's rectosigmoidectomy. Primary transanal endorectal pull-through and laparoscopic abdominal mobilisation for HD have gained popularity in the last few decades. The postoperative issues associated with endorectal pull-through include cuff abscess, cuff roll-back, and difficulty to adapt long segment HD [3-5].

The main complication associated with Swenson's procedure is the neurogenic bladder, due to extensive perirectal dissection. The postoperative problems in children seen after Modified Duhamel's operation include obstructive symptoms, soiling, and enterocolitis. The assessment of postoperative functional outcome after Duhamel's procedure and continence scoring systems are important considerations during follow-up of these children. Many existing

studies on the postoperative outcome of HD have not assessed the outcome based on objective criteria [5]. In the absence of a specific scoring system to assess the postoperative outcome in HD, objective evaluation of the results become difficult. The aim of the study was to assess the functional outcome in children under six years of age, who underwent Modified Duhamel's procedure for HD at a Tertiary Teaching Institution.

MATERIALS AND METHODS

The longitudinal study was conducted in the Tertiary Teaching Institution at Government Medical College, Thiruvananthapuram, Kerala, India. The duration of the study was one year and nine months, from February 2017 to November 2018. The study included cases, which had completed one, two and three years of postoperative follow-up. Prior clearance was obtained from the Institutional Ethics Committee (IEC) letter number (IECNo.0/3/29/2017/MCT).

Inclusion criteria: Children under six years of age with biopsy-proven HD, who underwent Modified Duhamel's procedure. Patients with short-segment disease, long-segment disease, and total colonic aganglionosis (based on barium enema findings, intraoperative findings, and histopathology report) were included in the study.

Exclusion criteria: Children, who underwent procedures other than Duhamel's operation, children with chromosomal anomalies like Down's syndrome/metabolic disorders/neurogenic impairment,

hypothyroidism, and children with syndromic disorders were excluded from the study.

Study Procedure

A total of 40 cases of biopsy-proven HD who underwent Modified Duhamel's procedure were analysed for the functional outcome [4]. Postoperative follow-up were done at one, two, and three years of definitive surgical procedure. The parameters studied in each patient included basic demographic data and the analysis for daily frequency of stools, constipation and incontinence (as an index of the sphincter control). The data was collected from the outpatient and inpatient records, operative notes, postoperative records, follow-up charts and the questionnaire administered to parents during regular follow-up period. In the present study, the Taylor-Duthie-Zachary continence scoring system, to assess sphincter control in postoperative anorectal malformation has been used to objectively assess the functional status in postoperative HD during successive years [6]. Patients were classified to have good, fair, and poor outcome depending upon the score. The highest total score was 5. The scoring assigned was: Good (3-5), Fair (1.5-3.5), and Poor (0-1.5). A digital rectal examination was done at the end of the examination period to assess the anal sphincter tone, rule out rectal spur and constipation with the loading of rectum.

STATISTICAL ANALYSIS

Statistical analysis was performed using SPSS software version 22.0. Patient's continence scores were analysed and categorised into good/fair/poor outcome groups. Continuous variables were expressed as mean Standard Deviation (SD) and interquartile ranges. Categorical variables were expressed as frequency and percentage. Pretest comparison of scores and p-value were analysed by Wilcoxon signed-rank test. A p-value <0.05 was considered as statistically significant.

RESULTS

The average age of the study population was 12.7±4.1 months and age ranges from three to 25 months. A total of 18 out of the 40 patients were of age 9-12 months [Table/Fig-1]. Out of the total 40 patients, 28 (70%) patients were male and 12 (30%) were females [Table/Fig-2]. Average weight of the patients was 8.6±1.3 kg ranges from 5-12 kg. About 28 (70%) patients were in the 7-9 kg weight group [Table/Fig-3].

Age (in months)	Frequency (n)	Percentage (%)
<9	7	17.5
9-12	18	45.0
>12	15	37.5
Total	40	100

[Table/Fig-1]: Age distribution of patients.

