

# Comparison of Long-term Outcomes of Hirschsprung Disease Patients after Soave and Duhamel Surgery based on Pediatric Incontinence and Constipation Scoring System : Retrospective Cohort Study

## Abstract

**Introduction:** Hirschsprung's disease is a congenital disease that causes intestinal motility disorders. Hirschsprung's disease is still the most common condition that causes intestinal obstruction in children. The incidence of Hirschsprung's disease is 1:5000 live births. The incidence ratio of this disease is around 4:1 between men and women, with a higher percentage of men<sup>1,3,4</sup>. Approximately 75% of cases of Hirschsprung's disease occur in the rectosigmoid colon area, 17% in the splenic flexure or transverse colon and 8% along the colon and terminal ileum. The Duhamel technique still remains a popular procedure most surgeons especially for long segment treatment and redo procedures.<sup>28,30</sup> The advantage of the Duhamel Procedure is that it is easy to perform, reduces the risk of damage to surrounding structures such as genitourinary innervation, but has the possibility of complications of anastomotic leakage which causes fistulas and abscesses.

**Materials and Methods:** Total subjects of this study was 441 patients with Hirschsprung's disease who were treated with the Soave procedure and the Duhamel procedure at Dr. Moewardi General Hospital in the period January 2017 - January 2022. This type of research is descriptive analytics with a retrospective cohort.

**Results:** The results of the study can be concluded that patients with HAEC increases the risk of constipation. Congenital abnormalities predispose to fecal incontinence. Differences in surgical techniques do not significantly influence or increase the risk of constipation and fecal incontinence based on statistical tests. However, the increased risk of constipation and fecal incontinence is higher in patients undergoing the Duhamel surgical procedure.

**Conclusion:** Conclusions that can be drawn from research regarding the comparison of long-term outcomes of Hirschsprung disease patients after Soave and Duhamel operations based on the Pediatric Incontinence and Constipation Scoring System at Dr. Moewardi General Hospital for period January 2017 – January 2022, it can be concluded that there is no significant difference in the outcome of patients who underwent the Soave or Duhamel

procedures. The Duhamel procedure has a higher prevalence of complications of constipation and fecal incontinence compared to the Soave procedure. The presence of HAEC before the surgical procedure increases the risk of constipation, significantly in the Soave and Duhamel procedures.

**Keywords:** hirschprung's disease, soave procedure, duhamel procedure, incontinence, constipation

## Introduction

Hirschprung's disease is a congenital disease that causes intestinal motility disorders. Hirschprung's disease is still the most common condition that causes intestinal obstruction in children. The incidence of Hirschprung's disease is 1:5000 live births.<sup>4, 5, 6</sup> The incidence ratio of this disease is around 4:1 between men and women, with a higher percentage of men.<sup>18, 19</sup> Approximately 75% of cases of Hirschprung's disease occur in the rectosigmoid colon area, 17% in the splenic flexure or transverse colon and 8% along the colon and terminal ileum.<sup>20, 23, 29</sup>

After making a diagnosis through history taking, physical examination and supporting examinations, patients with Hirschprung's disease must be given immediate treatment.<sup>28, 31, 32</sup> The principle of action in Hirschprung's disease is to resect the aganglionic intestine and pull the ganglionic intestine through a point just above the dentate line. There are several pull-through procedures for Hirschprung's disease such as Swenson, Soave, Duhamel and Rehbein with varying functional results.<sup>25, 26, 33</sup> Each procedure has its own advantages and disadvantages.

The Duhamel technique still remains a popular procedure most surgeons especially for long segment treatment and redo procedures.<sup>27, 29, 30</sup> The advantage of the Duhamel Procedure is that it is easy to perform, reduces the risk of damage to surrounding structures such as genitourinary innervation, but has the possibility of complications of anastomotic leakage which causes fistulas and abscesses.<sup>1, 4, 5</sup>

The principle of the Soave procedure is to make a circular cut through muscle layer of the large intestine in the pelvic peritoneal reflection.<sup>18, 20</sup> The advantage of this procedure is that it has a lower risk of pelvic dissection of intra-abdominal organs, such as bleeding, injury to other organs and the formation of adhesions. However, this procedure can cause functional obstruction due to the remaining seromuscular layer in the ganglionic segment.<sup>11, 12, 22</sup>

Although currently, the Duhamel procedure is one of the most is widely used to treat Hirschsprung's disease, but there is still some debate regarding which technique offers the best results.<sup>15, 16, 17</sup> The absence of incontinence and constipation after the procedure is the most important indicator in determining which procedure is better.<sup>2, 30</sup> This study aims to determine the long-term outcomes of Hirschsprung patients after Duhamel and Soave surgery at Dr. Moewardi General Hospital.

