

## Comparative Statistical Analysis of Surgical Procedures in the Management of Hirschsprung Disease

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### Rezumat

#### *Analiză statistică comparativă a procedurilor chirurgicale în managementul bolii Hirschsprung*

Acest studiu investighează corelația dintre diagnosticul de boală Hirschsprung (HD), confirmat sau infirmat histopatologic și tipul de intervenție chirurgicală efectuată asupra pacienților. Datele de la 24 de pacienți au fost analizate, aceștia fiind împărțiți în două categorii: HD confirmată (n=17) și HD infirmată (n=7). Rezultatele nu au identificat o asociere semnificativă statistic între diagnosticul de HD și sexul pacienților (valoarea  $p = 1.000000$ ). Sunt necesare studii suplimentare cu eșantioane mai mari pentru a clarifica această relație.

**Cuvinte cheie:** boala Hirschsprung, megacolon congenital, cazuri clinice, analiză statistică

### Abstract

This study investigates the correlation between the histopathologically confirmed or refuted diagnosis of Hirschsprung Disease (HD) and the type of surgery performed on patients. Data from 24 patients were analyzed, divided into two categories: confirmed HD (n=17) and refuted HD (n=7). The results did not identify a statistically significant association between the diagnosis of HD and the sex of the patients (p-value

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= 1.000000). Further studies with larger samples are needed to clarify this relationship.

**Key words:** Hirschsprung disease, congenital megacolon, clinical cases, statistical analysis

## Introduction

Hirschsprung Disease (HD) is a rare congenital condition affecting the large intestine, characterized by a lack of nerve cells in specific segments (1). This absence of nerve cells leads to severe constipation and other intestinal problems. Surgical interventions can be curative but may have significant complications. The choice of surgery type depends on the severity of the disease and the individual characteristics of the patient (2,3).

In this context, we analyzed a group of patients admitted between 2017 till 2024 at the Surgery Clinic of the "Grigore Alexandrescu" Emergency Clinical Hospital for Children. The study included patients diagnosed with congenital megacolon and treated for this condition. We considered this study feasible given the high patient influx at the source hospital, which receives cases from across the country. Data were collected from surgical protocols, general clinical observation records, the hospital's information system and histopathological results (4,5).

The study group was divided into two categories: Hirschsprung positive where the disease was confirmed histopathological and Hirschsprung negative where the disease was not confirmed histopathological. We selected and analyzed 24 patients diagnosed and treated for chronic constipation or enterocolitis caused by congenital megacolon. All patients included in the study had all the necessary clinical data for statistical processing (6).

The overall study is based on the comparative analysis between the positive and negative diagnosis of Hirschsprung disease, reported in relation to:

1. Patients' sex;

2. Type of surgery performed.

The retrospective clinical-statistical study utilized data from the archive of the "Grigore Alexandrescu" Emergency Clinical Hospital for Children in Bucharest and included all patients diagnosed and treated for Hirschsprung Disease during the analyzed period. The diagnosis was confirmed through radiologic and histologic methods, and the cases were reported to the Public Health Directorate.

For statistical analysis, the data were processed in Microsoft Excel and then analyzed using GraphPad and Epi Info software. Statistical verification involved applying Fisher's Exact Test, Mann-Whitney U Test, and Likelihood Ratio Test (7).

## Materials and Methods

This retrospective cohort study was designed to evaluate the types of surgeries performed on patients with Hirschsprung disease, diagnosed histopathologically.

The study included a total of 24 patients who were evaluated for suspected Hirschsprung disease at a specialized medical institution.

- Hirschsprung Disease Confirmed Group: 17 patients had a diagnosis of Hirschsprung disease confirmed through histopathological methods.
- Hirschsprung Disease Refuted Group: 7 patients had a diagnosis of Hirschsprung disease refuted through histopathological methods.

### *Inclusion Criteria:*

- Patients who were evaluated for

Hirschsprung disease and underwent surgical intervention (8).

- Hirschsprung disease diagnosis confirmed or refuted through histopathological methods (9).

