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Hirschsprung's Disease: a Comparison of Swenson's and Soave's Pull-through Methods

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Abstract

Background Hirschsprung's disease or the congenital intestinal aganglionosis is the result of arrested fetal development of the myentric nervous system, but the precise pathogenic mechanisms involved are unknown. The successful treatment of infants and children with Hirschsprung's disease depends on prompt diagnosis and early treatment.

Objective To compare the complications of Swenson's and Soave's pull-through procedures for the management of Hirschsprung's disease.

Methods This study was conducted by patient's relative interview in Central Teaching Hospital of Pediatrics in Baghdad for the period from June 2006 to June 2010 .All the patients were under (10) years old who were admitted and underwent surgical interference and followed up under the study group. This study included 40 infants and children with Hirschsprung's disease underwent either Swenson's or Soave's pull-through procedures. On average, the post operative follow-up period was six months after the last stage of operation (closure of colostomy).

Results The patients consisted of 32 males (80%) and eight females (20%), a ratio of 4:1. Their age ranged from 1 day to 10 years. History of failure to pass meconium was the commonest presentation and was found in (87.5%). Twenty-five patients (62.5%) underwent a Swenson's pull-through and fifteen patients (37.5%) underwent Soave's pull-through. The incidence of the complications following Swenson's procedure was 24% while after Soave's procedure was 20%. Wound infection and adhesive intestinal obstruction were the commonest complications after Swenson's pull-through and occurred in (12%) while anastomatic stricture was the commonest complication after Soave's procedure and occurred in (20%).

Conclusion The rate of complications was higher following Swenson's pull-through in comparison to Soave's pull-through procedure.

Keywords Hirschsprung's disease, Swenson's pull-through, Soave's pull through, complications.

Introduction

Harold Hirschsprung's, a Danish pediatrician is credited with the first definitive description of the disease in 1888 that bears his name [(1)]. Hirschsprung's disease or congenital megacolon is a developmental anomaly caused by migratory failure of neural crest cells. When these primitive neurogenic cells fail to take up positions in the Submucosal and intermyenteric plexus of the bowel from lips to anus, motility disturbances result; that most routinely present as chronic constipation in a newborn child [(1-4)].

Ninety-eight percent of babies pass meconium within 24-48 hours of birth. In babies with Hirschsprung's disease 90% fail to achieve this

passage. The disease progresses to abdominal distension, bilious vomiting, and possibly obstructive enterocolitis. If a neonate leaves the hospital without diagnosis, he will generally reappear with chronic constipation within two years. This constipation often accompanies a dietary change such as the change from breast milk to formula or formula to solid foods. Rarely, children escape diagnosis until more advanced ages when chronic constipation and failure to thrive are seen. The most common physical findings are abdominal distension, visible bowel loops with peristalsis, and poor muscle development secondary to poor nutrition. Rectal examination reveals a spastic rectum with no or little stool because stool is high above the spastic colonic segment.

When the clinical suspicion of Hirschsprung's disease has been raised, plain abdominal radiographs in the anteroposterior projection or in left lateral decubitus position are obtained to look for evidence of intestinal obstruction or free intraperitoneal air or both. This study can be followed by unprepared barium enema or anorectal manometry and full thickness or suction rectal biopsy. The specimen must be obtained at least 1.5 cm above dentate line [2-5].

Surgery is the only mode of treatment for Hirschsprung's disease. The child usually has a colostomy, which was placed several months previously. This colostomy decompresses the bowel and returns it to normal caliber. The definitive operation generally is performed when the child is 6 to 12 months old. Over the past 50 years, different methods of varying efficacy have been adopted to correct the underlying abnormality. The first definitive operation was described by Swenson's and Bill in 1948. This procedure involves resection of aganglionic colon bowel and anastomosis of the distal rectum to ganglionated colon by combined abdominoperineal approach. Essential to this operation is maintenance of dissection immediately adjacent to the rectal wall to avoid injury to the pelvic nerves responsible for rectal and bladder innervations and sexual function. Soave's method involves removing diseased mucosa and submucosa, followed by pulling the ganglionated intestine through the muscular cuff and end to end coloanal anastomosis [5-8].

