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POSTOPERATIVE FECAL INCONTINENCE IN HIRSCHSPRUNG'S DISEASE – TECHNICAL SURGICAL ERROR OR AN INEVITABLE COMPLICATION?

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Abstract: *Background.* Hirschsprung's disease is a congenital disorder of the enteric nervous system which determines functional bowel obstruction because of the absence of the ganglion cells in the muscular and submucosal layer. Postoperative fecal incontinence is a rare complication with life long implications. Surgical preventive measures and clinical protocols can prevent it and improve quality of care.

Objectives. The study evaluates the types and causes of postoperative fecal incontinence in our patients with Hirschsprung's disease.

Methods. This is retrospective study, single surgical team experience. The study included all patients operated for Hirschsprung's disease who presented for fecal incontinence and older than 3 years (age for toilet training). We examined the patients' surgical history (type of surgery, length of resected colon), evaluated the aspect of the colon with contrast enema and performed rectal examination under anesthesia to evaluate the integrity of the anal canal.

Results. Three patients had true fecal incontinence with disruption of the anal sphincters regardless of the retained colon length. Four patients with total aganglionosis had integral anal canal with short intestine but could obtain social continence with bowel management. The rest of the patients (14 patients) had false incontinence (encompresis) and were managed with laxatives and stool bulking agents for minimum 6 months.

Conclusions. Postoperative fecal incontinence in Hirschsprung's patients can and must be prevented. Management of these patients should be done following specific guidelines and in centers that can provide multidisciplinary services.

Keywords: Hirschsprung's disease, fecal incontinence, anal canal, bowel management, clinical protocols.

INTRODUCTION

Hirschsprung's disease is a developmental disorder of the enteric nervous system which affects the motility of the colon. The absence of ganglion cells in the submucosal and muscular plexus causes inability inmuscular relaxation of the bowel with fecal stasis, proximal distension and functional obstruction [1]. In healthy individuals there is a physiologic zone of aganglionosis immediately above the dentate line (up to approximately 1-1.5 cm, depending on age) [2] but lacks the presence hypertrophic nerves characteristic of Hirschsprung's disease [3].

The purpose of surgical treatment is removal of the aganglionic bowel segment while keeping intact the anal sphincter and pull through of the ganglionic bowel with preservation of fecal continence after surgery, no obstructive symptoms such as abdominal distension and postoperative enterocolitis.

Normal defecation requires integrity of the nerve supply, involuntary smooth muscles and voluntary striated muscle. Patients subjected to endorectal dissections (including the resection of the mucosa below the pectinate line) may suffer an important degree of incontinence consecutive to the loss of sensation although no muscle has been injured [4].

In 5% of the patients born with Hirschsprung's disease, fecal incontinence can occur postoperatively, usually because of a surgical cause – damage to the anal

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canal or sphincters [5]. The impact in the long term is extensive and irreversible in the life of the child [6]. Clinical protocols provide important guidance and direction to health professionals in their delivery of care. By detailing requirements and actions to be taken in specified circumstances or contexts, these documents may facilitate a quality in care [7].

In this study we evaluated the types and causes of postoperative fecal incontinence in our patients with Hirschsprung's disease.

MATERIAL AND METHODS

This study is a retrospective, single surgical team experience during a period of 12 years. We analyzed the medical records 24 consecutive patients who presented with symptoms of fecal incontinence or soiling after being operated for Hirschsprung's disease in our clinic or other institutions between 2007 and 2016. 3 patients were not compliant with evaluation and treatment recommendations and were lost to follow up. They were excluded. The study was approved by the hospital ethics committee. We defined continence as voluntary bowel movements without soiling in between and not under any treatment including bowel management. The study included patients operated with Duhamel, Soave and De La Torre Montdragon (transanal endorectal pull-through) procedure.We examined the patients' surgical history (type of surgery, length of colon resected), evaluate the aspect of the colon with contrast enema and performed rectal examination under anesthesia to evaluate the integrity of the anal canal. Rectal biopsies were done with patients with repeated enterocolitis.

RESULTS

We divided patients based on symptoms at presentation in 2 groups: group 1 (7 patients) with more than 5-6 involuntary bowel movements a day in small quantities, nocturnal incontinence or with defecation sensation present but the patient can't reach the toilet in time and group 2 (14 patients) with voluntary bowel movements but soiling in between and history of abdominal distention. The age of the patients was between 4 and 15 years old.

