

Are the complications after laparo-assisted endo-rectal pull-through for Hirschsprung disease related to the change of the anal tone?

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Abstract

The main advantage of the laparo-assisted transanal endorectal pull-through technique (LA - TERPT) for Hirschsprung Disease (HD) is the respect to the rectal-anal anatomy. Postoperative com-

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plications have been observed recently. The present study aims to determine how often these postoperative complications occur in these patients. From January 2009 to December 2018, a retrospective analysis was conducted on 36 children (25 males) with HD who underwent LA-TERPT. Data were collected on the age of diagnosis and surgery, sex, the presence of other pathologies, and cases of enterocolitis. In all cases, anorectal manometry (ARM) was performed to evaluate the anal tone. The median age at diagnosis was 2 months and the mean age at surgery was 5 months. Nine related pathologies were identified: five cases of Down syndrome, one case of hypertrophic stenosis of the pylorus, atresia of the esophagus, polydactyly, and anorectal malformation. A patient with total colonic aganglionosis was identified through laparoscopic seromuscular biopsies. Enterocolitis was diagnosed in 7 cases before and 6 after surgery. At follow-up, the complications recorded were: 5 cases of constipation (treated with fecal softeners), one case of anal stenosis (patient with anorectal malformation), 16 cases of soiling (treated with enemas) and 1 child with fecal incontinence (treated with a transanal irrigation system). The ARM was performed in all 36 cases and showed normal anal tone, except for one case with anal hypotonia. LA-TERPT is an important surgical technique for HD. According to the literature, soiling is the most main complication after HD surgery, probably due to “pseudo-incontinence” with normal anal sphincter tone.

Introduction

Hirschsprung’s Disease (HD) is a congenital disease of the digestive tract characterized by the absence of ganglion cells in the intestinal nerve plexus. It occurs in 1: 5000 live births and in 80% of cases it affects the rectum or rectosigmoid colon.¹⁻³ This anomaly results in an arrest of intestinal transit with a wide variety of symptoms depending on the length of the affected tract. The most characteristic sign, present in 90% of newborns with HD, is the delay in the first meconium passage. Other signs are represented by difficulty evacuation or abdominal distension in infants.⁴⁻⁶ The clinical presentation in older children may be chronic constipation, resistant to medical treatment. The gold standard for diagnosis is a rectal suction biopsy that shows the absence of ganglion cells, increased cholinergic fibers, calretinin, or enolase positivity.⁶⁻⁸ The treatment is surgical, the goal is the resection of the aganglionic bowel without damaging the function of the rectum. Over the years, a variety of surgical techniques have been suggested, with Duhamel, Soave, and Swenson laparoscopic-assisted techniques being the most frequently used procedures for HD.⁹⁻¹⁰ The Laparo-Assisted Transanal Endorectal Pull-Through Technique (LA-TERPT), proposed in 1995 by Keith E. Georgeson, is a procedure that allows a laparo-

scopic mesocolic dissection followed by a perineal endorectal pull-through of the normally innervated intestine through the rectum and the anus.^{11,12} The main advantages of these recent techniques are shorter hospitalization, absence of large abdominal scars, and greater respect for rectal-anal anatomy than traditional open techniques. As regards postoperative complications of LA-TERPT, recent works report an incidence between 25% and 37% for postoperative enterocolitis, 6% - 20% for fecal incontinence/ soiling, and 11% - 14% for constipation.¹³⁻¹⁵ The purpose of this retrospective study is to estimate the rate of these postoperative complications in HD patients undergoing LA-TERPT.