Enterocolitis of Hirschsprung's disease remains the major cause of significant morbidity and mortality today. The entity is manifested clinically by explosive diarrhea, abdominal distension, and fever. Pathologically, enterocolitis is defined as an acute inflammatory infiltrate into crypts and mucosa of either the colonic or the small intestinal epithelium [8,9].

Methods

Forty patients with a diagnosis of Hirschsprung's disease were admitted to Central Teaching hospital of pediatrics between 2006 and 2010. A standardized data sheets were prepared for collection of information including age, sex, body weight, and natal history, family history, age at presentation, associated anomalies, methods of diagnosis, type of the surgical procedure performed for treatment and post-operative complications. All our patients underwent three staged surgical procedures including colostomy which was placed in the ganglionic segment of colon as the first stage in order to decompress the colon and return it to normal caliber while the definitive surgical procedure done in the second stage after six months from the time of colostomy. The last stage is closure of colostomy which was done after two months from definitive surgery. On average, the post-operative follow-up period was six months after the closure of colostomy (third stage).

Results

During four years period from June 2006 to June 2010; forty cases of Hirschsprung's disease (congenital megacolon) were admitted to the hospital. There were 32 male (80%) and eight female (20%) patients, with male: female ratio being 4:1. Their ages ranged from 1 day to 10 years. Table 1 shows age distribution of the studied sample.

Table 1. Age distribution of the studied sample

Age	Number of patient	%
< 28 days	7	17.5%
28 days -1 year	23	57.5%
1 year – 6 years	7	17.5%
> 6 years	3	7.5%

History of delayed passage of meconium in the first 48 hours of life was the most common mode of presentation, occurring in 35 patients (87.5%), followed by bile stained vomiting in 25 patients (62.5%), while 28 patients(70%) had abdominal distension and 30 patients (75%) presented with constipation. Other modes of presentation include fecal impaction (10%), Encopresis (5%) ad Enterocolitis (5%) as noticed in table 2.

Table 2. The mode of presentation of patient with Hirschsprung's disease

Signs and symptoms	Number of patients	%
History of Failure to pass meconeum	35	87.5%
Bile-stained vomiting	25	62.5%
Abdominal distension	28	70%
Constipation	30	75%
Fecal impaction	4	10%
Encopresis	2	5%
Enterocolitis	2	5%

Rectal biopsy was performed in 37 patients, and three patients had been referred from other hospitals with rectal biopsy already done. All biopsies were performed 2-3 cm above the dentate line, four samples were initially reported as inconclusive, but the patients continued to have symptoms and biopsy was repeated, all these samples showed the absence of ganglion cells. No morbidity or mortality was associated with these biopsies during the follow-up period. Barium enema was performed in all our patients (even in patients with acute intestinal obstruction where Ba-enema done two weeks after creation of stoma) and was diagnostic only in thirty patients (75%) showing spastic distal segment. Down syndrome was the most common associated anomaly and was present in six patients (15%).

Congenital heart disease was present in four patients (10%), and congenital hip dislocation was observed in three patients (7.5%).Other associated anomalies malrotation (7.5%), inguinal hernia (5%), umbilical hernia in(5%), annular pancreas (2.5%), and ileal atresia in (2.5%). Long segment Hirschsprung's disease (extending to colon) was the most common type as shown in table 3.

Table 3. Types of Hirschsprung's disease according to extension of aganglionic cells (independent on Ba-enema and colonic biopsies).

Surgical procedure	No. of patients	%
Swenson's pull-through	25	62.5%
Soave's pull-through	15	37.5%
Total	40	100%

Twenty-five patients (62.5%) underwent Swenson's pull-through and fifteen patients (37.5%) underwent Soave's pull-through, based on surgeon preference and operative finding if there is adhesion between mucosal and muscular layers we prefer to do Swenson's pull-through and this is usually happened in those patients who had long history of presentation with recurrent enterocolitis, while if there is no adhesion and Submucosal dissection coming easy we prefer to do Soave's pull- through which usually occur in younger age and with those who had short history of presentation.