**Exclusion Criteria:**

- Patients with incomplete data or lack of histopathological confirmation.
- Patients who did not undergo any surgical intervention.

**Variables:**

*Dependent variable*

The type of surgery performed, which may vary depending on the diagnosis of Hirschsprung disease. Specific surgeries included in this variable are:

The surgical biopsy for Hirschsprung disease

Refers to performing a surgical biopsy to the patient without immediate surgical intervention for correction of the disease, in cases where the diagnosis is uncertain or surgery is not yet possible (10).

Soave endoanal pull-through procedure

A complex surgical procedure consists of a trans anal pull-through of the normally innervated colon into a rectal muscular cuff. It is often used to treat Hirschsprung disease by removing the affected segment of the intestine (2).

Colostomy

It is most commonly used in patients with severe enterocolitis. A colostomy is performed on the ascending or transverse colon and may be retained as a safety measure even after curative surgery (11).

De la Torre procedure

In 1998, Dr. Luis De la Torre-Mondragon described a novel, laparoscopy-unassisted, strictly transanal pull-through surgical procedure that adheres to the principles of the Soave procedure but is performed exclusively via a

perineal approach, eliminating the need for transabdominal mobilization (6,12).

Duhamel sphinctero-rectomy procedure

This surgical technique for the correction of congenital megacolon was developed in response to the technical difficulties and post-operative complications of previous methods. It aims to avoid dissection of the pelvic nerves located on the anterior and lateral surfaces of the rectum, as well as sectioning the remaining spastic anal sphincter, which can lead to subsequent enterocolitis. The procedure involves an anastomosis on only the posterior half of the rectum, reducing the risk of stenosis and dehiscence (8).

*Independent variable*

The diagnosis of Hirschsprung disease, which can be confirmed or refuted histopathologically.

*Demographic variables*

Includes the sex of the patients.

The collected information included:

- Histopathological Diagnosis of Hirschsprung Disease: This refers to the confirmation or refutation of the disease through histopathologic examination of tissue samples (12,13).
- Type of Surgery Performed: Surgical interventions were categorized based on the specific procedures used, including the surgical biopsy for Hirschsprung disease, Soave endoanal pull-through procedure, colostomy, De la Torre procedure, and Duhamel sphinctero-rectomy procedure (14,15).
- Patient Sex: Demographic information regarding the sex of the patients was recorded to examine possible differences in disease prevalence and types of surgeries performed (16,17).

**Table 1.** HD confirmed or refuted HP according to gender

Gender	HD confirmed HP (N=17)	HD refuted HP (N=7)	p_value
Female	2/17 (11,8%)	0/7 (0%)	1,000000 (Fisher's Exact Test)
Male	15/17 (88,2%)	7/7 (100%)	

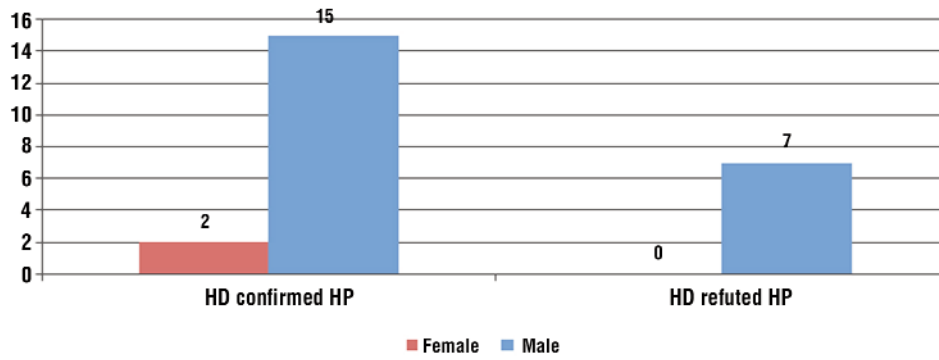


Figure 1. HD confirmed or refuted HP according to gender

**Data Analysis**

Descriptive analysis was employed to describe the demographic characteristics of the patients and the distribution of surgical types.