Materials and Methods

From January 2009 to December 2018, we retrospectively analyzed all patients with Hirschsprung's disease who underwent the laparoscopic assisted transanal endorectal pull-through procedure. Surgical treatment included serial intraoperative seromuscular biopsies to determine the length of the aganglionic tract and LA-TERPT, as described by Georgeson.¹¹ Each patient had their anastomosis site evaluated using a laparoscopic view at the end of the procedure to determine if the transition zone had been ruled out. Data was collected on the age of diagnosis and surgery, sex, the presence of other pathologies, and cases of enterocolitis before and after surgery. Each patient underwent a mean follow-up of 48 months (36 - 120 months) to evaluate problems and complications. In all cases, when the patient was at least 3 years old, Anorectal Manometry (ARM) was performed to evaluate the anal tone. Each patient had a bowel preparation with enema three days before the ARM, which was performed with the patient supine without sedation, using an open silicone catheter (2.0 mm diameter) with a side hole and perfused with sterile water (rate of 2 ml/min). The catheter was inserted into the anus with the lateral hole in the rectum so that the tone of the anal sphincter was measured at the withdrawal of the catheter at a rate of 0.5 cm for 30 sec. A pressure between 10 and 50 cmH₂O was considered normal, hypotonia if less than 10 cm H₂O, and hypertonia if greater than 50 cmH₂O.¹⁶ Each patient performed an anal calibration by sedation one month after the surgery and in case of stenosis (difficulty or bleeding during the use of Hegar 12 in children under 6 months of age), a new calibration after 15 days. One surgeon evaluated every patient for

symptom assessment. Constipation was defined if the child met at least two Rome III criteria, fecal incontinence was diagnosed in children over 3 years of age by the inability to check stool more than once per week, and soiling was defined as the stool was found around the anus for at least 50% of the time of the anus examination.¹⁷ All children with fecal problems (constipation, soiling, or fecal incontinence) underwent anal calibration (to rule out anal stenosis) and contrast enema for suspicion of obstructive disease (recurrence). Enterocolitis was diagnosed in cases of abdominal distension, vomiting, diarrhea (with or without bloody stools), and fever. The study was conducted according to the Helsinki principles and was approved by our ethics committee

Results

Thirty-six children with HD (25 males) were treated and included in the study. The median age at diagnosis was 2 months (range 7 days - 12 months) and the mean age at surgery was 5 months (range 1 month - 16 months). The associated pathologies were respectively: Down Syndrome (DS) in 5 cases, one Hypertrophic Stenosis of the Pylorus (HSP), one Esophageal Atresia (EA), two polydactyly and an intermediate form of Anorectal Malformation (AM). Laparoscopic seromuscular biopsies identified one case of Total Colonic Aganglionosis (TCA) (the patient underwent ileorectal anastomosis), 6 (16,7%) Long Segments (LS) (aganglionic tract up to the transverse colon) and 29 (80,5%) classic forms (Aganglionic Rectosigmoid Colon, ARC) (Table 1). There were no complications during surgery and in the immediate postoperative period. In the case of total colonic aganglionosis, seromuscular biopsies were performed several times and the treatment was the laparoscopic colectomy. Enterocolitis was diagnosed in 7 cases before and in 6 cases after surgery (16.7%), two children with Down syndrome (40% of cases of our series) had recurrent enterocolitis (twice) before and after the surgical treatment. At the average follow-up of 48 months (range: 36 - 120 months), the complications recorded (Table 2) were: 16 cases (44.4%) of soiling treated with enemas, 5 cases (13.9%) of constipation solved with fecal softeners, one anal stenosis (patient with anorectal malformation undergoing a one-stage PSARP) and 1 child with fecal incontinence treated with transanal irrigation system. The anorectal manometric study was performed in all 36

Table 1. Length of aganglionic colon after laparoscopic seromuscular biopsies.

HD type	N° of cases	Percentage (%)
Total colonic aganglionosis	1	2.8
Long segment	6	16.7
Rectosigmoid aganglionosis	29	80.5

Table 2. Type and cases of complications at follow up.

Complications	N° (%)	Comorbidity (N)	Length of segment (N)	Mean age at follow up (months)
Postoperative enterocolitis	6 (16.7)	DS (2), HSP (1)	LS (2), ARC (4)	6
Constipation	5 (13.9)	DS (1), EA (1)	ARC (5)	24
Soiling	16 (44.4)	DS (1)	LS (2), ARC (14)	40
Faecal incontinence	1 (2.8)		TCA (1)	24
Anal stenosis	1 (2.8)	AM (1)	ARC (1)	6

DS, Down syndrome; HSP, Hypertrophic stenosis of the pylorus; EA, esophageal atresia; AM, anorectal malformation; TCA, total colonic aganglionosis; LS, long segments, ARC; aganglionic rectosigmoid colon.