## Materials and Methods

Total subjects of this study was 441 patients with Hirschsprung's disease who were treated with the Soave procedure and the Duhamel procedure at Dr. Moewardi General Hospital in the period January 2017 - January 2022. This type of research is descriptive analytics with a retrospective cohort in patients with Hirschsprung's disease who were treated with the Soave procedure and the Duhamel procedure at Dr. Moewardi General Hospital.

The sampling technique in this research is purposive sampling, namely by determining subjects who meet the inclusion criteria and then including them in the research within a certain period of time so that the number of respondents can be met. Inclusion criteria in this study: 1. Patients with Hirschsprung's disease who were treated with the Soave procedure and the Duhamel procedure. 2. Patients who underwent surgery in the period January 2017 – January 2022.

The samples that will be taken are all Hirschsprung patients managed with the Soave and Duhamel procedure at Dr. Moewardi Hospital in the period January 2017 – January 2022 who met the inclusion and exclusion criteria.

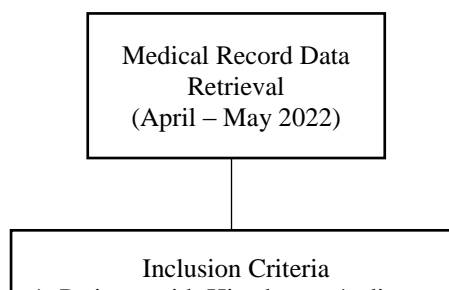
$$n = \left\{ \frac{Z\alpha + Z\beta}{0.5 \ln \left[ \frac{(1+r)}{(1-r)} \right]} \right\}^2 + 3$$

$$n = \left\{ \frac{1.65 + 0.842}{0.5 \ln \left[ \frac{(1+0.4)}{(1-0.4)} \right]} \right\}^2 + 3$$

For the minimum total sample , the formula is used Dahlan (2016)

n : 38 subjects (19 respondents for each group)

Information: n: number of subjects; Z : standard alpha value; Z : standard beta value; r: minimum relation coefficient.



UNDER PEER REVIEW

***List 1 : Paediatric Incontinence and Constipation Scoring System (PICSS)***

<b>Incontinence / Constipation Paediatric Score</b>			<b>Fecal Incontinence</b>			<b>Constipation</b>		
Does your child wear diapers at night?								
Always	Sometimes	No	0	2.5	5	-	-	-

Does your child wear diapers during the day?	0	2.5	5	-	-	-
Always	Sometimes	No				
How often your child defecates?	0	2.5	5			
Several times a day	1 time a day	Seldom				
What is the consistency of the stool?	0	2	4	1	0.5	0
Liquid	Variety	Solid				
Can your child hold back when he wants to defecate?	5	2.5	0	-	-	-
Yes, always	Sometimes	No				
Can your child differentiate between the urge to fart and defecate?	4	2	0	1	0.5	0
Yes, always	Sometimes	No				
Does your child often accidentally defecate in his underwear?	-	-	-	0	1	2
Yes, always	Sometimes	No				
Does your child often have incomplete emptying?	-	-	-	0	1.5	3
Yes, always	Sometimes	No				
Does your child feel pain when defecating?	2	1	0	0	2	4
Yes, always	Sometimes	No				
Does your child have to strain when defecating?	2	1	0	0	2	4
Yes	Normal	No				
Does your child fart a lot?	-	-	-	0	1	2
Yes, always	Sometimes	Never				
Does your child have difficult defecating?	-	-	-	0	2	4
Always	Sometimes	Never				
Does your child have abdominal pain?	-	-	-	0	2	4
Always	Sometimes	Never				
Total Score						
Chronic laxative use?	Yes			No		

## Results

### Characteristics of Research Subjects

General characteristics of research subjects are shown in **Table1.** including age, gender, prematurity status, congenital abnormalities, HAEC (Hirschprung Associated Enterocolitis), and patient outcomes (PICSS score, constipation, fecal incontinence) in each study group according to the surgical procedure performed.

