

ORIGINAL ARTICLE

Prognosis of Hirschsprung's Disease After Pull Through Surgery in Arifin Achmad General Hospital of Riau Province

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ABSTRACT

Introduction: Hirschsprung's disease (HSCR) is a congenital abnormality which marked as an absence of ganglion cells in the distal intestine. The definitive treatment for HSCR is pull-through surgery. It has been reported that some pull-through resulted in more complications than others. The aim of this study is to analyze the type of pull-through surgery as a prognostic factor that may affect the presence of Hirschsprung-associated enterocolitis (HAEC) and nutritional status as outcomes of the surgery. **Methods:** A retrospective cross-sectional analytic study was conducted from medical record data at Arifin Achmad General Hospital of Riau Province archives. The population of this study was all the patients who diagnosed with HSCR after pull-through (trans-anal and trans-abdominal) and confirmed by colon biopsy in the Pediatric Surgery Division between January 2016 and December 2018 period. HAEC was assessed using Delphi analysis with cutoff score ≥ 10 and nutritional status was assessed using WHO Z-score. Fisher exact test was used with ($p \leq 0.05$) determine significance. **Results:** No significant correlation found between the method of pull-through with the incidence of HAEC and nutritional status ($p > 0.05$). However, only patients who underwent Duhamel pull-through showed the presence of HAEC (23.5%). Presence of HAEC significantly correlates with nutritional status (p -value 0.005, OR 6.0, 95% CI 2.136 - 16.857). **Conclusion:** Findings of this study showed the presence of HAEC and poor nutritional status were likely to occur in patient underwent Duhamel rather than Soave procedure. The presence of HAEC may be the key to poor nutrition status that disrupt gastrointestinal function.

Keywords: Hirschsprung's disease, Nutritional status, Pull-through, HAEC

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INTRODUCTION

Hirschsprung's disease (HSCR) is a developmental abnormality in the enteric nervous system, starting from the internal anal sphincter that extends to the proximal bowel (1). In the general population, HSCR occurs in 1:5000 live birth and more often seen in males than females with a ratio of 4:1 (1,2). The exact incidence of HSCR in Indonesia is unknown, but one recent study in Yogyakarta found the incidence of approximately 1:3250 newborns (3). Based on the study conducted at the Arifin Achmad General Hospital of Riau Province, Indonesia, there were 127 patients diagnosed with HSCR from 2010 until 2016 (4).

The Diagnosis of Hirschsprung is made based on clinical manifestations, colon biopsy, anal manometry, and abdominal radiograph (2,5). Pull-through surgery is

the definitive treatment for HSCR. Duhamel and Soave are the most common method that usually used in pull-through surgery (6).

The most common complication that occurs after surgical treatment in HSCR is *Hirschsprung-associated Enterocolitis* (HAEC), with a high mortality rate. The incidence rate for HAEC after surgery varies from 5% to 42% (7,8). The incidence of HAEC may be affected by the technique of pull-through performed by the surgeon. Some studies reported a higher rate of HAEC in Duhamel pull-through procedure (6). Another research reported a higher rate of complication (e.g., constipation) in the Soave procedure (9). Based on these two statements, it can be observed that there is still some inconsistency regarding the rate of complication in a certain pull-through procedure.

In HAEC, several occur gut disorders disturb the digestion, absorption, metabolism, and excretion process that will interfere with the nutritional status of the patient that further worsens the patient condition. The nutritional status can be assessed using the World

Health Organization (WHO) 2005 nutritional status chart (11).

Although there is much research that has evaluated the prognosis after pull-through surgery worldwide(6,7,12), there is no data regarding prognosis based on the occurrence of HAEC in Riau Province, Indonesia. The main aim of this study was to evaluate the prognosis of the patient with Hirschsprung Disease after pull-through surgery with Duhamel and Soave procedures based on the incidence of HAEC and nutritional status at the Arifin Achmad General Hospital of Riau Province which serves as tertiary health care center in Riau Province. In addition to prognosis, this study also aims to evaluate the presence of HAEC as a risk factor for poor nutritional status.

MATERIALS AND METHODS

This retrospective cross-sectional analytical study was done in February 2019 at the Arifin Achmad General Hospital of Riau Province. This study was approved by the Ethical Clearance Board of Medical Faculty of Riau University, with a Certificate of Ethical Appreciation number: 030/UN.19.5.1 .1.8/UEPKK /2019.

