



# Quality of life of patients with Hirschsprung disease after Duhamel and Soave pull-through procedures: A mixed-methods sequential explanatory cohort study



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## ABSTRACT

**Background:** Quality of life (QoL) is one of the important outcomes for patients with Hirschsprung disease (HSCR) after pull-through that provides qualitative data concerning the long-term outcomes, however, it has not been well-studied. The HSCR/anorectal malformation quality of life questionnaire (HAQL) is considered valid and reliable to evaluate the QoL of HSCR patients.

**Material and methods:** A mixed-method sequential explanatory cohort study was conducted to compare the QoL of HSCR patients after Duhamel and Soave pull-through at Dr. Sardjito Hospital between 2013 and 2018 using an Indonesian adaptation of the HAQL, followed by a qualitative study.

**Results:** We ascertained eleven HSCR patients (Duhamel: five HAQL parents and one HAQL adolescent vs. Soave: four HAQL parents and one HAQL adult). For the quantitative study, the mean HAQL score was 2.50 and 2.79 for the Duhamel and Soave groups, respectively. For the qualitative study, interviewed patients' parents expressed how their child's life had improved after surgery. However, frequent bloating was a major complaint following Soave surgery, whereas hardened stools were a major problem after Duhamel procedure.

**Conclusion:** Here, for the first time using a mixed-method sequential explanatory cohort design, we show that patients with HSCR after Soave tended to have a higher overall QoL score compared to the Duhamel group. Further multicenter study with a larger sample size is mandatory to give better understanding about QoL of HSCR patients following pull-through.

## 1. Introduction

Hirschsprung disease (HSCR) is a complex genetic disorder defined as the absence of ganglion cells in the distal bowel extending proximally over a varying length of the intestines, resulting in functional obstruction in infants [1]. Its incidence is approximately 1 in 3250 to 5000 live births [1–3].

Surgical strategy for HSCR is to remove the aganglionic intestines and connect normal ganglionic intestines as distally as possible [4]. The most common procedures for HSCR are the Soave and Duhamel procedures [5–7].

Quality of life (QoL) is one of the important outcomes for patients

with HSCR after surgery [8]. The HSCR/anorectal malformation quality of life questionnaire (HAQL) is considered valid and reliable to evaluate the QoL of HSCR patients after definitive surgery in several languages [9–12]. We determined the QoL of HSCR patients after Duhamel and Soave procedures using an Indonesian adaptation of HAQL. An explanatory qualitative study was done afterwards to know how patients experience their condition within their daily life.

**Abbreviations:** HAQL, Hirschsprung's disease/anorectal malformation quality of life questionnaire; HSCR, Hirschsprung disease; QoL, quality of life

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## 2. Methods

### 2.1. Patient subjects

HSCR was diagnosed according to the clinical manifestations, contrast enema, and histopathological findings [2,5]. According to the current standard protocol, we utilized hematoxyline and eosin and S100 immunohistochemistry for the histopathological diagnosis of HSCR [2,5].

This cohort used a mixed-methods sequential explanatory research that consisted of a quantitative study followed by a qualitative study for patients with HSCR who underwent Duhamel and Soave procedures at Dr. Sardjito Hospital, Indonesia, from 2013 to 2018. During a 5-year period, 48 medical records of HSCR patients who underwent Duhamel (n = 20) and Soave (n = 28) pull-through at our institution were collected. One, 19 and 17 patients were excluded from this study because the patients' age was 1 year old, the parents' patients were unable to be contacted and they were not willing to join the study, respectively.

The research was approved by the Institutional Review Board of the Faculty of Medicine, Public Health and Nursing, Universitas Gadjah Mada/Dr. Sardjito Hospital (Ref: KE/FK/0791/EC/2018). Consecutive sampling was used to recruit participants for this study. After signing the informed consent form to participate, patients' parents were contacted by the attending physician either via phone calls or direct interview at the hospital during their visit to the outpatient clinic.

The work has been reported in line with the STROCSS criteria [13] and registered at Research Registry with UIN of researchregistry5218.