**Table1. General Characteristics of Research Subjects**

Characteristics	Procedure	
	Soave	Duhamel
Age (mean)	3,2	8,37
Sex		

Man	14	13
Woman	6	6
<b>Prematurity</b>		
Yes	1	1
No	19	18
<b>Congenital Abnormalities</b>		
Yes	1	1
No	19	18
<b>Hirschprung Associated Enterocolitis</b>		
Yes	1	3
No	19	16
<b>PICSS Score (mean)</b>	20,9	20,89
<b>Constipation</b>	3	6
<b>Fecal Incontinence</b>	4	6
<b>Without Complications</b>	13	7

**Table 1.** shows differences in age trends in Soave and Duhamel surgical procedures. In younger patients, the dominant procedures of choice are Soave and Duhamel in older patients. Therefore, to facilitate analysis in the next analysis, age was grouped into two categories, namely <6 months and >6 months. Regarding the gender of Hirschsprung disease patients, the prevalence is more male. Outcomes of constipation and fecal incontinence were measured using the PICSS score. From the table it can be seen that there are similar mean scores for the Soave and Duhamel procedures.

More specific characteristics of research subjects are shown in **Table2**. This table explains, in particular, several factors of research subjects on post-operative outcomes, namely the presence of complications in the form of constipation and fecal incontinence.

**Table 2. Characteristics of Differences in Surgical Techniques on Post-operative Outcomes**

Variable		Constipation	Fecal Incontinence	Normal Value
<6 months	Soave	2	4	6
	Duhamel	-	-	4

≥6 months	Soave	1	-	7
	Duhamel	6	6	3
Woman	Soave	1	1	4
	Duhamel	2	1	3
Man	Soave	2	3	9
	Duhamel	4	5	4
Prematurity	Soave	1	-	-
	Duhamel	-	-	1
Congenital Abnormalities	Soave	-	1	-
	Duhamel	-	1	-
HAEC	Soave	1	-	-
	Duhamel	2	-	1

**Table 2.** shows that younger patients in the Soave procedure are more likely to experience incontinence. Meanwhile, in the Duhamel procedure, the same prevalence of complications of constipation and fecal incontinence was found. Men experienced more complications than women in this study. Then, babies with congenital abnormalities tend to experience incontinence rather than constipation. Meanwhile, patients with HAEC tend to experience constipation rather than incontinence.

### Bivariate Analysis

The data was then analyzed using statistical tests to determine the significance value. **Table 3.** displays the results of the bivariate chi-square analysis test.

**Table 3. Chi-square Test Results**

Variable	Constipation		Fecal Incontinence	
	OR	P	OR	P
Age	4.000	0.103	1.400	0.468
Surgical Techniques	2.615	0.199	1.846	0.323
Sex	1.167	0.576	0.475	0.332
Prematurity	3.625	0.413	0.000	0.548
Congenital Abnormalities	0.000	0.587	Undefined	0.061
HAEC	14.500	0.032	0.000	0.289

The results of the study showed a significant comparison in the HAEC variable, where patients with HAEC had a 14.5 times risk of causing constipation compared to patients without HAEC. For other research results, the results were not statistically significant ( $p>0.05$ ). The table above also shows that prematurity and HAEC do not increase the risk of fecal incontinence compared to patients who are not premature and without HAEC.

The age variable which was divided into age <6 months and >6 months showed results that were not statistically significant, but from the odds ratio it could be concluded that age >6 months increased the risk of constipation 4 times and 1.4 times for fecal incontinence compared to <6 months of age.

In the surgical technique variable by comparing Soave and Duhamel, it was found that patients who underwent Duhamel surgery had a 2.6 times risk of experiencing constipation and a 1.8 times risk of experiencing fecal incontinence. However, the significance test is not statistically significant.

### Multivariate Analysis

The variables were tested using logistic regression for multivariate analysis. The results of the logistic regression statistical test are displayed in **Table 4.** and **Table 5.**

**Table 4. Logistic Regression Test**

Variable	P
Constipation	0,120
Fecal Incontinence	0,056

**Table 5. Partial Test**

Variable	P	
	Constipation	Fecal Incontinence
Age	0.249	0.269
Surgical Techniques	0.423	0.198
Sex	0.541	0.167
Prematurity	0.171	0.993
Congenital Abnormalities	0.340	0.008
HAEC	0.011	0.116

**Table 4.** shows the results of whether the independent variables generally influence the dependent variable. Obtained  $p > 0.05$  so that in general the independent variable does not affect the dependent variable. Then, the partial test results obtained in **Table 5.** show that HAEC significantly influences the incidence of constipation and congenital abnormalities most influence the incidence of fecal incontinence. For other variables, including surgical technique,  $p > 0.05$  was obtained so that this variable did not influence/correlate with the dependent variable.