Sex	Frequency (n)	Percentage (%)
Male	28	70
Female	12	30
Total	40	100

[Table/Fig-2]: Gender distribution of the subjects.

Weight (in kg)	Frequency (n)	Percentage (%)
<7	5	12.5
7-9	28	70
>9	7	17.5
Total	40	100

[Table/Fig-3]: Weight distribution of patients underwent Modified Duhamel's procedure for HD.

Two out of 40 (5%) patients have constipation after one year of surgery. Among the total of 40 patients studied, during successive years, only the cases with regular follow-up, complete follow-up data and filled questionnaires were included for analysis [Table/Fig-4]. The integrity of the anal sphincter and its postoperative state was assessed by digital rectal examination performed during follow-up. The functional status of the anal sphincter after surgery is an important determinant of the outcome. Out of 40 patients, 5 (12.5%) were noted to have lax anal sphincter and 35 (87.5%) were noted to have normal anal sphincter at the final per rectal examination. Two patients were found to have postoperative 'spur'. One patient underwent crushing of the spur and the other improved with conservative management. Median stool frequency per day after one, two and three years of follow-up was 6.0, 4.0 and 3.0, respectively. The observed difference in median frequency between after first year and second year, first year and third year and second year and third year was statistically significant (p<0.05). The incidence of postoperative soiling was found to be similar with an incidence of 5 (12.5%) patients. The stool frequency per day was significantly reduced in successive years [Table/Fig-5,6].

After one year		After two years	After three years
Constipation	n (%)	n (%)	n (%)
Yes	2 (5)	2 (5.6)	1 (3.3)
No	38 (95)	34 (94.4)	29 (96.7)
Total	40 (100)	36 (100)	30 (100)

[Table/Fig-4]: Incidence of postoperative constipation after Modified Duhamel's operation during follow-up at one year, two years and three years.

Stool frequency	Frequency (n)	Mean±SD	Median	Interquartile range
One year	39	6.2±1.6	6.0	5-7
Two years	34	4.2±0.9	4.0	4-5
Three years	24	2.7±0.6	3.0	2-3

[Table/Fig-5]: Postoperative incidence of stool frequency per day after Modified Duhamel's procedure during follow-up at one year, two years and three years. Stool frequency data was available for these patients only

Paired comparison	Wilcoxon signed-rank test	
	z-value	p-value
One year vs after two years	5.109	<0.001
One year vs after three years	4.243	<0.001
Two years vs after three years	4.132	<0.001

[Table/Fig-6]: Frequency of stools during postoperative period after Modified Duhamel's procedure; comparison of incidence at follow-up and statistical significance.

Median Taylor- Duthies- Zachary continence scores during postoperative follow-up at one year, two years, and three years were 2.3, 4.0 and 4.5, respectively. The observed difference in scores in successive years was statistically significant (p<0.05). There was a significant improvement in score after successive years [Table/Fig-7,8].

Continence score	n	Mean±SD	Median	Interquartile range
One year	40	2.3±0.6	2.3	2.0-2.5
Two years	36	4.00±0.7	4.0	3.5-4.5
Three years	24	4.6±0.4	4.5	4.5-5.0

[Table/Fig-7]: Taylor-Duthies Zachary continence score after Modified Duhamel's operation; comparison of score during follow-up at one year, two years and three years.

The outcome in 7 (17.5%) patients was poor according to continence score after one year. But during follow-up, only 1 (2.8%) patient was seen to have a poor outcome after two years and no case was observed to have poor outcome after three years [Table/Fig-9].

Paired comparison	Wilcoxon signed-rank test	
	z-value	p-value
One year vs after two years	5.263	<0.001
One year vs after three years	4.359	<0.001
Two years vs after three years	3.218	<0.001

[Table/Fig-8]: Taylor-Duthies Zachary continence score during postoperative follow-up; comparison of statistical significance of the change in score during successive years.

