Evaluation of Transanal Endorectal Pull-Through Results

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Abstract

Background: Harold Hirschprung, a Danish pediatrician, is credited with the first definitive description of the disease that bears his name. Over the next 30 years, poorly understood procedures that were intended to reduce sympathetic hyperactivity of the colon were reported to be, at least temporarily, successful in treating HD. Complications can be classified as either early (weeks to months) or late (months to years). The transanal endorectal pull-through approach represents a major revolution in treating Hirschsprung's disease.

Objectives: The aim of this study was to evaluate the outcome of transanal endorectal pull-through operation clinically and radio logically in patient with Hirschsprung's disease.

Patients and Methods: This prospective quasi-experimental cohort study was conducted on 40 children with Hirschsprung's disease operated beyond 2 years, in whom transanal endorectal pull-through was indicated. For each patient, the following was done prior to the surgical procedure: Full clinical history taking, Full physical examination, Laboratory investigations, Radiological examination and rectal biopsy.

Results: Our results showed that; As regards abdominal distension, 14 cases showed no distension, 24 cases showed mild distension and 2 cases showed moderate to severe abdominal distension. History of previous attack of enterocolitis was positive in all cases. Fourteen patients (35%) had a normal defecation. Fourteen patients (35%) had a frequency of defecation of 3-5 days. Eight patients (20%) had a frequency of defecation of 6-7 days. Four patients (10%) had a frequency of defecation of 8 or more days. Thirty two patients had true fecal incontinence and had a non-dilated colon on contrast enema and a tendency to have diarrhea and did not respond to medical treatment. Four patients had partial fetal incontinence and no patients had incontinence. Postoperative soiling accidents were observed occasionally in 4 patients (10%), often in 18 patients (45%). Perianal skin excoriation was absent in 36 patients (90%) and present in 4 patients only (10%). Rectal sensation was preserved in 30 patients (75%).

Conclusion: Transanal Endorectal Pull-Through (TEPT) is a good technique for surgical correction of Hirschsprung's disease with few operation-related complications. The transanal procedure needs a special experience and it has to be born in mind that laparotomy may be resorted to any time if any difficulty was encountered during the procedure. The clinical outcome is satisfactory. Many children with Hirschsprung's disease have a good outcome following surgical treatment, but long-term follow-up studies have identified a number of concerns. The predominant cause for persistent or recurrent unresponsive obstructive symptoms after initial pull-through procedure is incomplete resection of the transition zone. Fecal incontinence after operative management of Hirschsprung's disease represents a serious problem. To avoid incontinence after transanal endorectal pull-through, we recommend the following: overstretching of anal canal, very low colo-anal anastomosis, injury to sensory mucosa, injury to mussels and inadequate resection of dilated colon. The majority of long-term follow-up studies have concentrated only on the functional outcome. Although most patients had good or fair quality of life after surgical correction for Hirschsprung's disease, the long-term outcome and quality of life are not as good as surgeons expected. Satisfaction of the parents with the results is a subjective object varies with the socioeconomic levels of the parents, their education and their residency.

Keywords: Transanal Endorectal Pull-through Hirschsprung; Anal canal; Sbdominal distension

Introduction

Hirschsprung (HD) Disease is a relatively common neonatal developmental disorder of the enteric nervous system. It is characterized by the absence of ganglion cells in the myenteric and...
sub mucosal plexuses of the distal intestine. This results in absent peristalsis in the affected bowel, and the development of a functional intestinal obstruction. The pathogenesis and genetic basis of the disease is yet unclear [1]. The surgical management of Hirschsprung Disease (HD) has developed, from full-thickness recto sigmoid dissection (Swenson and Bill), an endorectal dissection (Soave) and retro rectal pouch procedure (Duhamel), to more recently a primary repair that can be done transanally [2]. Langer et al. [3] described the one-stage transanal pull-through operation for Hirschsprung’s disease. Since then, the procedure has become increasingly popular and compared with the traditional open laparotomy, the advantages of the operation are that it is minimally invasive, it eliminates the abdominal incision, so there is no abdominal scar, and it avoids complications of traditional laparotomy such as intestinal adhesion and wound infection postoperatively. The operating time and hospital stay are also shortened. The complications occurring after the surgical repair of Hirschsprung’s disease can be categorized into early and late complications. However, there is significant overlap in regard to the time period during which these may occur [4]. The early complications include wound infection, bleeding and anastomotic complications as (leak, pelvic abcess, retraction of pull-through segment) [4].

