Usefulness of laparoscopy for determining the location of transitional zone in patients with inconclusive barium enema for Hirschsprung's disease: a prospective clinical trial

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Objective **The present study aims to evaluate the efficacy of laparoscopy to localize the transitional zone in cases of Hirschsprung's disease (HD) with nondiagnostic contrast enema.**

Summary background data **In the treatment of HD, preoperative ascertainment of the extent of aganglionosis by contrast enema is crucial, as this affects the planning for transanal endorectal pull-through. Patients with aganglionic rectal biopsy with inconclusive barium enema present difficulty in determining the extent for transanal resection as the colon is not directly visualized as compared with transabdominal approach.**

Patients and methods **A study was conducted in our institution on 30 patients fulfilling such criteria with a median age at the time of surgery of 50 months. The location of the transitional zone by intraoperative laparoscopy view was used to plan further management.**

Results **The junction between normal dilated bowel with peristalsis and aganglionic collapsed bowel without peristalsis was marked by diathermy followed by transanal endorectal pull-through in the same setting in 14 (46.7%) cases. Subsequent histopathological examination of the excised specimens showed adequately ganglionated surgical margin in all 14 cases. In three (10%) patients, the transitional zone was seen in the most distal part of the rectum and the patients were diagnosed as suffering from very short-segment HD; therefore, anorectal myectomy was performed. In the remaining 13 patients no definite funnel was seen and leveling biopsy was done to exclude total colonic HD. Since frozen section biopsy was not available in our facility at the time of this study, all biopsies were sent for fixed paraffin sections. Further management for biopsied patients at later stage was in the form of either anorectal myectomy in five (16.7%) patients or Botox injection in eight (26.7%) patients.**

Conclusion **Laparoscopy was found to be a useful diagnostic and therapeutic tool for such patients.** ***Ann Pediatr Surg*** **14:146–150 © 2018 Annals of Pediatric Surgery.**

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Keywords: Hirschsprung's disease, laparoscopic biopsy, transanal endorectal pull-through, transition zone, ultra-short segment

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Introduction

Today, Hirschsprung's disease (HD) is treated using one-stage transanal endorectal pull-through (TEPT), either with [1–3] or without [4–6] laparoscopic assistance. Short-segment aganglionosis occurring distal to the sigmoid colon is a good indication for TEPT without laparoscopic assistance or laparotomy, while long-segment aganglionosis proximal to the descending colon is better treated using TEPT with laparoscopic assistance or laparotomy [7–9]. Since these surgical procedures are planned based on the extent of aganglionosis, accurate preoperative determination of this extent is crucial [7–10]. In some cases, the recognition of the transition zone might be challenging on contrast enema. A landmark by which the location of pull-through can be ascertained relatively accurately is thus required. We perform laparoscopy to mark the junction between the dilated peristaltic bowel and the collapsed nonperistaltic aganglionic bowel that is not clarified by preoperative contrast enema.

Patients and methods

During the period between September 2011 and June 2013, patients presenting to our institution with clinical presentation suspicious for HD were investigated. As part of the diagnostic workup for HD, we perform full-thickness rectal biopsy under general anesthesia at about 1 cm proximal to the dentate line regardless of the age, in addition to contrast enema to ascertain the radiographic transition zone. In 30 cases (27 males, three females), contrast enema was unclear concerning the transitional zone location (Fig. 1), despite confirming HD by rectal biopsy. In all cases, the pathologist confirms that the included specimens were of colonic origin with absence of ganglion cells from the intramural nerve plexuses, submucosa, and intermuscular regions with thickening of neural axon bundles. All biopsies were read by a pathologist experienced in pediatric colorectal diseases. After approval from the research ethics committee of Faculty of Medicine, Cairo University, patients were scheduled for intraoperative laparoscopy to define the location of the transitional zone. Median age at the time of surgery was 50 months (range: 4–96 months). Written informed consents were obtained from the parents prior to the surgery. Intraoperative findings, further management, and any encountered morbidity were reported.

The procedure followed steps similar to laparoscopic-assisted pull-through originally described by Georgeson *et al.* [1].

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The child was placed in LloydDavies position, cross table with table extension by table side arm extenders in children beyond infancy. When optimal muscle relaxation was guaranteed under general anesthesia, a pressure of 8–10 mmHg and flow set at 2 l/min in children below 1 year of age and at 4 l/min in children beyond that age were used. Three ports were used initially for marking, with additional fourth port added for biopsy taking or mobilization.