Continence score (Taylor-Duthies Zachary score)	After one year	After two years	After three years
	n (%)	n (%)	n (%)
Poor	7 (17.5)	1 (2.8)	0
Fair	33 (82.5)	10 (27.8)	0
Good	0	25 (69.4)	24 (66.7)
Total	40 (100)	36 (100)	24 (66.7)

[Table/Fig-9]: Taylor-Duthies Zachary continence score; the grading of outcome during postoperative follow-up as poor, fair and good.

DISCUSSION

Duhamel's procedure involves bringing the normal colon down through the bloodless plane between the rectum and the sacrum and joining the two walls to create a new lumen, which is aganglionic anteriorly and normally innervated posteriorly. Kocher clamp was used to join the walls initially and later Irani clamps were used to join the walls and left there till it falls off naturally, usually within a week. Presently, surgical staplers are almost always used instead [1,2]. The incidence of postoperative constipation was 5% in the first year, 5.6% in the second year and 3.3% after three years of postoperative period. Two patients were found to have postoperative 'spur'. One patient underwent crushing of the spur and the other improved with conservative management. Various previous studies shows that, the incidence of constipation is less in Duhamel's procedure. The incidence of constipation in various studies ranged from 4% to 13.4% [7-9]. The findings in the present study was associated with the previous studies. There was a steep decline in the incidence of frequency of stools, as the age advances. Six patients developed acute diarrhoeal disease and one patient developed enterocolitis, which were managed conservatively. Median stool frequency per day after one year, two year, three year of surgery was six, four and three, respectively and this finding has significant bearing on outcome. The incidence of stool frequency in various studies ranged from three to seven per day. These findings are correlated with the previous studies [9,10].

The continence score for the functional integrity of the anal sphincter was assessed. The score at one, two and three years after surgery was 2.3, 4 and 4.5, respectively. There is a significant improvement in score and it showed that, there was a good sphincter control over time. The findings of the present study also associated with previous studies [10,11]. In the present study, on digital rectal examination, 35 (87.5%) patients had a normal anal sphincter and 5 (12.5%) patients had lax anal sphincter. Functional improvement in the sphincter, occurred over the time and associated with previous studies [1,2,12-14]. The present study was compared with previous similar studies by Widyasari A et al., Aravind KL et al., and Shah S et al., as shown in [Table/Fig-10] on the functional outcome of HD after Duhamel's procedure [12-14]. The incidence of postoperative soiling was found to be similar with an incidence of 5/40 (12.5%) which is comparable. The incidence of constipation was found to be lesser than in most of the studies. The postoperative outcome was noted to be good/fair with normal continence in upto 38/40 (97.3%), which is higher than other studies.

Study details and parameters studied	Widyasari A et al., [12]	Aravind KL et al., [13]	Shah S et al., [14]	Present study
Age	<18 years	Three days to five years	One to nine years	Three to 25 months
Place of study	University Teaching Hospital, Yogyakarta, Indonesia	Mysore Medical College, India	LHMC, KSCH, New Delhi, India	SATH, GMC, Thiruvananthapuram, Kerala, India
Number of cases of Duhamel's procedure	28	17	30	40
Incidence of soiling (%)	18	-	10	12.5
Soiling frequency (%)	21	29.4	-	17.5
Constipation frequency (%)	4	11.76	13.4	3.3 to 5.6
Continence/voluntary bowel movements (%)	93	-	83.3	97.2

[Table/Fig-10]: Comparison of the present study with similar studies on the functional outcome of HD after Duhamel's procedure.

Limitation(s)

The relatively short follow-up period and the use of continence scoring system adapted from anorectal malformations were limitations of the study. Ideally, long term prospective studies using continence scoring systems specific to HD, are required for the assessment of postoperative functional outcome.

CONCLUSION(S)

The objective assessment of functional outcomes after surgery for HD using a continence scoring system gives valuable information regarding the status of bowel function. The majority of children were observed to have a good functional outcome with a low incidence of long-term postoperative complications. The continence score and bowel habits improved in successive years, as there was improvement in sphincter control.

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