This included presenting frequencies and percentages for variables such as patient sex, Hirschsprung disease diagnosis, and types of surgeries performed. The data were summarized in *Tables 1, 2* and *Figs. 1, 2* to provide a

Table 2. Types of surgical interventions in patients with positive or negative Hirschsprung Disease

Type of Surgery	HD confirmed HP (N=17)	HD refuted HP (N=7)	p_value 0,615863 (Likelihood Ratio)
The surgical Biopsy for Hirschsprung	1/17 (5,9%)	1/7 (14,3%)	
Soave Endoanal pull-through procedure	2/17 (11,8%)	0/7 (0%)	
Colostomy	1/17 (5,9%)	1/7 (14,3%)	
De la Torre Procedure	12/17(70,6%)	4/7 (57,1%)	
Duhamel sphinctero - rectomyotomy procedure	1/17 (5,9%)	1/7 (14,3%)	

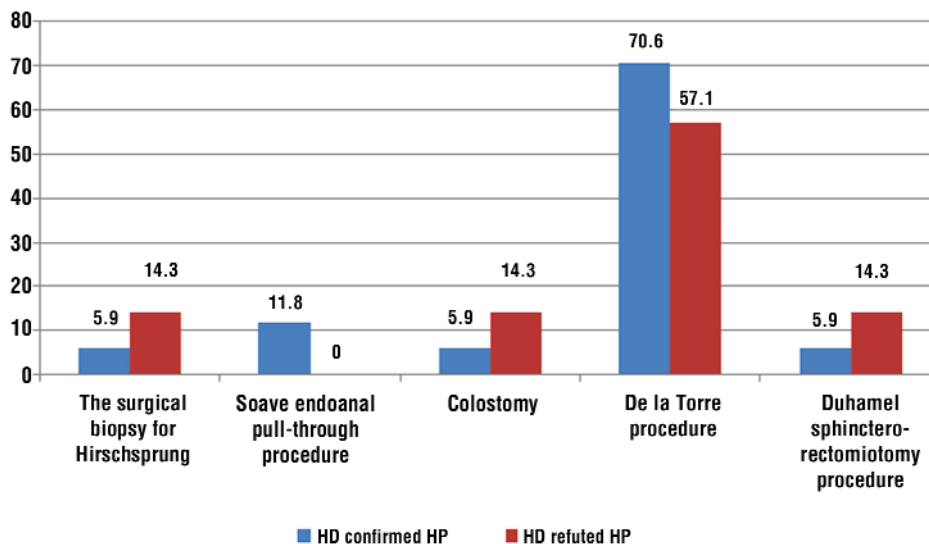


Figure 2. Types of surgical interventions in patients with positive or negative Hirschsprung disease

clear picture of the study population and the surgical interventions carried out (18).

Comparative analysis was conducted to compare the types of surgeries between patients with confirmed Hirschsprung disease and those with refuted Hirschsprung disease. To assess significant differences between these groups, appropriate statistical tests, such as the Chi-square test or Fisher's Exact Test, were used. These tests helped determine if there was a significant association between the Hirschsprung disease diagnosis and the type of surgery chosen, taking into account demographic variables such as patient sex (19,20).

By employing these analytical methods, the study was able to identify potential patterns and differences in surgical approaches, thereby contributing to a better understanding of optimal treatment strategies for patients with Hirschsprung disease.

### *Ethical Considerations*

The study was approved by the institutional ethics committee, and all patient data were anonymized to protect their confidentiality.

### **Results**

- Hirschsprung Disease Confirmed Group: 17 patients had a diagnosis of Hirschsprung disease confirmed through histopathological methods.
- Hirschsprung Disease Refuted Group: 7 patients had a diagnosis of Hirschsprung disease refuted through histopathological methods.

The study included two patients arms, differentiated based on the histopathological diagnosis of Hirschsprung disease: one group with confirmed disease and one group with refuted disease. The demographic analysis also focused on the sex of patients within each group.

#### *Confirmed Hirschsprung disease arm*

This cohort consisted of 17 patients, of whom 2 were female, representing 11.8% of the

group, and 15 were male, representing 88.2%. This indicates a significantly higher prevalence of Hirschsprung disease among male patients in this specific group.

