

A SEVEN YEARS EXPERIENCE IN HIRSCHSPRUNG'S DISEASE TREATMENT

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Abstract

Introduction. Hirschsprung's Disease (HD) is one of the main causes for the failure of meconium pass in the first 24-48 hours of life or later chronic constipation in infants or children. The diagnosis and the evolution after certain surgical treatment make HD a controversial topic for the pediatric surgeons where the success strongly relates to the postoperative issues.

Purpose. The feasibility and the safety of different diagnosis methods and operative techniques addressed in our team was the aim of our 7 years study.

Materials and methods. A retrospective study has been conducted over the cases of HD treated by our team in the last 7 years (January 2007 – June 2014). The study included 38 patients diagnosed by barium enema and/or suction rectal biopsy and further treated using 3 different operative techniques. Frozen tissue biopsy has been introduced in our team in the last year of our study and it has been used in 5 cases for intraoperative confirmation of the length of aganglionic bowel.

Results. Barium enema (used in 31 cases) was highly suggestive in diagnosis and preoperative imaging of the most cases of HD, but the gold standard for diagnosis is a rectal biopsy, which can be obtained safely using a mucosa-submucosa suction device (used in 14 cases). Modified Duhamel procedure was applied in 19 cases, Soave in 13 cases and De La Torre-Mondragon in 6 cases. The mean age was 28 months (the youngest patient having 7 weeks and the eldest 18 years old). The postoperative complications consisted in: residual septum, rectal bleeding, intestinal occlusion, enterocolitis, anastomotic stenosis, soiling and perianal rash.

Conclusions. The diagnosis quality in our clinic has increased after introducing the rectal suction biopsy and intraoperative frozen section biopsy. The use of the stapler device in the modified Duhamel procedure brings significant improvements regarding outcome. Soave is a valuable solution in total colonic HD and re-do surgery. De La Torre-Mondragon is an elegant procedure in the common type HD. Frozen tissue biopsy is a very safe method for intraoperative evaluation of the border between the aganglionic bowel and healthy colon.

Key words: Hirschsprung's Disease, surgical techniques, postoperative issues

Introduction

Hirschsprung's disease is the most common congenital anomaly in distal gut motility [1]. The intrinsic innervation of the gut wall is derived from neurons that are located entirely within the intramural ganglionic plexuses. There are two kinds of plexuses: myenteric (or Auerbach's) which lays within the muscularis externa between the circular and the longitudinal fiber layers, and there are two or more submucosal plexuses, the most superficial being the Meissner plexus [2]. HD is characterized by the absence of this intrinsic innervation in a variable length. The absence of propagation of the peristaltic wave is associated with the lack of submucosal and myenteric nervous plexuses making obstruction of the distal gut the primary clinical feature of HD [3]. Any child with history of chronic constipation or infant with failure of meconium pass in the first 2 days of life should be checked out for HD using appropriate diagnostic techniques [3, 4]. After the diagnosis of HD the surgical approach with the resection the aganglionic bowel segment is the current definitive treatment. On the other hand, the surgical approach over HD implies a lot of challenges in the matter of postoperative issues [1].

In our clinic, we recorded a significant improvement in the quality of diagnosis of HD by routine use of aspirative rectal biopsy and intraoperative frozen section histopathological examination. We also improved our surgical approach by replacing our classical Duhamel with the one-stage procedure using a stapler device and by introducing De La Torre-Mondragon surgical procedure.

Purpose

The aim of this study is to synthetize our last 7 years experience in HD management and to present the benefits or the disadvantages of different operative techniques used by our team. By using 3 different surgical approaches into treating HD patients and different diagnosis methods we've encountered varied aspects in the postoperative evolution. We are also claiming the importance of aspirative rectal suction biopsy and intraoperative frozen section biopsy.

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Materials and Methods

Our study focuses over the cases of HD treated by our team in the last 7 years (January 2007 – June 2014). We included in this retrospective analysis 38 patients who were diagnosed and surgically treated at different ages. The diagnosis was based by clinical picture, barium enema (26 cases), rectal suction biopsy (14 cases) and intraoperative frozen section biopsy (last consecutive 5 cases). In all cases, the diagnosis was confirmed by histopathological examination, using hematoxylin and eosin staining. We have used 3 different approaches: Modified Duhamel – one stage intervention using a stapler device (19 cases), Soave (13 cases) and De La Torre-Mondragon (6 cases). We have used modified Duhamel technique in common or high forms of congenital megacolon, and also in the majority of the cases admitted with an already performed colostomy. Soave procedure was preferred in high forms/ total aganglionosis and also in re-do pull-through surgeries. In the last years, based on the improved diagnostic means (rectal suction biopsy facility and intraoperative biopsy) we introduced De La Torre-Mondragon technique. In present it is our surgical procedure of choice for patients with common type of HD.

Results

We have studied 38 cases of HD treated by a single team over the last 7 years. The sex ratio was 3.2:1 (29 males and 9 females). The mean age of intervention was 28 months. The youngest patient we have treated was 7 days and the oldest was of 18 years of age.

All cases showed constipation as major clinical sign. 8 patients experienced episodes of enterocolitis, previous to surgery. 12 children presented in emergency with acute signs of bowel obstruction. 15 cases suffered initial enterostomy (10 colostomies and 5 ileostomies), in 9 of them stoma being performed in other centers.

Barium enema was performed in 31 patients. In 9 cases the result was inconclusive (29%). In 6 patients repeated barium enemas were done, without obtaining suggestive images for HD.