The population of this study was all of the patients in the Pediatric Surgery ward at Arifin Achmad General Hospital between January 2016 – December 2018 diagnosed with HSCR that treated with pull-through surgery either with Duhamel or Soave procedures. The diagnosis of Hirschsprung disease was made based on clinical manifestations, colon biopsy, anal manometry, and abdominal radiograph. Our source of data was medical record files retrieved from hospital archives. Only patients diagnosed by Pediatric surgeon or Pediatricians in the medical records with HSCR and confirmed by colon biopsy were included in this study.

HAEC was assessed by using the HAEC scoring system that was adapted from another study. The scoring includes 18 items that must be fulfilled by history taking, physical examination, radiologic examination, and laboratory tests (12). The nutritional status was assessed according to the subject’s age; the nutritional status was assessed by using a standardized WHO Z-score. We define a good nutritional status as Z score between -2 until 2, and poor nutritional status as Z score <-2 or >2 (11).

In our tertiary health center, we chose both Soave (trans-anal endorectal) and Duhamel (trans-abdominal retrorectal) pull-through operation as our definite operation based on the standard operating procedure in our hospital. In trans-anal (Soave) pull-through operation, all the patient underwent the operation as a one-step operation, that makes it is possible for all the children to get definitive operation before six months old. In trans-abdominal (Duhamel) pull-through operation, all the

patient must at least underwent colostomy procedure before the HSCR operation. After the operation, the presence of HAEC and nutritional status was assessed eight weeks after the operation. The reason for this approach was, we believe that after eight weeks, the operation wound will be fully healed and able to reflect the post-operation condition. To further reduce the chance of bias for all assessments, we assess the variable according to respective specialization (i.e., nutritional status assessed by our dietitian, and HAEC assessed by our pediatric surgeon). There are two pediatric surgeons in our health center performed the pull-through surgery, and both of their data included in this study.

A Chi-square test was used to determine the significant correlation between attributes. If the chi-sample size is smaller than 50, then the Fisher exact test will be used to determine the significance rather than Chi-square. The result was regarded as statistically significant if the p-value less than 0.05 (p<0.05). All statistical analysis was performed with SPSS 20.0.

RESULTS

In our study population, there was a male to female ratio of 1.75:1 (14 boys and 8 girls) patients with HSCR. Most patients (40.9%) are diagnosed at 1-6 months old. In our study, most patients have good nutritional status after pull-through surgery (68.2%), and most surgeries were done with Duhamel procedure rather than the Soave procedure. Also, five patients (22.7%) developed and diagnosed with HAEC after surgery. The characteristics of the subject were shown in Table I.

Among 17 patients that underwent Duhamel procedure

Table I: Characteristic of subjects

| Variabel | n | % |
|---------------------------------------|----|------|
| Gender | | |
| Male | 14 | 63.6 |
| Female | 8 | 36.4 |
| Age | | |
| <1 Month | 6 | 27.3 |
| 1 – 6 Month | 9 | 40.9 |
| >6 Month – 1 Year | 4 | 18.2 |
| >1 Year | 3 | 13.6 |
| Nutritional Status After Pull Through | | |
| Good | 15 | 68.2 |
| Poor | 7 | 31.8 |
| Types of Pull Through | | |
| Duhamel | 17 | 77.3 |
| Soave | 5 | 22.7 |
| HAEC | | |
| Yes | 4 | 18.2 |
| No | 18 | 81.8 |

HAEC: Hirschsprung-associated enterocolitis

in this study, four patients developed HAEC. All patients who underwent the Soave procedure (5 patients) did not develop HAEC. Since the data did not meet the requirements for Chi-Square test analysis, then Fisher's Exact test was used to determine the significance of the result. Based on the results of the statistical analysis, there was no significant correlation between the type of pull-through procedures (Soave and Duhamel) and the incidence of HAEC in HSCR (Table II).

Table II: The correlation between types of pull through and the Incidence of Hirschsprung-associated Enterocolitis (HAEC)

| Types of Pull Through | HAEC | | | | Total | | p-value |
|-----------------------|------|------|-----|------|-------|-----|---------|
| | No | % | Yes | % | N | % | |
| Duhamel | 13 | 76.5 | 4 | 23.5 | 17 | 100 | 0.5325 |
| Soave | 5 | 100 | 0 | 0 | 5 | 100 | |

HAEC: Hirschsprung-associated enterocolitis

Among 17 patient that underwent Duhamel procedure in this study, six patient showed poor nutritional status (35.3%) while only one patient (20%) showed poor nutritional status when underwent Soave procedure. The statistical analysis showed there was no significant correlation between the type of pull-through procedures and nutritional status in our patient (Table III).

