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Original

# Surgical treatment of Hirschsprung's disease at the National Children's Hospital "Dr. Carlos Saenz Herrera" during the period 2000-2010

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## Abstract

**Background:** Hirschsprung´s disease (congenital megacolon) is a common cause of lower bowel obstruction in neonates. Its main characteristic is that a section of the bowel is unable to relax, causing functional colonic obstruction. Treatment for this disease requires the resection of the abnormal bowel segment in order to restore the functions of the healthy part of the intestine. The management and its epidemiology of this disease at the National Children´s Hospital during the period 2000 to 2010 were analyzed.

**Methods:** observational and retrospective study, 130 files were reviewed using Microsoft Excel and the JMP® statistical software.

**Results:** a male: female ratio was 4:1, most cases occurred in full term babies, and the most common initial symptoms were abdominal distension and bilious vomiting. In 78% of the cases the age at which the disease occurred was less than one month. Among the surgical techniques used, the Duhamel technique was the most used (48%), followed by the transanal pull-through technique (35%). The most common postoperative complication for the Duhamel group was rectal septum (8 out of 21 cases), and enterocolitis in the transanal pull-through group (14 out of 19 cases).

**Conclusion:** the clinical and epidemiological characteristics of the cases of Hirschsprung's Disease analyzed are similar to those reported in the literature. The postoperative complications and morbidity was similar for both surgical techniques.

**Keywords:** Hirschsprung disease, colonic aganglionosis, Congenital Megacolon.

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**Abbreviations:** TERPT, transanal endorectal pull-through; NCH, National Children's Hospital "Dr. Carlos Sáenz Herrera".

Hirschsprung's disease is a congenital disorder characterized by the absence of ganglion cells in the myenteric and submucosal plexuses of the intestine.[1] It is a common cause of intestinal obstruction in children, especially during the neonatal period.[2, 3]

The most common presentation of the disease is the location in the rectosigmoid. However, different parts of the colon can be affected, and also completely; part of the small intestine is very rarely affected.[3, 4]

Early diagnosis and management avoids the potential complications of patients with Hirschsprung's disease. Among the initial studies to be performed, are contrast enemas, rectal manometry and rectal biopsy, which eventually confirms the diagnosis.[5-9] Disease management has evolved over the years from very invasive procedures performed in several stages including

three surgeries, to minimally invasive procedures carried out in a single surgical intervention.[10-14]

The goal of the study was to analyze and compare the surgical techniques used in the National Children's Hospital Dr. Carlos Sáenz Herrera (NCH), for the treatment of this condition and its results in the short and medium term. In addition, the epidemiology of the disease was examined.

## Methods

Upon approval of the study by the institutional bioethics committee, the medical records were reviewed as population taking all patients discharged from the National Children's Hospital diagnosed with Hirschsprung's disease from January 2000 to December 2010.

Patients with the diagnosis of Hirschsprung's disease were included during the established term, who had undergone surgery. Cases with incomplete records and all patients who had Hirschsprung's disease ruled out were excluded from the analysis.

Records that met the inclusion criteria were reviewed. Epidemiological data was analyzed, symptomatology, compromised intestinal segment, diagnostic procedures, surgical intervention, postoperative complications according to surgical technique and associations.

Microsoft Excel was used to analyze the data, obtaining the results and completion of the graphics.

JMP® statistical software was used to digitize the data of the 87 patients treated with one of the two surgical methods (transanal endorectal pull-through -TERPT -or Duhamel) and a logistic regression to determine if there were differences in treatment effects was built.

## Results

A total of 130 records were obtained in the Statistics Service list, with discharge diagnosis of aganglionic megacolon or Hirschsprung's disease. 26 records were excluded: 11 for being incomplete and 15 for presenting positive biopsies for ganglion cells, which allowed to rule out Hirschsprung's disease, so that a population of 104 patients were left who met the inclusion criteria.

A relationship male: female ratio of 4:1 was found, for 80 men and 24 women, and an increased incidence in infants born at term (83 cases). Initial symptoms were analyzed and identified bloating as the most frequent, followed by bilious vomiting, terminal meconium and constipation, for a 74%, 52%, 36% and 19%, respectively. At the same time, it showed other forms of initial presentation less often than the previous ones, such as oral intolerance, vomiting clear or dairy, intestinal obstruction, intestinal perforation, jaundice, irritability, among others.

Most cases occurred in less than one month: 78 cases out of 104. No difference was found between the group of one month to six months and that over twelve months, 11 cases diagnosed for the first group and 10 for the second. In the group of six to twelve months only 2 cases occurred.

Most patients did not associate enterocolitis while presenting symptoms of Hirschsprung's disease. At the time of diagnosis, enterocolitis was evident only in 18 patients.

Data was obtained from the abdominal radiograph only in 70% of the 104 patients. In 17% a plain abdominal radiography was not performed and in 13% no information was provided.