## Conclusion

Based on the results of data analysis and discussion, the author obtained. Conclusions that can be drawn from research regarding the comparison of long-term outcomes of Hirschsprung disease patients after Soave and Duhamel operations based on the Pediatric Incontinence and Constipation Scoring System at Dr. Moewardi General Hospital for period January 2017 – January 2022, it can be concluded that there is no significant difference in the outcome of patients who underwent the Soave or Duhamel procedures. The Duhamel procedure has a higher prevalence of complications of constipation and fecal incontinence compared to the Soave procedure. The presence of HAEC before the surgical procedure increases the risk of constipation, significantly in the Soave and Duhamel procedures.

## References

1. Adhi, M. *et al.* (2012) 'Duhamel ' s Procedure for Adult Hirschsprung ' s Disease Duhamel ' s Procedure for Adult Hirschsprung ' s Disease', 22(218), pp. 395–398.
2. Aworanti, O. M. *et al.* (2012) 'Comparative review of functional outcomes post surgery for Hirschsprung's disease utilizing the paediatric incontinence and constipation scoring system', *Pediatric Surgery International*, 28(11), pp. 1071–1078. doi: 10.1007/s00383- 012-3170-y.
3. Bradnock, T. J. and Walker, G. M. (2013) 'Open Duhamel Pull-Through', in *Basic Techniques in Pediatric Surgery*. Butler Tjaden, N. E. and Trainor, P. A. (2013) 'The developmental etiology and pathogenesis of Hirschsprung disease', *Translational Research*. Mosby, Inc., 162(1), pp. 1–15. doi: 10.1016/j.trsl.2013.03.001.
4. Cheng, Lily S., Dana M. Schwartz, Ryo Hotta, Hannah K. Graham, and Allan M. Goldstein. "Bowel Dysfunction following Pullthrough Surgery Is Associated with an

- Overabundance of Nitrergic Neurons in Hirschsprung Disease." *Journal of Pediatric Surgery* 51, no. 11 (2016): 1834. Accessed September 2, 2023. <https://doi.org/10.1016/j.jpedsurg.2016.08.001>.
5. Chhabra, S. and Kenny, S. E. (2016) 'Hirschsprung's disease', *Surgery (United Kingdom)*. Elsevier Ltd, 34(12), pp. 628–632. doi: 10.1016/j.mpsur.2016.10.002.
  6. Chistoper P., C. (2014) 'Pediatric Surgery: Diagnosis and Treatment', in.
  7. Demehri, F. R. *et al.* (2013) 'Hirschsprung-associated enterocolitis: Pathogenesis, treatment and prevention', *Pediatric Surgery International*, 29(9), pp. 873–881. doi: 10.1007/s00383-013-3353-1.
  8. Elsherbeny, M. and Abdelhay, S. (2019) 'Obstructive complications after pull-through for Hirschsprung's disease: different causes and tailored management', *Annals of Pediatric Surgery*. *Annals of Pediatric Surgery*, 15(1), pp. 0–4. doi: 10.1186/s43159-019-0003-y.
  9. Frykman, P. K. and Scott S. Short (2012) 'Hirschsprung-Associated Enterocolitis: Prevention and Therapy', *Seminars in Pediatric Surgery*, 21(4), pp. 328–335.
  10. Gao, Tingting, Weijue Xu, Qingfeng Sheng, Ting Xu, Wei Wu, and Zhibao Lv. "Clinical Outcomes and Risk Factors for Postoperative Complications in Children with Hirschsprung's Disease." *American Journal of Translational Research* 14, no. 7 (2022): 4830-4837. Accessed September 2, 2023. <https://www.ncbi.nlm.nih.gov/pmc/articles/PMC9360846/>.
  11. George W, H. I. (2014) *Ashcraft's Pediatric Surgery 6th edition*. Elsevier.
  12. Gershon EM, Rodriguez L, Arbizu RA. Hirschsprung's disease associated enterocolitis: A comprehensive review. *World J Clin Pediatr* 2023; 12(3): 68-76 [PMID: 37342453 DOI: 10.5409/wjcp.v12.i3.68]
  13. Hasserius, Johan, Josefine Hedbys, Christina Graneli, Kristine Hagelsteen, and Pernilla Stenström. "Treatment and Patient Reported Outcome in Children with Hirschsprung Disease and Concomitant Congenital Heart Disease." *BioMed Research International* 2017, (2017). Accessed September 2, 2023. <https://doi.org/10.1155/2017/1703483>.
  14. Joseph R. Davidson and others, Comparative cohort study of Duhamel and endorectal pull through for Hirschsprung's disease, *BJS Open*, Volume 6, Issue 1, February 2022, zrab143, <https://doi.org/10.1093/bjsopen/zrab143>
  15. Kathryn L., M. (2014) *Pathophysiology The Biologic Basis for Disease in Adults and Children 7th Edition*.