With the camera tower opposite the foot of the patient and surgeon standing at patient's head, the first cannula (5 mm diameter trocar) was inserted into the umbilical scar to allow for CO_2 insufflation and 30° telescope for vision. The second (the right one), in the right lower quadrant, was used for the surgeon's right-handed instrument. The third (the left one), in the left lower quadrant in a mirror image to cannula 2, was used either by the surgeon's left hand in case of marking or for the assistant to grasp the bowel segment to be biopsied. The fourth cannula was inserted halfway between the xiphoid and the umbilicus, in the middle of the epigastrium and slightly to the right in small infants but more to the right and lower down in older children and was used for the surgeon's left-handed instrument or the assistant during biopsy taking or mobilization.

Usually, it is immediately clear whether the patient had classic funnel or not. The bowel wall in the transitional zone became progressively thickened toward the normoganglionic segment. In case of HD limited to rectum marking by diathermy for transitional zone was done followed by shift to classic TEPT (Fig. 2); otherwise, for rectosigmoid cases mobilization was done.

If the intraoperative appearance was misleading and the proximal extension of the disease cannot be delineated, pathological examination of serial seromuscular biopsies was done to determine the level of aganglionosis. These biopsies were taken with a Kelly forceps in one hand and a pair of scissors on the other (Fig. 3) from the low rectum, rectosigmoid, mid-transverse colon, and the right colon. In some cases, the rectosigmoid biopsy was taken by pulling the colon through the assistant port on the left side, holding it with stay sutures and taking the seromuscular biopsy extracorporeally (Fig. 4). The biopsy sites were marked by nonabsorbable 3/0 silk sutures (Fig. 5).

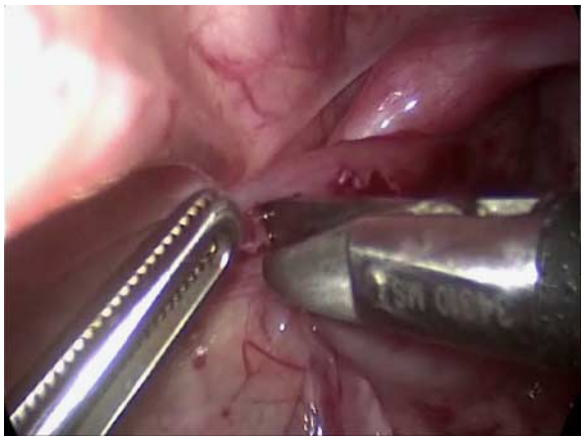
Fig. 1

Patient with inconclusive barium enema and aganglionic rectal biopsy.

Fig. 2

Intraoperative laparoscopic view for the extent of disease with marking site of transitional zone.

Fig. 3



Laparoscopic intracorporeal extramucosal biopsy taking.

Results

The transitional zone was visualized by laparoscopy in 17 (56.7%) patients. However, in the remaining 13 (43.4%) patients laparoscopy failed to identify the transitional zone. In 14 (46.6%) patients, the transitional zone was distal to the sigmoid colon denoting rectosigmoid aganglionosis. Therefore, TEPT was performed after laparoscopic diathermy marking immediately distal to the presumed anastomotic site followed by mobilization of the transitional zone. Nevertheless, the transitional zone was distal to the peritoneal reflection as denoted by hypertrophy and prominent vascularity of the proximal third of rectum in the remaining three (10%) patients. As a result, those patients were subjected to anorectal myectomy.

In patients with unclear transitional zone leveling biopsies were done to exclude total colon aganglionosis. Unfortunately, due to lack of frozen section at our institution during the study time period, all biopsies were sent for analysis by fixed paraffin sections and further management was postponed (Table 1).

In patients subjected to TEPT, subsequent histopathological examination of excised specimens proved HD, in addition to adequately ganglionated surgical margin in all 14 cases. For patients who underwent laparoscopic leveling biopsies (13 patients), histopathological results showed ganglion cells in all laparoscopic biopsies. The cause of constipation in this group was unclear. According to the grade of constipation those patients were further subdivided (Table 2):

(1) Group A: patients with constipation more than grade 1 (grade 2 or 3). Those patients were subjected to anorectal myectomy.
(2) Group B: patients with grade 1 constipation. For such patients, intrasphincteric Botox injection at a dose of 25 IU per quadrant was applied in four quadrants at the level of dentate line.

We termed the patient as responding if the child had at least one motion daily; otherwise, the patient was marked as relapsed. The mean follow-up period was 3.5 months. Unfortunately, two patients from group B were lost during the follow-up.

Fig. 4

Extracorporeal seromuscular biopsy through the left side port.

Fig. 5

Marking biopsy sites.