#### *Refuted Hirschsprung disease arm*

This cohort included 7 patients, all of whom were male, meaning that 0% of the patients were female and 100% were male.

#### *Comparative analysis*

The distribution of sex between the two arms suggests a predominance of male patients in both the confirmed and refuted Hirschsprung disease arms. However, the difference is more pronounced in the confirmed disease cohort, where there is a small proportion of females (11.8%), while in the refuted disease cohort, there are no female patients. There were no statistically significant gender differences identified.

#### Analysis of surgical types in patients with confirmed Hirschsprung disease

Hirschsprung Disease is a congenital condition where ganglion cells are absent in a segment of the large intestine, leading to difficulties with stool passage and intestinal obstruction. In the group of patients with confirmed Hirschsprung disease, various types of surgical interventions were performed, each tailored to the specifics of the case. Here is a detailed explanation of the types of surgeries performed:

- The surgical biopsy for Hirschsprung disease: 2 patient (5.9%). One patient identified in each arm.
- Soave Endoanal pull-through procedure: 2 patients (11.8%). Two patients were identified in the Hirschsprung histopathological confirmed arm and 0 subjects in the Hirschsprung histopathological refuted arm.
- Colostomy: 2 patients (5.9%). One patient identified in each arm.
- De la Torre Procedure: 16 patients (70.6%). The majority of patients were identified in the Hirschsprung histopathological confirmed arm, but also 4 subjects were identified in the Hirschsprung

histopathological refuted arm.

- Duhamel Sphincter-Rectomy: 2 patient (5.9%). One patient identified in each arm.

## Discussions

In 2022, Joseph R. and colleagues published a comparative study titled 'Comparative cohort study of Duhamel and endorectal pull-through for Hirschsprung's disease' in *BJS Open*. This study highlighted the limited data available for comparing outcomes between surgical approaches for Hirschsprung's disease. Duhamel and endorectal pull-through (ERPT) are two of the most common procedures performed worldwide. Objective outcomes were compared between contemporary cohorts (aged 4-32 years) undergoing Duhamel or ERPT using a case-control methodology. Data were collected prospectively using standardized questionnaires assessing bowel and bladder function, and quality of life (Pediatric Quality of Life Inventory, Short Form 36, and Gastrointestinal Quality of Life Index).

For the above comparative studies, the aim was to identify if there is a connection between the type of surgery and sex but it seems there were no statistical relevance for this aspect, mostly the type of surgery is dictated by the surgeon based on his experience and capacities but also in regards with patient capabilities.

The second purpose of this study was to emphasize if there is a connection between the type of surgeries and HD, based on the histopathological status and this was also not find as significant from statistical purpose.

## Conclusions

The statistical significance was not achieved. By analyzing the distribution of Hirschsprung disease cases by patient sex, the study may help identify demographic patterns that could influence the diagnosis and treatment of the condition. Although Hirschsprung disease appears to be significantly more prevalent in males than in females, no statistically

significant association was found between the diagnosis of Hirschsprung disease and patient sex.

Even though the diagnosis of Hirschsprung disease was disproved for these patients, their symptoms and clinical needs necessitated various surgical interventions. The De la Torre procedure was the most frequently used, indicating a preference for minimally invasive approaches that enable rapid recovery and reduce the risk of complications. Other procedures, including colostomy and Duhamel sphincter-rectomy, were applied based on the specifics of each case, demonstrating the surgical team's adaptability to the individual needs of the patients.

The data on the types of surgeries performed in patients with confirmed Hirschsprung disease highlight the diversity of surgical approaches utilized in treating this condition. Each surgical option is chosen based on the severity and extent of the disease, the patient's age and overall health, and the experience of the surgical team. The frequent use of the De la Torre procedure suggests a strong preference for minimally invasive techniques that promote quicker recovery and reduce the risk of complications. This approach reflects the ongoing evolution in surgical practices aimed at optimizing patient outcomes in Hirschsprung disease.

## Conflicts of Interests

The authors declare no conflicts of interests.

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