SURGERY TECHNIQUE, GENDER, AGE ON HIRSCHSPRUNG ASSOCIATED ENTEROCOLITIS SCORE OF POST-PULL THROUGH HIRSCHPRUNG PATIENTS

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ABSTRACT

Knowing if there are differences in HAEC scores following pull-through procedures between groups due to the interaction of surgical techniques with gender by controlling the influence of patient age is as important as determining if there are differences in Pull-Through through the procedure between male and female patients. The purpose of this study was to compare HAEC score profiles after Pull-Through surgery using Duhamel's surgery technique and TEPT surgery technique to see whether there was a difference through the process of comparing HAEC scores after the Pull between Duhamel's surgical technique and TEPT surgery technique while controlling the patient's age impact. The study, which compares Duhamel's method with the TEPT process, is an ex-post facto descriptive and inferential study with the aim of determining the profile and variation of post-pull-through HAEC scores. Patients at RSUP Dr. M. Djamil Padang who suffered from Hirschsprung disease between 2018 and 2022 were the study population. All patients in the study sample, compiled using the method of complete sampling, were between 0 and 5 years old. The HAEC scores of individuals with Hirschsprung's disease are determined by a combination of age, gender, and surgical approach. HAEC scores are strongly influenced by age variables alone, the older the patient, the higher the HAEC score. There was a difference in the average HAEC score between the group of patients undergoing Duhamel surgery and the group undergoing TEPT surgery, with the HAEC score of the Duhamel engineering group being higher than that of the TEPT surgery technique group. The average HAEC score for the male and female patient groups was the same.

INTRODUCTION

Hirschsprung Disease in simple language is referred to as a condition in which the colonic area cannot push the stool as it should so that it can cause partial or complete blockage of the colon. Gunadi, with more technical language, said that Hirschsprung disease is a congenital birth defect that occurs in the intestinal nervous system caused by the absence of intramural ganglion cells in the plexuses of Auerbach and Meissner, resulting in functional obstruction of the colon (Gunadi et al., 2020).

Many researchers suggest Hirschsprung disease occurs in about 1 in 5000 live births (Pecoraro et al., 2021), (Hagens et al., n.d.). The incidence of Hirschsprung disease also differs among the breeds. Research by Anderson et al, for example, showed that among the black race, children of the African-American race had the highest incidence of 4.05 out of 10,000 live births, children of the Asia-Pacific islands 2.45 out of 10,000 live births. As for children of the white and
Surgery Technique, Gender, Age on Hirschsprung Associated Enterocolitis Score of Post-Pull through Hirschprung Patients

Caucasian races the figure is 1.89 out of 10,000 live births (Anderson et al., 2018). In Indonesia, cases of Hirschsprung disease are higher, namely 1 in 3,250 live births (Gabriela et al., 2020). In addition, research on Hirschsprung disease, in addition to involving race, also involves other variables such as the sex of people with Hirschsprung disease. Most of the studies found that male sufferers were significantly more numerous than female sufferers (Anderson et al., 2018; Oh et al., 2020; Parahita et al., n.d.).

One of the complications of Hirschsprung disease is the serious and life-threatening Hirschsprung-associated enterocolitis (HAEC). It was Harold Hirschsprung, a Danish pediatrician, who first described congenital megacolons in 1886 based on his observations of two children who died at the age of 7 and 11 months, allegedly due to repeated HAEC attacks (Demehri et al., 2013). The causal relationship between Hirschsprung disease and HAEC was recognized by Swenson and Fisher in 1956 and the process was later described in detail by Bill and Chapman in 1962.

Although with the advances that have been made in the treatment of Hirschsprung disease, HAEC remains a frequent Hirschsprung disease complication with apparent morbidity and mortality, and its etiology and pathophysiology are still poorly understood (Shono et al., 2020). Significant advances in the treatment of Hirschsprung disease have developed in the last 50 years, starting with Swenson and Bill in 1948 and then surgery by Duhamel, Soave, and others.