The main finding on the plain abdominal radiograph was diffuse distension of loops in 74 cases (85%), followed by lack of distal air in 34 (39%), edema in between loops in 29 (33%), fluid levels in 10 (11.5%), coprostasis in 9 (10%), ground glass appearance in 5 (6%) and pneumoperitoneum in 4 cases (4,5 %).

Barium enema was performed in 66 patients (63%), 29 (28%) did not undergo the procedure, for unspecified reasons on file, and for 9 (9%) patients, it was unable to obtain information about whether or not they performed a barium enema, at the moment of diagnosis.

Of the 66 patients who underwent barium enema, a transition zone was evident in 31, which corresponds to 47%. No transition zone was revealed in 35 patients (53%), of which 20 (30%) had undergone a rectal procedure such as irrigations, digital rectal exam or stimulation with thermometer, so that it can be shown as false negatives in the barium enema.

Of the 31 studies that the transition zone was demonstrated, only 16 cases (52%) were correlated with the true transition zone, according to the final report of Pathology, it was not correlated in 8 cases (26%), and in 7 (22%) no information was found in the record to indicate at what level the transition zone was observed on the barium enema study.

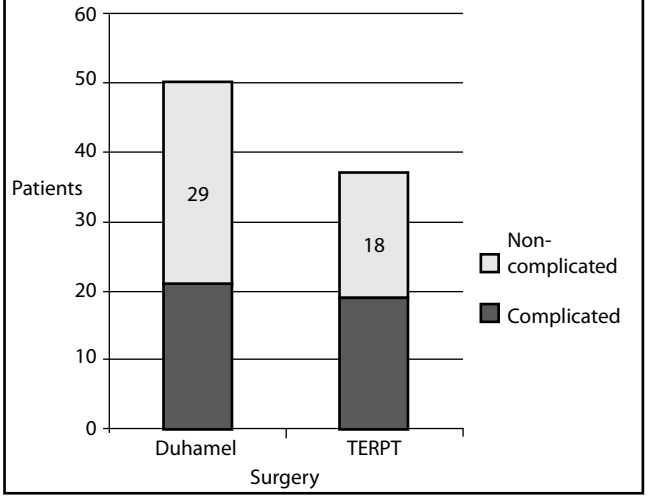
The length of the compromised intestinal segment in 12 patients was ultrashort, short in 60 cases, long in 23, and had total megacolon in 7 cases; and in 2 cases no documentation was found for the compromised segment.

Of the 104 patients in the study, 22 showed association with other diseases: mostly congenital heart diseases (10 patients), followed by Down syndrome (8 patients), among several others. (Table 1)

**Table 1. Associated pathology in patients with Hirschsprung's disease , NCH 2000-2010**

| Associated Pathology | # of patients |
|---|---|
| Congenital heart diseases | 10 |
| Down Syndrome | 8 |
| Meckel's Diverticulum | 2 |
| Dysmorphism | 1 |
| Monorenal | 1 |
| Repetitive bronchopneumonia | 1 |
| Gastroesophageal reflux | 3 |
| Intestinal malrotation | 4 |
| Hypospadias | 1 |
| Cryptorchidism | 1 |
| Corpus callosum agenesis | 1 |
| Bardet-biedl syndrome | 1 |
| Hypogonadism | 1 |
| Polydactyly | 2 |
| Mental retardation | 1 |
| Asthma | 1 |
| Bronchopulmonary dysplasia | 1 |
| Oxygen dependent | 1 |
| Club foot | 1 |
| VACTERL syndrome | 2 |
| Cystic adenomatoid malformation of the lung | 1 |
| Hydroureteronephrosis | 1 |
| Heterotaxy syndrome | 1 |

**Figure 1. Hirschsprung's disease patients with postoperative complications, according to surgical technique. CNH 2000-2010**




Regarding the type of surgery performed, the first surgery that was performed on 48 patients was a colostomy; on 37, one TERPT; on 13, ileostomy, and on 6, sphincter myotomy. As a final surgery for the ostomy patients, Duhamel was conducted on 50 patients, and Soave-Boley and Lester Martin in 1 patient each. 9 cases were documented in patients who have not had their definitive surgery or died without it.

**Figure 2. Causes of reoperation, according to surgical technique in patients with Hirschsprung's disease. CNH 2000-2010**