### 2.2. Soave and Duhamel procedures

Soave and Duhamel procedures were performed as previously reported [5–7]. Soave surgery involves removing the mucosa and submucosa of the rectum and pulling the ganglionic bowel down into the aganglionic muscular cuff of the rectum, followed by primary anastomosis above the dentate line [5,7]. Duhamel technique consists of resecting the aganglionic colon and bringing down the ganglionic colon via the retrorectal space, followed by colorectal anastomosis and elimination of the septum between the rectum and the ganglionic colon [5,6]. Soave and Duhamel procedures were chosen according to attending physician preference in our hospital.

### 2.3. The HSCR/Anorectal malformation quality of life questionnaire (HAQL)

We used the HAQL because it is already considered valid and reliable for evaluation of QoL of patients with HSCR in various populations using different languages [9–12].

In the original HAQL, the minimum age of a patient to participate is 6 years old [9]. We modified the minimum age of participants to be 3 years old for two reasons: 1) more patients could take part since there is a limited number of participants from our institution for this study, and 2) by the age of three, most children are already given toilet training by their parents [5]. According to a previous report, the HAQL is divided into the parents-reported questionnaire for children aged 3–11 years, adolescent reported questionnaire for ages 12–16 years and adult reported questionnaire for ages 17 years and above [9].

The HAQL for parents, adolescents and adults used in this study had a total of 36 items to be answered by patients. These items were divided into 8 dimensions which were: laxative diet, constipating diet, presence of diarrhea, fecal continence, urinary continence, social functioning, emotional functioning and body image and physical functioning [9,10]. A score from 0 to 3 was given in response for each item and a better QoL was indicated by a higher score. For each of the dimensions, the scores were computed as the sum of the item scores which was then divided by the number of items answered in that dimension [10]. The overall QoL

score was determined by calculating the total mean of the eight dimension scores for each type of surgery.

### 2.4. Qualitative study

Patient's parents from each type of surgery based on their availability were invited to participate. All interviews were conducted via telephone in Indonesian language to allow them to express their experiences with fewer communication barriers. The duration of the interview with the parents of patients who underwent Soave procedure was about 15 min while for the patients who underwent Duhamel procedure it was 13 min. Prior to the interview, the participants were briefed that their experiences after the surgery and contribution to the research may help other patients who have not completely recovered after the surgery to improve their QoL. Participants were asked open ended questions exploring their experiences related to their answers from the HAQL. The conversations were recorded for validation of the content. The interview was then translated from Indonesian to English and transcribed by the first author to be analyzed and used in the results of this study. The second author read through the transcript to identify meaning units related to the surgeries. Repeated patterns of how patients experience their daily life were developed from those content comparisons, including how the patients experienced life after surgery and what were the major problems faced after the pull-through procedure. Representative samples of the parents' quotations were selected to demonstrate the difference in the participants' responses.

## 3. Results

### 3.1. Baseline characteristics

We ascertained eleven HSCR patients (Duhamel: five HAQL parents and one HAQL adolescent vs. Soave: four HAQL parents and one HAQL adults) (Table 1).

### 3.2. Quantitative study

Here, we report the QoL of patients with HSCR after pull-through using an Indonesian adaptation of the HAQL. We found that the Soave group tended to have a higher overall score of HAQL than the Duhamel group (Table 2).

Analysis of individual dimensions showed that in some dimensions such as presence of diarrhea and urinary continence, the scores for Duhamel pull-through patients and Soave pull-through patients were similar (Table 2). This result showed that the patients from both surgeries experienced somewhat similar effects for items under both dimensions. There was a large difference in QoL score between the two

**Table 1**

Characteristics of HSCR subjects who underwent Duhamel and Soave pull-through procedures at Dr. Sardjito Hospital, Indonesia.

Characteristics	Duhamel (n)	Soave (n)
Age (years)		
- 3–11 (HAQL parents)	5	4
- 12–16 (HAQL adolescent)	1	0
- > 16 (HAQL adults)	0	1
Gender		
- Male	4	5
- Female	2	0
Duration of follow-up after surgery (years)		
- < 3		
- 3–5	3	0
- > 5	2	3
	1	2

HAQL, Hirschsprung's disease/anorectal malformation quality of life questionnaire.