Table III: The correlation between types of pull through and nutritional status

| Types of Pull Through | Nutritional status | | | | Total | | P |
|-----------------------|--------------------|-------|------|-------|-------|-----|-------|
| | Good | % | Poor | % | N | % | |
| Duhamel | 11 | 64.7% | 6 | 35.3% | 17 | 100 | 0,477 |
| Soave | 4 | 80% | 1 | 20% | 5 | 100 | |

All the patients with HAEC showed poor nutritional status, while only three patients (16.7%) showed poor nutritional status when there was no presence of HAEC. The statistical analysis showed there was a significant correlation between HAEC and the nutritional status of the patient (p-value 0.005, OR 6.0, 95% CI 2.136 - 16.857) (Table IV).

Table IV: The correlation between Hirschsprung associated enterocolitis (HAEC) and nutritional status

| HAEC | Nutritional status | | | | Total | | P-value | OR (95% CI) |
|------|--------------------|-------|------|-------|-------|-----|---------|----------------------|
| | Good | % | Poor | % | N | % | | |
| Yes | 0 | 0% | 4 | 100% | 4 | 100 | 0.005 | 6.0 (2.136 – 16.857) |
| No | 15 | 83.3% | 3 | 16.7% | 18 | 100 | | |

HAEC: Hirschsprung-associated enterocolitis

DISCUSSION

In this study, the frequency of HAEC was higher through the Duhamel procedure than the Soave procedure. This may be possible because the Duhamel technique still leaves a small segment of the aganglionic colon (13). Despite that, the Duhamel procedure could minimize

the risk of constipation (9). This study is in accordance with the study conducted by Parahita et al., which showed that HAEC frequencies after pull-through surgery are seen more often with the Duhamel procedure rather than the Soave procedure (6).

The result of the statistical analysis showed that there was no significant correlation between surgical procedure chosen and the development of HAEC. Conversely, a study from Parahita et al., suggested the frequency of HAEC was significantly higher after the Duhamel than the Soave procedure. This difference may be caused by our small sample size in the study. Despite the difference in the statistical analysis, our result showed a similar incidence of HAEC with Parahita et al. study, which showed the presence of HAEC in 23.5% patient underwent Duhamel procedure, and no patient underwent Soave procedure showed sign of HAEC (6).

According to some studies, there are several factors that may increase the likelihood of HAEC after surgical treatment of patients with Hirschsprung disease, such as patient's age, anomalies (e.g., Down syndrome), technical errors during surgery, patients with the previous history of HAEC, and the level of aganglionosis. Some research suggested that sex and type of surgery could be possible risk factors that increase the likelihood of HAEC in patients with HSCR, but this finding is still controversial. According to Menezes M and colleagues, currently, there is no high-quality research between the type of pull-through operation and development of HAEC.(7,14)

Nutritional status represents the balance between nutrition intake and the requirement for biological function in the body. Our statistical analysis showed there was no significant correlation between nutritional status and type of pull-through procedures. However, our result showed higher poor nutritional status in patient underwent Duhamel procedures. In addition, only patient underwent Duhamel procedures showed the presence of HAEC. All the patients with HAEC showed poor nutritional status. Our statistical analysis showed a significant correlation between HAEC and nutritional status. One of the factors that may affect the nutritional requirement is the presence of gastrointestinal disease (15,16). Patients that received pull-through surgery may experience complications such as constipation, HAEC, or incontinence, which results in the accumulation of faeces in the intestine. Furthermore, abnormal bacterial activity may disrupt intestinal homeostasis, causes nutrients not to be appropriately metabolized, inhibits absorption, and excretion, leading to poor nutritional status (8,10,11).

The author acknowledges that there is some limitation in this study including small sample size, the imbalance frequency of procedures used in the pull-through surgery, and the chance of some bias (e.g., Recall

bias). Despite this limitation, this was the first study conducted at Riau Province, Indonesia, that evaluates the surgery procedure regarding prognosis after pull-through surgery in patients with Hirschsprung Disease. Nevertheless, further study with a bigger population should be conducted to evaluate the prognosis of pull-through surgery in Hirschsprung disease and to further evaluate the occurrence of HAEC with different surgical procedures and associated risk factors. Future meta-analysis to evaluate the nutritional status based on the procedures of pull-through surgery (e.g., Soave/Duhamel) is also recommended.

CONCLUSION

Our study showed that there was no significant correlation between HAEC and nutritional status after pull-through surgery, findings in our study showed a higher frequency of HAEC and poor nutritional status with Duhamel procedures. Our finding showed there is a significant correlation between HAEC and nutritional status. Patients with HAEC tended to have poor nutritional status. Therefore, the presence of HAEC may be the key to poor nutrition status that disrupt gastrointestinal function. However, the result in our study may need further investigation due to the small sample size. Our study showed that there is still some inconsistency between the occurrence of HAEC and nutritional status with the surgical procedures chosen.

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