Table 3. Correlation between manometric data and complications.

Manometric data	Constipation	FI	Soiling	POE
Normal anal tone	5	0	16	6
Anal hypotonia	0	1	0	0
RAIR present	5	0	16	0
RAIR not present	0	0	0	0

FI, faecal incontinence; POE, postoperative enterocolitis.

cases, at a mean age of 40 months (range: 36 - 48 months), and showed normal anal tone, with a mean pressure tone of 35 cmH₂O (range; 25 - 45 cmH₂O); in one case (patient with fecal incontinence) an anal hypotonia was revealed. RAIR was tested in 21 children (58%) (soiling and constipation) and it was found to be present (Table 3).

Discussion

The LA-TERPT is currently one of the most commonly used treatments for Hirschsprung's disease in various centers of Pediatric Surgery. The outcome is good for several reasons such as short hospitalization, aesthetics, faecal continence, and time of recovery. Nevertheless, even with this surgery, there are complications common to other techniques reported in the literature.¹²⁻¹⁴ Different complications for each technique are reported in some articles, making it difficult to determine which is the best option for the patients.¹⁵ Our series includes patients who were treated with the same technique (LA-TERPT) for aganglionosis of a distinct tract of the colon (total colonic, long segment, or sigmoid colon). Therefore, the analysis of the data is representative of the choice of the surgical technique.

We have observed, after ARM and clinical follow-up, that despite normal anal tone, constipation is present in 13.9% of cases and is easily treated with emollients, while most of the cases (44.4%) present soiling. This disorder has been reported to occur in all techniques,¹⁸⁻²⁴ but it appears to be the most significant problem in LA-TERPT.²⁵⁻²⁷ Enemas can easily be used to treat postoperative soiling, even daily, but the patient does not always follow the treatment and reports this condition as significant. A literature review of long-term complications after LA-TERPT shows an incidence of enterocolitis (16.7% in our series) ranging from 3.05% to 34%, constipation (13.9% in our series) from 0.09% to 38.09% and soiling (44.4% of our patients) from 4.4% to 88%.^{11,19,21} It is interesting to note that this last data is reported more frequently in small than large series: the reason may be that for a small number of patients, more attention is given to minor disorders like soiling.

Faecal soiling is a term used to describe involuntary stool that is deposited in the underwear, regardless of the presence of an organic or anatomic lesion.¹⁷ The cause remains uncertain. Faecal continence involves the ability to recognize the fullness of the rectum, distinguish between solid, liquid, or gas, and retain the rectal content. Faecal incontinence or soiling¹⁷ is a result of losing one of these mechanisms. In the literature, soiling after pull-through is considered due to "pseudo-incontinence", abnormal sensation, or inadequate sphincter control. "Pseudo-incontinence" is a clinical condition where the anatomy and physiological continence mechanisms are preserved. The hypermotility of the rectum is a result of the colon segment's inability to properly fill and empty. Therefore, the natural function of the reservoir is not maintained with consequent soiling. The variability of the

volume of distention of the operated rectum and the loss of discrimination capacity are associated with an abnormal sensation. Excessive muscle stretching at the end of the procedure can cause injury to the anal sphincter. In our opinion, extreme care must be taken during the dissection of the submucosal from the muscle plane and the execution of the pull-through.

Anorectal manometry is useful in distinguishing "pseudo-incontinence" from soiling from other problems, such as fecal incontinence, sphincter elongation after pull-through, or persistence of the transition zone.³⁰⁻³³ In the case of soiling in patients undergoing this procedure, it is necessary to exclude obstructive causes such as anal stenosis or recurrence, therefore we recommend carrying out an anal calibration and a contrast enema.

Soiling is the most common complication in our series, and the study with ARM has indicated that these patients do not have any changes in anal tone. Probably the disorder is not caused by an evident anatomical disorder.

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