**Table 2**  
Comparison of QoL between Duhamel and Soave pull-through patients based on each dimension using HAQL.

HAQL Dimensions	HAQL Scoring		
	Duhamel (n = 6) (mean ± SD)	Soave (n = 5) (mean ± SD)	Δ
Laxative diet	2.08	2.60	0.52
Constipating diet	2.50	3.00	0.50
Presence of diarrhea	2.58	2.60	0.02
Faecal continence	2.06	2.83	0.77
Urinary continence	2.96	3.00	0.04
Social functioning	2.81	3.00	0.19
Emotional functioning and body image	2.69	2.83	0.14
Physical functioning	2.30	2.43	0.13
Overall	2.50 ± 0.33	2.79 ± 0.22	0.29

Δ, HAQL scoring of Soave – HAQL scoring of Duhamel; HAQL, Hirschsprung's disease/anorectal malformation quality of life questionnaire; QoL, quality of life; SD, standard deviation.

procedures for dimensions such as fecal continence, constipating diet and laxative diet compared to the scores of other dimensions (Table 2). Moreover, for soiling frequency which comes under the fecal continence dimension and voluntary bowel movement which comes under the physical functioning dimension, the Soave procedure group also tended to have a better score compared to the Duhamel group (Table 2). However, due to the small sample size used in this study, the statistical significance in the differences between the scores of the two procedures cannot be accurately determined.

### 3.3. Qualitative study

Following the quantitative study, a qualitative study was conducted which addressed major questions based on the HAQL dimensions to further explain how patient's experience their new condition after surgery and how to cope some difficulties. To know the condition of both procedures, we selected one patient of each procedure. We decided to interview the parents of selected patients in order to generate rich information due to better communication ability. As a representative sample for the Soave procedure group, quotations from one interview of a parent of a 7-year-old-male patient were selected, while for the Duhamel procedure group, one parent of a 5-year-old-male patient was used to demonstrate the difference in content. Those patients had the duration of follow-up after surgery before the interview of 5 years and 1 month and 2 years and 4 months for Soave and Duhamel patients, respectively.

During the interviews, the parents of both patients expressed that they felt that their child's life had improved after the surgery. This was shared by most parents in the study but some differences were expressed. The major problems faced by the patient post Soave pull-through was their child remained thin despite eating in large quantities (good appetite) and had frequent bloating.

“... Do you know how big the portion of KFC rice is? Yes, that is still little for him ... although he is eating and drinking a lot, he has a problem in his large intestine. Surgery has been done a few times for him ... Sometimes the stomach is good and sometimes there is bloating too ... Sometimes, during bloating, his flatulence gives out a very strong stench ... If he is bloated, he has an explosive bowel movement.” (Mother of male Soave patient, 7 years old).

The parent of the Soave patient also mentioned that her child experienced fecal incontinence post-surgery very rarely.

“Waking up there are spots that look like feces. Yeah, like that but just a little. Even so, every day, at least once every 2 months or 3 months ...” (Mother of male Soave patient, 7 years old).

Additionally, the parent of the Soave patient shared that she relied

on home remedies to deal with problems in her child's daily life post-surgery. According to the parent, orange juice consumption usually relieves the hardened stools in her child. Besides that, to overcome her child's bloating stomach, the parent encourages her child to drink warm water first thing in the morning and positioned her child in a frog-like position to allow air from stomach to be released.

These experiences were different from that of the selected representative patient post Duhamel pull-through who reported hardened stools and poor appetite as the major problem.

“About defecating, you can't predict it. Not every day, maybe morning or afternoon or evening ... he defecates at least two days once and the feces is hard, he feels pain ... He doesn't want the food that children normally eat. He only eats rice, he doesn't want bread, doesn't want fruit. He doesn't want juice either. If he does want to try those things, he just wants a little.” (Mother of male Duhamel patient, 5 years old).

On the topic of fecal incontinence, the parent of the Duhamel patient said that at times, her child had experienced passing of feces in his diapers at night.

“At night, I still give diapers. Sometimes, at night while sleeping, he has a bowel movement in the diapers.” (Mother of male Duhamel patient, 5 years old).

## 4. Discussion

One advantage of QoL studies is providing qualitative data concerning the long-term outcomes of patients after a surgical procedure [8]. There are important effects on personal, social and professional spheres due to reoccurring constipation and/or fecal incontinence which will be reflected later during the patients' life [8]. QoL data involve subjective information consisting of several areas that should be assessed individually and according to the expectations of patients and their relatives [8].