Discussion

HD is a developmental disorder of enteric nervous system characterized by absence of ganglion cells in the myenteric and submucosal plexuses along variable portion of the distal intestine. The target of the surgical management of HD is to remove the aganglionic segment of the bowel and bring normally innervated

Table 2 Summarizes further interventions done in patients with ganglionated laparoscopic biopsy

	Total	Responded [*n* (%)]	Relapsed [*n* (%)]	Drop outs [*n* (%)]
Group A	5	3 (10)	2 (6.7)	–
Group B	8	3 (10)	3 (10)	2 (6.7)

Table 1 Demonstrating the various interventions done laparoscopically in the patients studied

Laparoscopic finding	Final diagnosis	Procedure done	*n* (%)
Normal looking colon all through (no identified transitional zone)	Suspected total colonic aganglionosis	Leveling biopsy	13 (43.3)
Visible funnel at distal rectum	Rectosigmoid aganglionosis	TEPT	14 (46.6)
Funnel distal to peritoneal reflection	Short-segment rectal aganglionosis	Anorectal myectomy	3 (10)

TEPT, transanal endorectal pull-through.

part to the anus with preservation of anal sphincter function [11].

Indeed, patients with misleading contrast enema represent a challenge to the surgeon. The majority of short and very short-segment HD represents a problem in their diagnosis and management. Those patients suffer from symptoms of HD; whereas in most cases the barium enema study is not conclusive [12]. Such patients were intentionally selected in our study to try to stratify management and to depict essentiality of laparoscopy for those patients. It was noted that a barium enema could not always demonstrate a definite transitional zone in every patient [13,14]. Location of the radiographic transition zone as assessed by contrast enema with reversed rectosigmoid index matches aganglionic bowel in 62.5–79% of all cases [8,9]. This figure is much lower for long-segment or total colon aganglionosis, at 25–31% [8,9]. The transition zone is undetected in 11–27.3% of all cases, and this is most troublesome, since about 10% of cases diagnosed as short-segment aganglionosis according to contrast enema actually display long-segment or total colon aganglionosis [10].

Thus, in our study, case selection was to target mainly cases with nonconclusive barium enema to evaluate the role of laparoscopy in diagnosing cases with short and very short-segment HD by either direct intra-abdominal visualization of the transition zone or the hypertrophied proximal colon with its prominent vascularity denoting distal obstruction, or to take biopsies in case laparoscopy failed to reveal any hypertrophied proximal bowel.

The correct location of the transitional zone is important to obviate an improper pull-through surgery. In our experience, the transitional zone could usually be recognized by the picture of hypertrophic intestinal wall under a laparoscopy-magnified view, even in small babies.

No radiographic transition zone was visualized in all cases. However, in 17 out of the 30 (56.6%) patients, the transitional zone finding was ascertainable by laparoscopy. Based on these findings, patients were diagnosed as either short-segment aganglionosis and treated by laparoscopic-assisted endorectal pull-through in 14 cases or very short-segment HD in which anorectal myectomy was done in the same setting in three cases. Pathological and intraoperative findings were matched for all patients. All patients showed uneventful postoperative period with improvement of symptoms.

While in 43.3% cases laparoscopy failed to show the macroscopic appearance of the transitional zone, it was a guide to another entity of patients with very short HD that could be improved by Botox injection. Improvement of symptoms occurs in 20% of patients either by anorectal myectomy or Botox injection. Despite the fact that all cases included in the study were subjected to preoperative rectal biopsy, which proves to be aganglionic in all of them, three (10%) cases out of the total 30 cases show improvement on Botox injection having motion once daily. This shows another benefit of laparoscopy; otherwise, those patients would be subjected to unnecessary pull-through or anorectal myectomy procedures with its complications such as incontinence and anastomotic stricture.

Sauer *et al.* [15] have proposed obtaining seromuscular biopsies through an open transumbilical incision instead of laparoscopy. Although this allows histopathologic guidance of the extent of resection, some segments of the bowel, such as the rectum, may not be accessible by this technique. Furthermore, dissection of the mesentery for mobilization is not feasible [15]. We were able to replicate such technique to obtain high rectal biopsy in small infants using left side port, which offers better cosmesis. Yamataka *et al.* [16] reported suction colonic biopsy with laparoscopic assistance as a technique to determine the location of pull-through, but this technique is limited to short-segment aganglionosis and is technically complex.

Conclusion

Although diagnosis of HD can be achieved in more than 95% of cases using rectal biopsy [16], the accurate delineation of transitional zone using contrast enema represents a diagnostic challenge, especially in children with short-segment aganglionosis. Therefore, the use of intraoperative laparoscopy for such cases is highly recommended. Nevertheless, a larger sized sample and longer follow-up period is needed for more reliable results.

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Conflicts of interest

There are no conflicts of interest.

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