There are three definitive pull-through procedures that have been introduced for Hirschsprung disease namely Swenson, Soave, and Duhamel Surgery Techniques (Holschneider & Puri, 2008), which can be performed using an open or minimally invasive approach (Taguchi et al., 2008). In 1963, Soave introduced transanal endorectal pull-through (TEPT) as its new operating technique. The basic concept of this operation is surgery from the opening (Taguchi et al., 2008). This procedure was actually first introduced by Rehbein in 1959 for surgical procedures on high-lying anorectal malformations. But by Soave was introduced for the definitive surgical procedure of Hirschsprung disease. The main purpose of this Soave procedure is to remove the aganglionic rectal mucosa, then pulled over the ganglionic proximal colon erobos into the peeled lumen rectum (Grosfeld et al., 2006; Peña & Levitt, 2011).

Although endorectal pull-throughs for Hirschsprung disease are performed correctly, about 40% of Hirschsprung disease patients are estimated to have HAEC. HAEC which can simply be interpreted as an inflammatory state of the intestine pre or postoperative in Hirschsprung disease. Postoperative HAEC is one of the complications with high mortality and morbidity (Plekhova et al., 2021). However, there is still little information obtained through research on its effectiveness in reducing the risk of HAEC after surgery.

The etiology of HAEC, especially after surgery is not yet widely known. Some of the symptoms experienced by HAEC patients include diarrhea, flatulence, and vomiting, besides that it is also accompanied by fever, and finally septic shock (Singer et al., 2018). HAEC is the cause of morbidity and mortality of people with Hirschsprung disease. The incidence of HAEC worldwide ranges from 6-58% (Nakamura et al., 2018). The mortality rate in HAEC is quite high, which is between 6-30% and in patients accompanied by tuberculosis the mortality rate can reach 50% (Song et al., 2021).
Surgery Technique, Gender, Age on Hirschsprung Associated Enterocolitis Score of Post-Pull through Hirschprung Patients

This study intends to compare HAEC scores in post-pull-through HAEC patients between TEPT Surgery Technique and Duhamel Surgery Technique from a gender angle by controlling the age of patients at RSUP Dr. M. Djamil Padang. This study intends to compare HAEC scores in post-pull-through HAEC patients between TEPT Surgery Technique and Duhamel Surgery Technique from a gender angle by controlling the age of patients at Dr. M. Djamil Padang Hospital.

This study aims to find out how the profile of HAEC scores after Pull-Through action according to Duhamel surgery technique and TEPT surgery technique, find out whether there is a difference in HAEC score after Pull-Through between Duhamel's surgery technique and TEPT surgery technique by controlling the influence of patient age, find out whether there is a difference in HAEC score after Pull-Through between male and female patients by controlling the influence of patient age and knowing whether there were differences in HAEC scores after pull-through procedures between the groups resulting from the interaction of surgical techniques with gender by controlling the influence of the patient's age.

The results of this study are expected to be useful to increase understanding of the incidence of HAEC after the Pull-Through procedure both through the Duhamel surgery technique and through the TEPT surgery technique, especially for children suffering from Hirschsprung's disease, and can be used as a basis for thinking about the consideration of medical actions that will be carried out further after the Pull-Through procedure both through the Duhamel Surgery Technique and through the TEPT surgery technique to reduce the severity of HAEC.

METHOD

This study is a descriptive and inferential ex-post facto study that aims to investigate the profile and difference in post-pull-through HAEC scores according to operating techniques between the Duhamel procedure and the TEPT procedure. Haec score differences by sex between men and women by controlling the age of patients as covariates. This research was conducted for two months, namely from May to August 10, 2022. The data collection of this study was carried out at RSUP Dr. M. Djamil Padang which is a type A hospital and is the main referral hospital for the West Sumatra region. The population of this study was all patients suffering from Hirschsprung disease at RSUP Dr. M. Djamil Padang in 2018-2022. The samples in this study were taken using the total sampling technique, where all patients were aged 0-5 years.