Most studies compared the QoL of HSCR patients with healthy controls [14–19], while other studies focused on evaluation of one procedure for HSCR patients [20,21]. In one study, patients with HSCR had a lower psychosocial QoL scores, but not physical and total QoL scores, compared with healthy control children [14]. This difference was because the children with HSCR had poorer functional outcomes [14]. Interestingly, HSCR children scored lower QoL scores than the healthy children, whereas the adult patients showed significantly higher scores than their healthy counterparts [15]. This finding might be due to the adult patients having better coping approaches to face their symptoms than the children with HSCR [15]. Moreover, while patients showed poor functional outcomes, they showed a comparable overall QoL scores with healthy subjects [16,21].

To the best of our knowledge, our study is the first report that compared the QoL of patients with HSCR between two transabdominal surgeries, particularly the Soave and Duhamel procedures. One recent study compared the QoL between Duhamel and transanal Soave procedures and focused on the occurrence of failed pull-through [16]. We used the HAQL adapted into the Indonesian language for evaluation of QoL of our patients. This form might be useful for other populations with other Asian languages as well, besides those already translated [9–12]. Another novelty of our study is using a mixed-methods sequential explanatory design to evaluate the QoL of HSCR patients after surgery.

Our qualitative study found that the QoL of both groups of patients were affected in similar ways that were commonly expressed in the interviews. Interestingly, as frequently found in Asian countries, some patients rely on traditional home remedies to deal with problems in their daily lives post-surgery.

However, in addition to some parents' reticence in the interviews, our findings should be interpreted cautiously because of the small sample size. The main reason was because many patients' parents were not willing to participate in this study since they did not find their

participation in this study beneficial as their children's QoL had already improved after surgery.

## 5. Conclusions

Here, for the first time using a mixed-method sequential explanatory cohort design, we show that patients with HSCR after Soave tended to have a higher overall QoL score compared to the Duhamel group. Further multicenter study with a larger sample size is mandatory to give better understanding on QoL of HSCR patients after pull-through.

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The study was funded by the Indonesian Ministry of Research and Technology/National Agency for Research and Innovation.

## Ethical Approval

This study has been approved by the Ethical Committee of Faculty of Medicine, Public Health and Nursing, Universitas Gadjah Mada/Dr. Sardjito Hospital (Ref: KE/FK/0791/EC/2018).

## Author contribution

Michelle Raj Saysoo, Fatwa Sari Tetra Dewi and Gunadi conceived the study. Michelle Raj Saysoo conducted the study and drafted the manuscript, while Fatwa Sari Tetra Dewi and Gunadi critically revised the manuscript for important intellectual content. Gunadi facilitated all project-related tasks. All authors read and approved the final manuscript.

## Registration of Research Studies

Name of the registry: Research Registry  
 Unique Identifying number or registration ID: researchregistry5218  
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## Guarantor

Gunadi

## Consent

Written informed consent was obtained from the parents before joining the study. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

## Declaration of competing interest

No potential conflict of interest relevant to this article was reported.

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## Appendix A. Supplementary data

Supplementary data to this article can be found online at <https://doi.org/10.1016/j.amsu.2020.05.043>.

## Provenance and peer review

Not commissioned, externally peer reviewed.