The late complications are usually present within the first few postoperative months. They include: enterocolitis, stricture and constipation [5]. In another review of the patients undergoing Swenson procedure, Kim et al. [6] noted that continence as Fecal soiling has the greatest negative impact on the quality of life in children with Hirschsprung’s disease, 12% of their patients less than 5 years of age had some degree of soiling, however between 10 and 15 years age its declined to only 6%. The incidence of soiling decreased from 8% at 5 years old follow-up to less than 2% at 20 years old follow-up. Finally, for post-operative Duhamel patients, they noted that only 5.3% showed evidence of soiling. Rates of fecal incontinence post pull-through for HD ranges from 3% to 53% at long term follow-up only 10.3% had fecal incontinence. The anorectal function are greatly affected after the surgical treatment of the Hirschsprung’s disease. It can be assessed clinically by interviews and questionnaires to the patients and the parents. It also can be assessed by investigations as endorectal ultrasound, anorectal manometer, Colonic transit time and defecography [7]. The aim of this study is to evaluate the outcome of transanal endorectal pull-through operation clinically and radio logically in patient with Hirschsprung’s disease.

**Patients and Methods**

This prospective quasi-experimental cohort study was conducted on 40 children with Hirschsprung’s disease operated beyond 2 years, in whom transanal endorectal pull-through was indicated.

**Inclusion criteria**

- Patients with histopathologically documented Hirschsprung disease operated on by transanal endorectal pull-through in the last 5 years.
- Transitional zone in the rectosegmoid colon (classic type of Hirschsprung’s disease) diagnosed by contrast enema.

**Exclusion criteria**

- Patients operated in the last 2 years.
- Previous colorectal surgery for Hirschsprung’s disease other than rectal biopsy.
- Associated congenital syndromes (e.g. Down syndrome).
- Neurological impairment (e.g. menengiomyelocele).

**Methods**

For each patient, the following was done prior to the surgical procedure:

- Full clinical history taking.
- **Full physical examination:** General condition, weight, abdominal examination: abdominal distension, palpable colon, and rectal examination.
- **Laboratory investigations:** CBC, coagulation profile, liver and renal function tests and CRP.
- **Radiological examination:** Plain X-ray of the abdomen (erect and supine) for suspected HAEC and Contrast [Barium or Gastrograffin (GG)] enema.
- Rectal biopsy.

**Evaluation criteria**

- Age at presentation.
- Age at surgery.
• Associated major congenital anomalies.
• Defecation frequency per Day.
• Rectal sensation is persevered or not.
• Soiling of clothes in between defecation times.
• If incontinence present, It is for stool only, or flatus only or both.
• Number of attacks of enterocolitis in general.
• Number of attacks of enterocolitis required hospital admission.
• Abdominal distension.
• Faecal Impaction.
• Perianal skin excoriation.
• Anal sphincter tone.
• Feeding.

Results

We followed up 40 pediatric patients with Hirschsprung's disease, in whom transanal endorectal pull-through was indicated. There were 32 males and 8 females. Mean age was 9.2 months (Figure 1 and Table 1). As regards abdominal distension, 14 cases showed no distension, 24 cases showed mild distension and 2 cases showed moderate to severe abdominal distension (Figure 2). Fourteen patients (35%) had a normal defecation. Fourteen patients (35%) had a frequency of defecation of 3-5 days. Eight patients (20%) had a frequency of defecation of 6-7 days. Four patients (10%) had a frequency of defecation of 8 or more days (Figure 3). Thirty two patients had true fecal continence and had a non-dilated colon on contrast enema and a tendency to have diarrhea and did not respond to medical treatment. Four patients had partial fetal incontinence and no patients had incontinence (Figure 4).

Postoperative soiling accidents were observed occasionally in 4 patients (10%), often in 18 patients (45%). Soiling was not observed in 18 patients (45%) (Figure 5). Perianal skin excoriation was absent in 36 patients (90%) and present in 4 patients only (10%) (Figure 6). Rectal sensation was preserved in 30 patients (75%) (Figure 7).