The data collected in this study are data on HAEC scores, Hirschsprung disease surgery techniques used, patient sex and patient age. The data needed is secondary to the recording of medical records of patients suffering from Hirschsprung disease in the medical record subdivision for the 2018-2022 period at RSUP Dr. M. Djamil Padang.

The flow of the study is described as follows:
The collected data is processed with the following steps so that it is ready for analysis.

1) Editing: Editing is done to check the accuracy and completeness of the data. If the data is incomplete or there is an error, the data is completed by re-looking at the medical record;
2) Coding: Data that has been collected and corrected for accuracy and completeness is then coded by the researcher manually before being processed into a computer;
3) Entry: The cleaned data is then entered into a statistical processing program;
4) Cleaning: Re-checking all data that has been entered into the computer to avoid errors in data entry;
5) Analyzing: Inferential analysis aims to test the research hypothesis by comparing HAEC scores among: (1) the group of operating techniques, namely the HAEC scores between duhamel operation techniques and TEPT operation techniques; (2) comparing HAEC scores between sex groups, namely HAEC scores between men and women; and (3) comparing HAEC scores between groups generated by interactions between surgical techniques and sexes (Duhamel-Male: Duhamel-Female, TEPT-Male: TEPT-Female), by controlling the patient's age variable as covariate.

The data were analyzed statistically using with the help of the Windows program SPSS 24.0 (Statistical Package for Social Science). There are two analytical techniques used, namely covariance analysis (Anakova) and Chi-Square.

RESULT AND DISCUSSION

Inferential Analysis Results

Test Analysis Requirements

There are at least two requirements anakova that need to be met, namely the normality test and the homogeneity test. The study also tested both requirements with the following results.
Surgery Technique, Gender, Age on Hirschsprung Associated Enterocolitis Score of Post-Pull through Hirschprung Patients

The normality test is concluded in table 1. Both with the Kolmogorov-Smirlov test with a significance coefficient of 0.200 and with the Shapiro-Wilk test with a significance coefficient of 0.341 which is greater than 0.05, the data are normally distributed.

<table>
<thead>
<tr>
<th>Table 1. Normality Test Conclusion</th>
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<tbody>
<tr>
<td></td>
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<tr>
<td>Kolmogorov-Smirnov</td>
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<tr>
<td>Statistic</td>
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<tr>
<td>Residual Score Total</td>
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</table>

The variance homogeneity test for comparing the compared groups (Duhamel vs TEPT; male vs female; and Duhamel-male vs Duhamel-female vs TEPT-male vs TEPT-female) was tested with the Lavene test whose results as concluded in table 2 successively produced homogeneity coefficients of 0.444; 0.846 and 0.714 (appendix). Since all these homogeneity coefficients are greater than 0.05 then the compared group is homogeneous. From the results of the normality and homogeneity test, it can be implied that the requirements for covariance analysis (Anakova) of nature to test the research hypothesis have been met.

<table>
<thead>
<tr>
<th>Table 2. Homogeneity Test of Lavene Variance</th>
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<td>.477</td>
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**Hypothesis Test**

The results of the analysis to test research hypotheses related to the influence of independent variables on HAEC scores are concluded in table 3. The model in the source column with a significance of .000 as shown in table 3 means that the variables of patient age, surgical technique and gender jointly affect the HAEC score of Hirschsprung disease patients.

Furthermore, the age variable as a covariate with a significance of 0.002 has a significant effect on the HAEC score.

<table>
<thead>
<tr>
<th>Table 3. Test of the Effect of Independent Variables on HAEC total score</th>
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<tbody>
<tr>
<td>Source</td>
</tr>
<tr>
<td>Model</td>
</tr>
<tr>
<td>Age</td>
</tr>
<tr>
<td>Operating technique</td>
</tr>
<tr>
<td>Gender</td>
</tr>
<tr>
<td>Surgery technique * Gender</td>
</tr>
<tr>
<td>Error</td>
</tr>
<tr>
<td>Total</td>
</tr>
</tbody>
</table>
Furthermore, the magnitude in table 3 is used to test the research hypothesis as follows.