16. Kemenkes RI (2017) 'Pedoman Nasional Pelayanan Kedokteran Tata Laksana Penyakit Hirschsprung', *Keputusan Menteri Kesehatan RI*, Nomor HK.0.
17. Khazdouz, M. *et al.* (2015) 'Clinical outcome and bowel function after surgical treatment in Hirschsprungs disease', *African Journal of Paediatric Surgery*, 12(2), pp. 143–147. doi: 10.4103/0189-6725.160403.
18. Kyrklund, K. *et al.* (2020) 'ERNICA guidelines for the management of rectosigmoid Hirschsprung's disease', *Orphanet journal of rare diseases*. Orphanet Journal of Rare Diseases, 15(1), p. 164. doi: 10.1186/s13023-020-01362-3.
19. Langer, J. C. and Levitt, M. A. (2020) 'Hirschsprung Disease', *Current Treatment Options in Pediatrics*. Current Treatment Options in Pediatrics, 6(3), pp. 128–139. doi: 10.1007/s40746-020-00195-3.
20. Levitt, M. A. *et al.* (2009) 'Hirschsprung disease and fecal incontinence: diagnostic and management strategies', *Journal of Pediatric Surgery*. Elsevier Inc., 44(1), pp. 271–277. doi: 10.1016/j.jpedsurg.2008.10.053.
21. Łukasz, S. (2014) 'Diagnosis of Hirschsprung's disease with particular emphasis on histopathology. A systematic review of current literature', *Przegląd Gastroenterologiczny*, 9(5), pp. 264–269.
22. Mohajerzadeh, L. *et al.* (2015) 'Comparison Between Swenson and Soave Pull-Through in Hirschsprung Disease', *Annals of Colorectal Research*, 3(4). doi: 10.5812/acr.20746.
23. Mohamed, O. (2016) 'How to manage a late diagnosed Hirschsprung's disease', *African Journal of Paediatric Surgery*. doi: 10.4103/0189-6725.182562.
24. Moore, S. W. (2016) 'Hirschsprung disease: current perspectives', *Open Access Surgery*, p. 39. doi: 10.2147/oas.s81552.
25. Nabi, Z. *et al.* (2018) 'Diagnosis of Hirschsprung's disease in children: Preliminary evaluation of a novel endoscopic technique for rectal biopsy', *JGH Open*, 2(6), pp. 322– 326. doi: 10.1002/jgh3.12092.
26. NHS (2019) *Outcome Frameworks*, National Health Scotland. Available at: <http://www.healthscotland.com/ofhi/faq.html>.
27. Orvar, S. (2021) 'Hirschsprung's Disease: A Review', *AAP Publication*, 27710(5), pp. 1521–1523.
28. Parahita *et al.*, (2017). Comparison of Hirschsprung-associated enterocolitis following Soave and Duhamel procedures, *Journal of Pediatric Surgery*, Volume 53, Issue 7, P1351-1354, July 2017

29. Prem, P. (2011) *Pediatric Surgery: General Principles and Newborn Surgery* 1st ed 2020 Edition
30. Scholfield, D. W. and Ram, A. D. (2016) 'Laparoscopic duhamel procedure for hirschsprung's disease: Systematic review and meta-analysis' *Journal of Laparoendoscopic and Advanced Surgical Techniques*, 26(1), pp. 53–61. doi: 10.1089/lap.2015.0121.
31. Singh, S. (2017) "' The Laparoscopic Assisted Duhamel Pull through Procedure for Hirschsprung's Disease: Our Technique and Short Term Results '", *Journal of Pediatrics & Neonatal Care*, 7(2). doi: 10.15406/jpnc.2017.07.00281.
32. Wagner, J. P. (2014) 'Hirschsprung Disease Treatment & Management'. Available at: <http://emedicine.medscape.com/article/178493-treatment#a1128>.
33. Widyasari, A. *et al.* (2018) 'Functional outcomes in Hirschsprung disease patients after transabdominal Soave and Duhamel procedures', *BMC Gastroenterology*. *BMC Gastroenterology*, 18(1), pp. 4–9. doi: 10.1186/s12876-018-0783-1.
34. Zakaria, O. M., El Labban, G. M. and Shams, M. E. (2012) 'Fecal incontinence after single-stage Soave's pull-through: Abdominal versus transanal endorectal pull-through', *Annals of Pediatric Surgery*, 8(1), pp. 5–8. doi: 10.1097/01.XPS.0000407759.30719.57.