In the last 14 cases we improved our diagnosis methods by introducing suction rectal biopsy. In 11 patients the results were positive for HD (78.6%). In 3 of them the result was unsatisfying, because of the quality of the obtained specimen (absence or not enough submucosa).

Frozen tissue biopsy was introduced in our clinic in the last year and was used in the last 5 consecutive cases. In all of them the length of the aganglionic segment was demonstrated.

In our series we encountered 5 difficult cases, misdiagnosed in other centers, with previous failed operations, in which the definitive diagnosis was established by serial open full thickness biopsies [12].

In all operated cases the diagnosis was confirmed by histopathological examination, using hematoxylin and eosin staining.

26 of our cases had common type of HD, 5 patients had a long colonic segment affected, 2 patients with short HD and 5 patients with total colonic HD.

We have used 3 different approaches: Modified Duhamel procedure using a stapler device (19 cases), Soave technique (13 cases) and De La Torre-Mondragon (6 cases).

After modified-Duhamel procedure we noticed the following complications [11]: 5 cases of minor rectal bleeding, subocclusive symptoms do to remnant septum with subsequent fecaloma formation in the rectal ampula in 4 cases, repeated enterocolitis in 3 cases, mechanical occlusion or adhesion in one case.

In patients who underwent Soave procedure we encountered 9 cases of significant perianal rash, and 7 cases with night soiling. We must mention that night soiling cannot be attributed to the technique itself, but to the length of the aganglionic segment and/or to the re-do surgery situation in 5 patients.

Analyzing the total/subtotal colonic HD cases that were operated using Soave technique we noted a medium frequency of stools in the first month after surgery of 7-15/day. Nevertheless, the frequency of stools decreased gradually in all cases as in after two years follow-up the frequency reduced to 4-6/day. In this group we noted 2 cases having postoperative enterocolitis.

Despite the limited number of De La Torre Mondragon patients, we can affirm that this procedure gave us the best results, with only one complication reported – anastomotic stenosis successfully treated by serial dilatations.

Discussions

The rectum is always affected in HD, but the length of the additional involved proximal situated bowel varies widely [7].

In most of the cases, contrast enema, as the traditional way of diagnosis, may help the surgeon not only to presume the diagnosis of HD, but also to estimate the length of the aganglionic segment. In particular situations, as in neonates, in operated patients and in total aganglionosis forms, the diagnostic value of contrast enema diminishes considerable. This is the reason why the aspirative rectal biopsy has become a gold standard for the HD [8]. The procedure can be done at the bedside or in an ambulatory setting without the need for general anesthesia. Adequate tissue is obtained for analysis in the majority of patients. Repeated suction biopsies or full-thickness biopsies can be performed if the initial biopsy is equivocal.

A study made in 2005 [10] presents the sensitivity and specificity of the three main diagnosis methods used in HD (see table 1). It mentions rectal suction biopsy with a sensitivity of 93% and a specificity of 100%, contrast enema with a sensitivity of 76% and a specificity of 97% and rectal manometry with a sensitivity of 83% and specificity of 93%.

Other studies report a variability in contrast enema test, with a sensibility that can vary from 65% to 80% and a specificity of diagnosis from 65% to 100% [3, 6]. The normally innervated proximal colon may undergo progressive dilation, while anganglionic region may have a grossly normal or contracted appearance but it isn't effective in all forms of the disease – for example, in a total colonic the diagnosis is difficult to establish using radiologic

studies. Most cases are limited to the rectum and sigmoid colon [6, 7]. The length of the aganglionic small bowel and the age of the patient can influence the radiological findings in total colonic HD. The transitional zone can be false-positive in total colonic HD. The colon can appear normal. Total colonic HD may be assumed if the contrast enema study is normal but the patient remains symptomatic and other causes of distal bowel obstruction have been excluded [5]. On the other hand the barium enema is a good screening test for HD since it correlates with rectal biopsy as confirmation diagnosis [6].

In the investigation of neonates with functional large-bowel obstruction and older children with severe constipation, rectal biopsies remain the main diagnosis method. In our department we've used a RBI2 suction rectal biopsy device without manometric control. This technique's principle is getting small amounts of submucosa by direct suction of superficial rectal layers using an aspiration endorectal device. The correct gathered submucosa is sent to the Pathology Department for histological examination. The procedure can be done at the bedside or in an ambulatory setting without the need for general anesthesia. Adequate tissue is obtained for analysis in the majority of patients. Repeat suction biopsies or full-thickness biopsies under general anesthesia can be performed if the initial biopsy is equivocal [8]

An important mention would be that recent studies are trying to include full colonoscopy as a useful tool in determining the transition zone in transanal endo-rectal pull-through in HD (De la Torre-Mondragon technique) but none of them are conclusive [9].

Conclusions

Introducing rectal suction biopsy in our clinic made possible reducing the age of intervention due to the early diagnosis of the disease and also the necessity of colostomies in common type and short HD cases. The relatively small percentage of positive results obtained in our series (78.6%) may be attributed to the learning curve of the procedure.

Frozen tissue histopathological examination should be a mandatory intraoperative step in evaluating the length of the affected colon segment, in order to decide the right spot for the future anastomosis. This quick procedure can prevent re-do surgery and future complications.

Soave procedure has been confirmed as the best choice for high forms of HD and for re-do surgery in misdiagnosed and mistreated cases. All cases of re-do surgery have reported variable night soiling as a complication.

De La Torre Mondragon became in our team the surgical procedure of choice for common type of HD, due to rapid postoperative recovery and lack of complications.

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