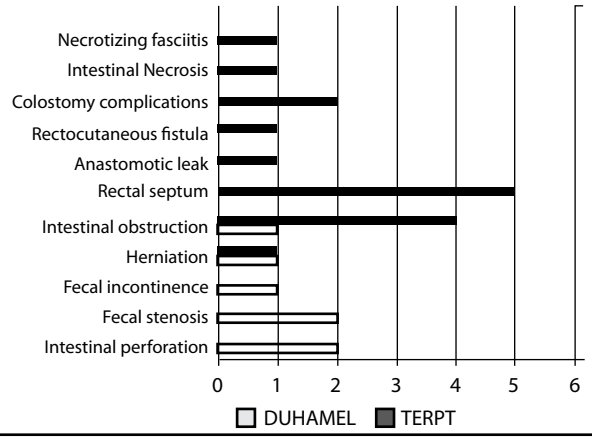
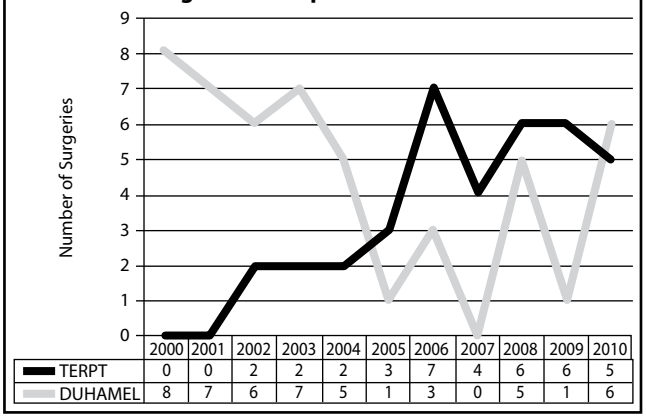



**Figure 3. Surgeries performed per year, according to surgical technique. CNH 2000-2010**




| | 2000 | 2001 | 2002 | 2003 | 2004 | 2005 | 2006 | 2007 | 2008 | 2009 | 2010 |
|---|---|---|---|---|---|---|---|---|---|---|---|
| TERPT | 0 | 0 | 2 | 2 | 2 | 3 | 7 | 4 | 6 | 6 | 5 |
| DUHAMEL | 8 | 7 | 6 | 7 | 5 | 1 | 3 | 0 | 5 | 1 | 6 |

Nineteen of the 37 patients operated on with the TERPT technique, and 21 of the 50 patients undergoing surgery with Duhamel, had postoperative complications. (Figure 1)

Logistic regression was constructed using the JPM statistical program to determine if there are differences in treatment effects of the TERPT and Duhamel; the type of treatment used- TERPT and Duhamel was taken as dependent variable, and postoperative complications were taken as an independent variable. This procedure resulted in a probability associated to the logistic regression model of 0.30; so with a confidence level of 95% that indicates that there is not enough evidence to affirm that there are significant differences in the application of the above treatments.

The most common complications in the group of patients undergoing TERPT were: enterocolitis, rectal stenosis, constipation and intestinal obstruction. Only 7 patients had complications that required reintervention. (Figure 2)

In the intervention group with the Duhamel technique, the most common postoperative complications were: rectal septum, intestinal obstruction, enterocolitis and gastrointestinal bleeding. Among these, 16 patients required surgical reintervention. (Figure 2)

Several patients had more than one complication during the postoperative follow-up period, and some may have needed surgical reintervention on more than one occasion.

The use of each surgical technique for both TERPT and Duhamel was analyzed per year. (Figure 3).

## Discussion

The literature describes a predominance of Hirschsprung's disease in males, from 3:1 to 4:1,[1, 4,15] and more frequent in term newborns.[15] In this study a relationship was found male: female ratio of 4:1, and 77% of the cases occurred in term newborns.

The symptoms that were presented in order of frequency: abdominal distension, bilious vomiting, terminal meconium and constipation, which are correlated with that described in the literature.[4, 15,16] Most patients had symptoms before the first year of life and only 10% did so after the year, mainly showing chronic constipation, as described by various authors.[17]

Unlike what is described in the literature on the manifestation of enterocolitis the time of diagnosis in 50% of cases,[16] in this study only 17% of patients had enterocolitis when presenting initial symptoms.

We found a 25% association with other pathologies, slightly lower percentage than 30% found in other studies.[15, 18,19]

The barium enema showed a limited value as a diagnostic method, since only a quarter of the studies was able to identify the actual transition zone, which implies a lower number than found in the literature.[5, 8,9,11]

The most commonly used surgery was Duhamel procedure in 50 of the 104 cases; the TERPT was used in 37 cases; the sphincter myotomy in 7 patients; Soave-Boley in 1 case, and Lester Martin in another. This difference of 13 cases between Duhamel and TERPT could show that the TERPT began to practice in 2002, therefore no cases were reported in the first two years during the study; in addition, there is no trend or pattern of use between both surgical techniques, during the 10 years covered.

Regarding postoperative complications, they occurred in 51% of the TERPT cases and 42% of Duhamnel cases. This is different from what was described by several authors, who report a higher tolerance by the child and a decreased risk of complications of the TERPT technique,[13,14,20,21] since there is no significant difference between the percentage of patients who became complicated while using both techniques.

A higher percentage of patients requiring reoperation were found in the TERPT group, 81%, versus 68% of the Duhamel group.

One limitation of the study is its retrospective design based on gathering record information, with the disadvantages that this confers, as no other variables were analyzed that resulted in being interesting to compare the results and would enrich the quality of research. Another limitation found was the lack of data in some cases, which prevents analyzing such variables.

**Conflict of Interest:** There is no conflict of interest.

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