**First Hypothesis**
1) $H_{0A1}$: There was no significant significant difference in the average haec score after pull-through between Duhamel's surgery technique and TEPT surgery technique after controlling patient life;
2) $H_{1A2}$: There was a significant difference in the average HAEC score after pull-through between Duhamel's surgery technique and TEPT surgery technique after controlling patient life.

**Statistical Hypothesis**
1) $H_0: \mu_{YA1} = \mu_{YA2}$;
2) $H_1: \mu_{YA1} \neq \mu_{YA2}$

It turned out that the results of the analysis resulted in a significance level of 0.043. This result is smaller than 0.05. This means that there is a difference in the average HAEC score between Duhamel's operating technique and tept surgery technique. Hence the null hypothesis was rejected, meaning that the research hypothesis stating there was a difference in the average HAEC score between duhamel's surgery technique and tept surgery technique was supported by research data.

**Second Hypothesis**
1) $H_{0B1}$: There was no significant difference in the average haec score after pull-through between male patients and female patients after controlling the age of the patients;
2) $H_{1B2}$: There was a significant difference in the average haec score after pull-through between male patients and female patients after controlling the age of the patients.

**Statistical Hypothesis**
1) $H_0: \mu_{YB1} = \mu_{YB2}$ (no difference);
2) $H_1: \mu_{YB1} \neq \mu_{YB2}$ (there is a difference)

It turned out that the results of the analysis resulted in a significance level of 0.278. This result is greater than 0.05. This means there is no difference in average HAEC scores between male patients and female patients. Hence the null hypothesis is accepted, meaning that the research hypothesis that states there is a difference in the average HAEC score between male patients and female patients is not supported by research data.

**Third Hypothesis**
1) $H_{0AB}$: There was no effect of interaction between surgical techniques and sex on the average HAEC score after pull-through after controlling the patient's lifespan;
2) HAAB: There was an effect of interactions between surgical techniques and sex on the average HAEC score after pull-through after controlling the patient’s lifespan.

**Statistical Hypothesis**

1) \( H_0: \mu_{Y\text{A1B1}} = \mu_{Y\text{A1B2}} = \mu_{Y\text{A2B1}} = \mu_{Y\text{A2B2}} \) (no difference);

2) \( H_0: \mu_{Y\text{A1B1}} \neq \mu_{Y\text{A1B2}} \neq \mu_{Y\text{A2B1}} \neq \mu_{Y\text{A2B2}} \) (there is a difference)

It turned out that the results of the analysis resulted in a significance level of 0.417. This result is greater than 0.05. This means there is no difference in average HAEC scores between the groups of interaction results between the surgical technique and the gender (Duhamel-male, Duhamel-female, TEPT-male and TEPT-female groups). Hence the null hypothesis is accepted, meaning that the research hypothesis stating there is a difference in the average HAEC score among these groups is not supported by research data.

**Fourth Hypothesis**

1) \( H_0: \) There was no significant difference in the proportion of HAEC patients after pull-through between Duhamel's surgery technique and TEPT surgery technique;

2) \( H_1: \) There was a significant difference in the proportion of HAEC suffersers after pull-through between Duhamel's surgery technique and TEPT surgery technique.

**Statistical Hypothesis**

1) \( H_0: p_{\text{haecD}} = p_{\text{haecT}} \);

2) \( H_1: p_{\text{haecD}} \neq p_{\text{haecT}} \)

The fourth hypothesis was tested with Chi-Square whose results are summed up in Table 4.10. It turned out that the results of the analysis using Chi-Square showed Pierson's Chi-Square coefficient of 5.030 with a significance level \( p = 0.025 \) which was smaller than 0.05. This means that there is a difference in the proportion of HAEC sufferers after surgery with the Duhamel surgery technique compared to the TEPT surgery technique. Therefore, the null hypothesis was rejected, meaning that the research hypothesis that stated there was a difference in the proportion of HAEC sufferers among the group of patients operated on by duhamel surgery technique with TEPT surgery technique supported by research data.

