



## Hirschsprung disease: do risk factors of poor surgical outcome exist?

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### Abstract

**Introduction:** Long-term results after surgical treatment of Hirschsprung disease (HSCR) are not always as good as a surgeon may perceive. Several studies have tried to correlate preoperative features to the surgical outcome of HSCR, but none came to definitive conclusions. Our study is aimed at identifying risk factors of poorer long-term outcome after surgery for HSCR.

**Materials and Methods:** One hundred two patients with HSCR were included. Eighty had rectosigmoid aganglionosis and 22 had total colonic aganglionosis (TCSA). Preoperative variables were sex, associated anomalies, delayed meconium passage, preoperative enterocolitis, preoperative bowel obstruction, age at surgery, and number of pull-through procedures performed. Outcome measures were surgical complications, postoperative enterocolitis, perineal excoriations, constipation, continence, psychological self acceptance, and patients' perspectives. These were evaluated with regard to preoperative features and length of aganglionosis.

**Results:** Results of patients with TCSA proved to be significantly worse than those of patients with classic HSCR (constipation and poor continence excluded). Male patients with TCSA proved to have a significantly higher incidence of complications (100% vs 38.5%) and poor psychological self acceptance (100% vs 46.2%). Patients with associated anomalies (central nervous system) experienced a significantly higher incidence of postoperative constipation (43% vs 13%). Patients who described failure or delayed meconium passage complained of significantly worse continence (28.3% vs 7.1% of fair to poor continence). Preoperative enterocolitis proved to be significantly correlated to postoperative enterocolitis (16.4% vs 37.1%) and perineal excoriations (13.7% vs 30.9%). Patients with preoperative intestinal obstructions complained of significantly worse psychological self acceptance (37.8% vs 12.5%).

**Conclusions:** Length of aganglionosis has the greatest impact on overall surgical outcome of HSCR. Other minor risk factors have been identified, namely, male sex, associated central nervous system anomalies, failure to pass meconium, and preoperative enterocolitis occurrences. Age at surgery and redo procedures proved not to significantly interfere with the outcome. Our study provides comprehensive and useful data to inform parents and families of a baby with HSCR with regard to

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expected results and long-term outcomes of surgery basing on preoperative features. Further studies on larger series are strongly recommended.

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Long-term results after surgical treatment of Hirschsprung disease (HSCR) appear to be satisfactory. Nonetheless, overall outcome is not always as good as a surgeon may perceive. In fact, various authors described a higher than expected incidence of problems in the long-term follow-up. These include enterocolitis, constipation, failure to thrive, fecal soiling, and incontinence, which are the most frequent complaints regardless of the technique adopted [1-3]. In particular, some reports demonstrated that soiling and incontinence have an important impact on psychosocial functioning and parental criticism, in one word, on quality of life [2].

Surgeons' perspectives of overall outcome tend to differ from that of patients who learn to cope with minor and sometimes major functional disturbances occurring during childhood. Therefore, their perspective of overall outcome can be sometimes overestimated because they adapt and are more satisfied than expected [3]. On the other hand, surgeons can underestimate the impact of the disease on bowel function and quality of life [4]. Nonetheless, overall outcome of HSCR tends to improve with time; and this occurs in both classic and ultralong forms during the whole childhood up to adolescence or adulthood [5,6]. Subsequently, long-term follow-up is mandatory to perceive changes, improvements, and final bowel adaptation after surgery for HSCR.

Reliable criteria to predict the results of treatment would be very helpful in implementing appropriate consent forms and in improving and personalizing the management according to each patient's situation. In the last decade, various reports have tried to correlate the outcome of surgery of HSCR with specific risk factors such as sex, length of aganglionosis, age at operation, preoperative enterocolitis, associated anomalies, and genetic background [1,7-11]. Enterocolitis, length of aganglionosis, and Down syndrome [7-11] proved to be significantly correlated to a poorer outcome, whereas other factors did not show any significant relationship with long-term results of surgery.

The authors will evaluate the results of a series of patients operated on for HSCR at the Gaslini Children's Hospital from January 1993 onward. The aim of this article is to correlate some major outcome measures of treatment of HSCR to specific preoperative features to identify possible risk factors of poor outcome. Classic HSCR and total colonic aganglionosis (TCSA) will be evaluated separately.

## 1. Materials and methods

### 1.1. Patients

All children with HSCR who underwent a pull-through procedure at the Giannina Gaslini Children's Hospital

from January 1993 onward were included in this study. Other inclusion criteria were minimum follow-up of 6 months and surgical procedure performed by the same experienced senior consultant and his staff (VJ) with adequate intraoperative evaluation of the extent of aganglionosis (histochemistry). We submitted to all patients a questionnaire based on what was published by Hanneman et al in 2001 [12], which was added with a group of preoperative items to achieve complete and exhaustive demographic and clinical data. Furthermore, hospital records and outpatient follow-up records of each patient were reviewed to achieve detailed information on surgical procedure, extent of the disease, and perioperative morbidity and mortality.

### 1.2. Variables and outcome measures

Potentially predictive variables for HSCR were sex, presence of associated anomalies (syndromic HSCR, Table 1), length of aganglionosis, delayed passage of meconium (questionnaire and hospital notes matched information considering delay when later than 24 hours of life), preoperative enterocolitis (diagnosed according to Elhalaby et al [13]) preoperative bowel obstruction, age at surgery, and reoperations (redo pull through). We chose the following outcome measures:

1. Postoperative *enterocolitis* (diagnosed according to Elhalaby et al [13])
2. Postoperative surgical *complications* divided into 4 main types (Table 1). In particular, residual anal achalasia was a clinical diagnosis based on the following parameters: (1) absence of residual aganglionosis or dysganglionosis; (2) absence of stricture; and (3) improvement after anal dilatation, Botox (Allergan Inc, Irvin, CA) injection, or isosorbide dinitrate ointment application.
3. Presence of severe postoperative buttocks *excoriations* lasting longer than 3 months
4. *Constipation* according to the modified Rome II criteria [14]
5. *Psychological self acceptance* (patients' perspective) according to modified Hanneman et al [12] scoring system (excellent, fair, poor) combining emotional functioning (body image, shyness, peer relationship, etc) and social functioning (social life, sport activity, etc) results. This score was assessed in patients older than 6 years [12].

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