In the first group, 4 patients had total colonic aganglionosis, were operated with Soave procedure, and the main concern was nocturnal incontinence which determined them to wear diapers at night. During the day, they were continent but avoided practicing sports and required to always locate a toilet, easy to reach, when going out. The other 3 patients in the first group, operated in another center, were wearing diapers or pads at all times. One 7 years old patient with Down syndrome had a Soave procedure with unknown location of the transition zone. Second patient had descending colon aganglionosis, was operated with transanal endorectal pull-through procedure, had a history of anastomosis dehiscence, rectal abscess, and posterior sphincterotomy after the patient developed abdominal distention with repeated episodes of enterocolitis. The 3rd patient with transverse colon aganglionosis operated with Soave procedure, dehiscence of the anastomosis, rectal abscess, ileostomy, redo-pull though and multiple ano-rectal fistulae.

In the second group, all patients had a transverse colon or descending colon aganglionosis, 5 patients were operated with Duhamel procedure, 3 with Soave procedure and 6 patients had a transanal endorectal pull-through.

Contrast enema was performed in all cases with evidence of colon dilatation and fecal impaction in the 2nd group. In the 1st group the colon had a normal aspect. All rectal biopsies showed ganglion cells present and normal terminal nerves.

Examination under anesthesia, showed circumferential disruption of the dentate line in the patient with Down syndrome with colonic anastomosis at the anal skin and severe perianal dermatitis (Fig. 1A and 1B). The 2nd patient had partial disruption of the dental line in the posterior half with important fibrosis. The 3rd patient had a patulous anal opening, with fibrosis of the ano-rectal wall and partial disruption of the dentate line. All patients with a damaged anal canal were from the group of true fecal incontinence.

In the second group, examination under anesthesia showed no pathologic modifications except in 2 patients who had a Duhamel procedure, with long septum and fecal impaction of the rectal pouch (Fig. 2). These patients were operated with mechanical section of the septum and prior desimpaction.

The group of patients with total aganglionosis and fecal incontinence had integral anal canal but because the entire colon and terminal ileum has been removed, could not form solid stool. They received treatment with Loperamide, constipating diet and bulking agents. All 4 patients became continent during the day and night and stopped wearing underwear pads. The compliance to treatment was also determined by the fact that they were all adolescents and were motivated to be able to fully integrate with their peers. The 3 patients with true incontinence because of the