<table>
<thead>
<tr>
<th>Table 4. Chi-Square Test</th>
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<tbody>
<tr>
<td><strong>Value</strong></td>
</tr>
<tr>
<td>Pearson Chi-Square</td>
</tr>
<tr>
<td>Continuity Correction(^b)</td>
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<tr>
<td>Likelihood Ratio</td>
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</tbody>
</table>

Based on the results of the research findings as stated in the previous section, there are several things that need to be discussed. First, the data showed that most (66%) of Duhamel's surgical techniques were used for Hirschsprung disease at the Pediatric Surgery Poly of RSUP Dr. M.
Surgery Technique, Gender, Age on Hirschsprung Associated Enterocolitis Score of Post-Pull through Hirschprung Patients

Djamil Padang. This may be due to duhamel's surgical technique being invented first and has been used since 1956 (Parahita et al., n.d.; Pecoraro et al., 2021; Youn et al., 2021), while TEPT operating techniques were only introduced in the 1990s (Gunadi et al., 2021; Schleef et al., 2021; Taguchi et al., 2008). This time difference is thought to have also affected the expertise of doctors, other medical personnel and also the availability of logistics in the field so that Duhamel's surgical technique is more widely used.

Next, it turns out that people with Hirschsprung disease at RSUP Dr. M. Djamil Padang are more men than women. It is indeed linear with many studies conducted elsewhere (Bawazir, 2020; Halleran et al., 2020; Oh et al., 2020). However, it is not yet clear why men suffer more from Hirschsprung disease.

Furthermore, when the HAEC score was used to determine whether a patient had HAEC, using the Pastor's instrument with a cut-off of 10, it turned out that out of 53 patients 26 people (49%) had HAEC. This finding is almost the same as the findings of studies that have been carried out by several researchers which found that around 40% of people with Hirschsprung disease will experience complications of HAEC (Chan et al., 2021; Gunadi et al., 2020; Schleef et al., 2021).

If the HAEC score is further used to test whether there is a difference in the proportion of HAEC sufferers between the group operated on by duhamel surgery technique and TEPT surgery technique. It turned out that in this study the proportion of HAEC sufferers after surgery with duhamel surgery technique was greater than after surgery with TEPT surgery technique and was statistically significant. The author has not succeeded in finding other studies that compare Duhamel's surgical technique with TEPT surgery technique in terms of the proportion of HAEC sufferers after surgery.

Parahita et al, for example, compared the Duhamel surgery technique with the Soave surgery technique found the HAEC frequency was significantly higher after the Duhamel procedure than the Soave procedure. In addition, patients with preoperative enterocolitis are likely to develop HAEC (Parahita et al., n.d.).

Duhamel and TEPT surgical techniques are often used to treat Hirschsprung disease. The results of these two techniques varied among published studies, and there was no consensus regarding the most effective techniques for treating Hirschsprung disease. Some compare postoperative results after Duhamel pull-through versus TEPT in terms of postoperative constipation, reducing the risk of constipation, and the patient's bowel movement patterns (Seo et al., 2018)

CONCLUSION

Factors of age, gender and surgical technique together determine the HAEC score of Hirschsprung disease patients. In itself the age variable significantly determines the HAEC score, the higher the patient's lifespan the higher the HAEC score.

There was a difference in the average HAEC score between the group of patients with the Duhamel surgery technique and the TEPT surgery technique group, where the HAEC score of the group of patients operated on the Duhamel technique was higher than the group operated on with
the TEPT surgery technique. There was no difference in the average HAEC score between the male patient group and the female group.

There was no difference in the average HAEC scores of patients grouped based on the interaction between the surgical technique and the sex, in other words there was no interaction between the surgical technique and the sex in determining the average HAEC score. The proportion of HAEC sufferers after surgery with Duhamel's surgical technique is more than that of tept surgery technique, the difference is statistically significant.

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Surgery Technique, Gender, Age on Hirschsprung Associated Enterocolitis Score of Post-Pull through Hirschprung Patients


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