Discussion

Hirschsprung’s disease is one of the most common congenital malformations, which occurs in 1:2000 to 5000 live births. There are three ways that Hirschsprung's disease characteristically presents neonatal bowel obstruction, chronic constipation, and enterocolitis [8]. The treatment of Hirschsprung's disease is primarily surgical. Surgical therapy for Hirschsprung's disease implies removal of aganglionic bowel and bringing of normally innervated intestine to the anus. Although all of the three most commonly performed basic techniques for the correction of Hirschsprung's disease (Swenson, Soave and Duhmel) accomplish this goal, the modified Soave operation has the important advantage of helping to avoid injury to the pelvic nerves responsible for urinary incontinence and sexual dysfunction [9]. The use of transanal approach is relatively a new concept. The risks of contamination and adhesion formation are eliminated. The procedure does not damage the pelvic structures, is not expensive, and has the most optimal cosmetic result [10]. The aim of this study is to evaluate the outcome of transanal endorectal pull-through operation clinically and radio logically in patient with Hirschsprung’s disease. The study was performed on 40 patients with Hirschsprung disease operated on by transanal endorectal pull-through in the last 5 years. Zhang et al. [11] performed long-term follow up of 73 children who had undergone surgical correction of Hirschsprung’s disease with transanal-one stage pull-through operation, of these children, 58 responded and 15 were lost to follow up; the losing rate was 20%. We exclude patients with Hirschsprung's disease operated in the last 2 years. Fourteen patients (35%) had a normal defecation. Fourteen patients (35%) had a frequency of defecation of 3-5 days. Eight patients (20%) had a frequency of defecation of 6-7 days. Four patients (10%) had a frequency of defecation of 8 or more days. Thirty two patients had true fecal incontinence and had a non-dilated colon on contrast enema and a tendency to have diarrhea and did not respond to medical treatment. Four patients had partial fetal incontinence and no patients had incontinence. As regards abdominal distension, 14 cases showed no distension, 24 cases showed mild distension and 2 cases showed moderate to severe abdominal distension. History of previous attack of enterocolitis was positive in all cases. Hadidi et al. [12] reported a 4.4% enterocolitis rate utilizing the transanal approach in comparison to a 12% incidence in a preceding series of open pull-through procedures. This is very low incidence of post pull-through enterocolitis which differs from most of studies reported in the literature. The rate of enterocolitis varies widely in published reports, from a higher rate of 54% by Van Leeuwen et al. [13] to a lower rate of 4.55% in Zhang et al. [11] study of which may attribute to criteria of enterocolitis used in different studies.

Postoperative soiling accidents were observed occasionally in 4
patients (10%), often in 18 patients (45%). Soiling was not observed in 18 patients (45%). Soiling was observed at night in 16 patients (40%) and at day and night in 6 patients (15%). Most episodes of enterocolitis occur within the first two years after the pull-through procedure. Engum et al. [14] have noted the infants that experience enterocolitis before operative intervention have an increased risk of occurrence of this complication following the pull-through procedure. Zhang et al. [11] studied 58 patients who underwent transanal pull-through operation for Hirschsprung’s disease, the results were soiling in 9 patients (15.5%) and constipation in 5 (8.6%) and no incontinence was obtained in any patient. Indications for the second pull-through include: retained or acquired aganglionosis, severe stricture, dysfunctional bowel segment, marked dilation of the bowel as a result of years of constipation, anocutaneous fistula, and intestinal neuronal dysplasia (Gobran et al., 2007). El-Sawaf et al. [15] studied 41 patients who underwent transanal pull-through operation for Hirschsprung’s disease, there was a 53.7% overall incidence of post-pull-through enterocolitis. This higher rate of enterocolitis may be because of low threshold in diagnosing early cases of enterocolitis. Levitt et al. [16] reported that postoperative enterocolitis was more frequent in patients who had enterocolitis before the pull-through procedure. We explained that satisfaction of the parents with the results is a subjective object varies with the socioeconomic levels of the parents; their education and their residency (rural or urban). Moreover, one other point deserving recognition is that, currently, the purpose of the management of Hirschsprung’s disease is not only to ensure survival, but also to achieve normal bowel function and, more importantly, to achieve and good quality of life in all possible ways. Surgery is not the end of the treatment. We feel it is necessary to emphasize the long-term regular follow up of patients after surgery for Hirschsprung’s disease because it is the prerequisite for improvement of the quality of life. Strategies, including psychological counseling, medication therapy, and toilet training, are needed.

**Conclusion**

- Transanal Endorectal Pull-Through (TEPT) is a good technique for surgical correction of Hirschsprung’s disease with few operation-related complications.
- The transanal procedure needs a special experience and it has to be born in mind that laparotomy may be resorted to any time if any difficulty was encountered during the procedure.
- The clinical outcome is satisfactory.
- Many children with Hirschsprung’s disease have a good outcome following surgical treatment, but long-term follow-up studies have identified a number of concerns.
- The predominant cause for persistent or recurrent unresponsive obstructive symptoms after initial pull-through procedure is incomplete resection of the transition zone.
- Fecal incontinence after operative management of Hirschsprung’s disease represents a serious problem.
- To avoid incontinence after transanal endorectal pull-through, we recommend the following: overstretching of anal canal, very low colo-anal anastomosis, injury to sensory mucosa, injury to mussels and inadequate resection of dilated colon.
- The majority of long-term follow-up studies have concentrated only on the functional outcome.

- Although most patients had good or fair quality of life after surgical correction for Hirschsprung’s disease, the long-term outcome and quality of life are not as good as surgeons expected.
- Satisfaction of the parents with the results is a subjective object varies with the socioeconomic levels of the parents; their education and their residency.

**References**