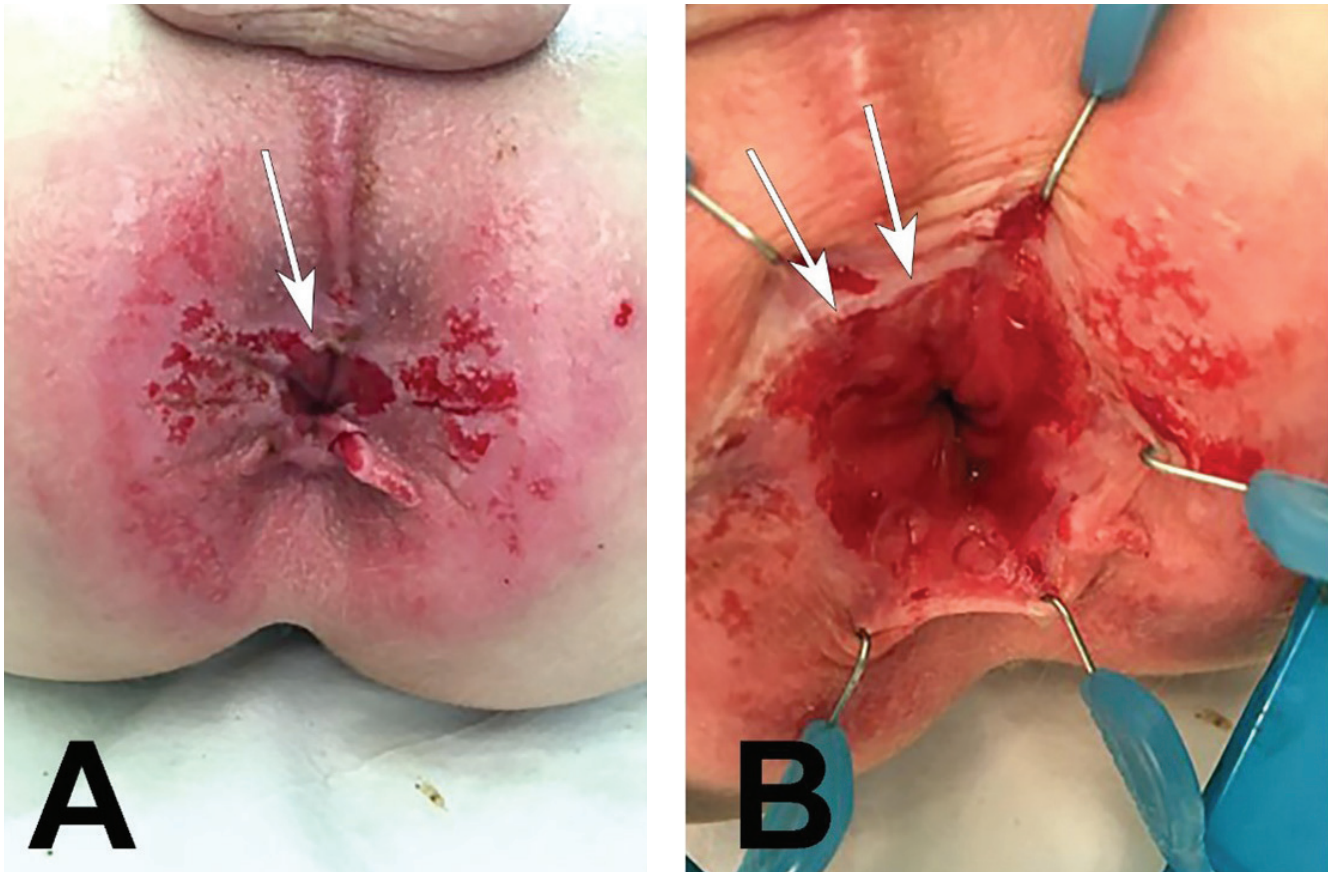

Figure 1. Perineal aspect with A - severe dermatitis and visualization of the colonic anastomosis at the skin level, B- anal canal with disrupted dental line.

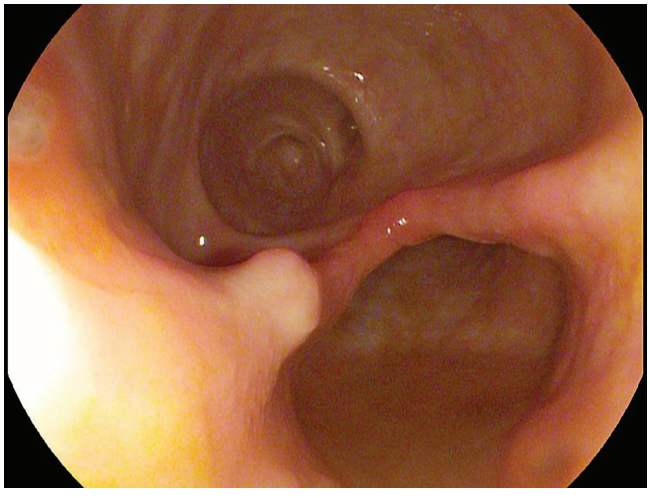

Figure 2. Colonoscopic image of the Duchamel rectal pouch and long remnant septum.

disrupted anal canal were started on bowel enemas to empty the colon and constipating diet. The patient with Down syndrome is still wearing diapers because of the difficulty in implementing and adherence to the bowel management program caused by the cognitive impairment. The 2nd patient has been very compliant and success in implementing the bowel management program and is clean for 24 hours. The 3rd patient chose to have a permanent ileostomy.

In the 2nd group all patients were treated with initial bowel enemas for desimpaction of the colon, bulking agent of the stool and Senna laxatives for a minimum of 6 months. After 6 months patients can stop the laxative and try only diet because, usually, this interval of time allows the colon to recover to a normal size and the child learns to relax efferently their sphincters during bowel movement. In 5 patients they required Senna treatment for more than 6 months and are currently under follow up.