## References

- [1] J.M. Tilghman, A.Y. Ling, T.N. Turner, et al., Molecular genetic anatomy and risk profile of Hirschsprung's disease, *N. Engl. J. Med.* 380 (2019) 1421–1432.
- [2] Iskandar K. Gunadi, A. Makhmudi, A. Kapoor, Combined genetic effects of RET and NRG1 susceptibility variants on multifactorial Hirschsprung disease in Indonesia, *J. Surg. Res.* 233 (2019) 96–99.
- [3] A. Kapoor, Q. Jiang, S. Chatterjee, P. Chakraborty, M.X. Sosa, C. Berrios, A. Chakravarti, Population variation in total genetic risk of Hirschsprung disease from common RET, SEMA3 and NRG1 susceptibility polymorphisms, *Hum. Mol. Genet.* 24 (2015) 2997–3003.
- [4] P.K. Tam, Hirschsprung's disease: a bridge for science and surgery, *J. Pediatr. Surg.* 51 (2016) 18–22.
- [5] A. Widayarsi, W. Pravitasari, A. Dwihtantoro, Gunadi, Functional outcomes in Hirschsprung disease patients after transabdominal Soave and Duhamel procedures, *BMC Gastroenterol.* 18 (2018) 56.
- [6] S.A. Nah, P. de Coppi, E.M. Kiely, et al., Duhamel pull-through for Hirschsprung disease: a comparison of open and laparoscopic techniques, *J. Pediatr. Surg.* 47 (2012) 308–312.
- [7] J.H. Lefèvre, Y. Parc, Soave procedure, *J. Vis. Surg.* 148 (2011) e262–e266.
- [8] A. Mathias, A. Tannuri, M. Ferreira, M. Santos, U. Tannuri, Validation of questionnaires to assess quality of life related to fecal incontinence in children with anorectal malformations and Hirschsprung's disease, *Rev. Paul Pediatr.* 34 (2016) 99–105.
- [9] M.J.G. Hanneman, M.A.G. Sprangers, E.L. De Mik, L.W.E. van Heurn, Z.J. De Langen, N. Looyard, et al., Quality of life in patients with anorectal malformation or Hirschsprung's disease development of a disease-specific questionnaire, *Dis. Colon Rectum* 44 (2001) 1650–1660.
- [10] C. Baayen, F. Feuillet, P. Clermidi, C. Crétolle, S. Sarnacki, G. Podevin, et al., Validation of the French versions of the Hirschsprung's disease and anorectal malformations Quality of Life (HAQL) questionnaires for adolescents and adults, *Health Qual. Life Outcome* 15 (2017) 24.
- [11] C. Grano, D. Aminoff, F. Lucidi, C. Violani, Disease-specific quality of life in children and adults with anorectal malformations, *Pediatr. Surg. Int.* 26 (2010) 151–155.
- [12] H. Wigander, B. Frenckner, T. Wester, M. Nisell, M. Öjmyr-Joelsson, Translation and cultural adaptation of the Hirschsprung's disease/anorectal malformation quality of life questionnaire (HAQL) into Swedish, *Pediatr. Surg. Int.* 30 (2014) 401–406.
- [13] R. Agha, A. Abdall-Razak, E. Crossley, N. Dowlut, C. Iosifidis, G. Mathew, For the STROCSS group. The STROCSS 2019 guideline: strengthening the reporting of cohort studies in surgery, *Int. J. Surg.* 72 (2019) 156–165.
- [14] L. Collins, B. Collis, M. Trajanovska, R. Khanal, J.M. Hutson, W.J. Teague, S.K. King, Quality of life outcomes in children with Hirschsprung disease, *J. Pediatr. Surg.* 52 (2017) 2006–2010.
- [15] R. Meinds, A. van der Steeg, C. Sloots, M. Witvliet, I. de Blaauw, W. van Gemert, M. Trzpis, P. Broens, Long-term functional outcomes and quality of life in patients with Hirschsprung's disease, *Br. J. Surg.* 106 (2019) 499–507.
- [16] O.G. Townley, R.M. Lindley, M.C. Cohen, G.V. Murthi, Functional outcome, quality of life, and 'failures' following pull-through surgery for Hirschsprung's disease: a review of practice at a single-center, *J. Pediatr. Surg.* 55 (2020) 273–277.
- [17] E.E. Hartman, F.J. Oort, D.C. Aronson, M.A. Sprangers, Quality of life and disease-specific functioning of patients with anorectal malformations or Hirschsprung's disease: a review, *Arch. Dis. Child.* 96 (2011) 398–406.
- [18] F. Drissi, G. Meurette, C. Baayen, et al., Long-term outcome of Hirschsprung disease: impact on quality of life and social condition at adult age, *Dis. Colon Rectum* 62 (2019) 727–732.
- [19] S. Sood, R. Lim, L. Collins, et al., The long-term quality of life outcomes in adolescents with Hirschsprung disease, *J. Pediatr. Surg.* 53 (2018) 2430–2434.
- [20] S. Onishi, K. Nakame, T. Kaji, et al., The bowel function and quality of life of Hirschsprung disease patients who have reached 18 years of age or older - the long-term outcomes after undergoing the transabdominal soave procedure, *J. Pediatr. Surg.* 52 (2017) 2001–2005.
- [21] M.I. Neuvonen, K. Kyrklund, R.J. Rintala, M.P. Pakarinen, Bowel function and quality of life after transanal endorectal pull-through for Hirschsprung disease: controlled outcomes up to adulthood, *Ann. Surg.* 265 (2017) 622–629.