All patients were followed between 7 and 15 months.

DISCUSSION

The central nervous system, the afferent and efferent pathways and receptors are essential factors in fecal continence along with the integrity of the complex sphincter, anal canal sensation and colon motility. The voluntary muscle structures are represented by the

levators, the muscle complex and the external sphincter. They are used only for brief periods when stool in the rectum reaches the anorectal area pushed by the involuntary peristaltic contraction of the recto sigmoid. These muscles also voluntarily relax at the appropriate time to allow the stool to exit the rectum. Both the complex muscle sphincter and the anal canal is normal in patients with Hirschsprung's disease. Recto sigmoid colon has an important role as a bowel reservoir and together with the rest of the colon participates in forming solid stools [8].

Fecal incontinence after surgical management of Hirschsprung's disease should not occur because these patients are born with an anatomically intact continence mechanism including the intact dentate line and intact voluntary sphincters that surround the anus. In fact, all available surgical techniques were designed to preserve the anal canal and, therefore, sensation and the sphincter mechanism [9].

An endorectal dissection can safely be performed without jeopardizing fecal continence provided the mucosa is preserved from about 1 cm above the pectinate line and down whether it is a submucosal or a full-thickness dissection, the muscle layers are divided above the internal anal sphincter. If the pull-through is done via a transanal approach it is extremely important to avoid using retractors and stretch the external sphincters excessively [10].

In cases operated with Duhamel procedure the introduction of mechanical suture devices to the colorectal anastomosis has led to a significant facilitation in division of the common wall, represented by a lower percentage of postoperative complications [11]. Stensrud *et al*. [12] performed anal endosonography and manometry after transanal endorectal pull-through, with or without laparotomy or laparoscopy and showed that postoperativeinternal anal sphincter defects were frequently detected, were associated with daily fecal incontinence and it occurred more often after transanal procedures.

Even when the traction sutures are placed above (proximal to) the pectinate area, circumferential incision and placement of the sutures during the anastomosis shorten the initially proposed length between the rectum and anal canal. As a result, the anastomosis could be completed in a lower (distal) location, damaging the anal canal [13].

The success in treating patients with Hirschsprung's disease and postoperative continence problems can be enhanced by having in place a team who can provide bowel management services to patients and their families [14]. In most cases, patients have encopresis as the cause of their soiling which is treatable or as in the case of patients with total aganglionosis where even if the entire colon is removed, an intact anal canal with the help of bowel management can help achieve full continence.

Studies have shown that Hirschsprung's disease patients have a lower quality of life in the long term and into adulthood depending on the length of aganglionosis and postoperative complications. They have a significantly higher number of bowel movements per week, higher incidence of soiling, urgency, permanent stomas, use of laxatives, enemas and loperamide and also scored significantly lower in their satisfaction with their bowel function. They also reported higher incidence of negative impact on daily life, social interaction and ability to go on vacation [15].

Taking in consideration that fecal incontinence can be avoided, it's wide psychological and socio-economic implications, it depends on us to take all the necessary measures to anticipate factors and situations at risk. It is important that these types of cases are concentrated in tertiary centers with a high volume of cases per year, to have protocols highlighting the surgical principles in Hirschsprung's disease, to have a dedicated histopathology center with frozen section and periodic multidisciplinary meetings.

Protocols should be elaborated by expert consensus based on current knowledge regarding causes, methods of diagnosis, and treatment approaches. Some postoperative complications such as enterocolitis cannot be avoided, only decreased same as in other conditions as for example Chron disease [16]. The principle recommendations include: positive diagnosis of Hirschsprung's disease should be done with rectal biopsy (suction or transrectal); location of the transition zone must be evaluated with contrast enema; dissection in the anal canal should start 1-1.5 cm above the dentate line and must avoid unnecessary stretching of the muscles;the level of pull-through should be assessed with intraoperatory frozen biopsy; postoperative care and follow-up is important to prevent the occurrence of false incontinence (encompresis).

In conclusion, the management of patients with Hirschsprung's disease is complex. Fecal incontinence in Hirschsprung's disease can and should be prevented. The psychological and socio-economic impact is for life. Centers that choose to treat this type of congenital malformation must take all the necessary measures to provide best of care.

Conflict of interest

The authors declare that they have no conflict of interest.

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