Table 4. The surgical methods used in treating Hirschsprung's disease

Type	No. of patients	%
Short segment(localized to rectosigmoid)	11	27.5%
Long segment (extend to colon)	28	70%
Total colonic aganglionosis	1	2.5%
Total	40	100%

The incidence of complications following Swenson's pull-through was 24% and that after Soave's pull-through was 20%. The most common complications after Swenson's pull-through were wound infection and adhesive intestinal obstruction (12%), one of those patient with adhesive intestinal obstruction not responding to conservative treatment and underwent surgical treatment.

Anastomatic stricture was most common complication after Soave's pull-through and occurred in three patients (20%) all of them responding to the frequent dilatation under general anesthesia. Enterocolitis was most common after Soave's pull-through (13.4%) in compare to Swenson's pull-through (8%) all of them were admitted to hospital and treated with bowel rest and parental antibiotics. Two patients (8%) continued to be constipated after Swenson's pull-through while no patient remained constipated after Soave's pull-through (Table 5).

Table 5: Complications after pull-through method

Complication	Surgical procedure	
	Swenson's pull-through	Soave's pull-through
Wound infection	2(4%)	1(6.7%)
Anastomatic leak	1(4%)	0
Anastomatic stricture	2(8%)	3(20.1%)
Adhesive intestinal obstruction	3(12%)	1(6.7%)
Constipation	2(8%)	0
Fecal incontinence	1(4%)	0
Enterocolitis	2(8%)	2(13.4%)
Voiding dysfunction	2(8%)	0

Voiding dysfunction occurred in two patients (8%) after Swenson's pull-through while no patients developed such problem after Soave's pull-through. All those patients with voiding dysfunction treated with urinary bladder catheterization for one month; then intermittent catheterization done for another one month until they become with normal urination.

Discussion

Since 1888, when Harold Hirschsprung's presented his classical description of congenital megacolon for the first time, numerous approaches have adopted for the diagnosis and surgical treatment of Hirschsprung's disease. In this study, delayed passage of meconium was the most common presentation (87.5%) followed by bile-stained vomiting (62.5%) which similar to O'Donovan et al [5], but differ from Taxman et al (70%) [6], Stockman and Philippart (80%) [7]. Barium enema and rectal biopsy used as diagnostic procedures, barium enema was diagnostic in 75% showing spastic distal colonic segment and this result was similar to O'Donovan et al (77%) [5], but differ from Taxman et al [6] (80%) and Stockman and Philippart (85%).

We performed rectal biopsy in 37 patients using open method under general anesthesia. The failure in first attempted biopsy happened in 4 biopsies (8%), which is higher than range reported by Andrassy et al [8] (2.3%), but low in comparison to Ghos and Griffen study [9] (14%). This failure rate attributed to technical error resulting in biopsy being taken close to dentate line or too superficially.

In our center, both the multistage Swenson's and Soave's approach were used, with colostomy being performed within one month from the time of rectal biopsy .There were no intraoperative or early post-operative deaths during the period of this study. These results similar to Fortuna et al[10] and Taxman et al (70%) [6] who reported no mortality, but it was differ from those shown by Ikeda and Goto [11] as well as Kleinhaus et al [12] ,where the mortality was 0.3%.

The incidence of complications following Swenson's procedure was 24% whereas after Soave's procedure was 20%, this results are comparable with the series reported by Stockman and Philippart, who observed similar complication rates following Swenson's (22%) and Soave's procedures (18%), whereas Fortuna et al reported higher complication rate after Soave's (30%) and Swenson's (40%) procedures. In our study adhesive intestinal obstruction was commonest complication after Swenson's procedure (12%) while (6.7%) after Soave's procedure, a rate higher than Ikeda and Goto study (8% after Swenson's 4.2% after Soave's procedures).

Anastomatic stricture was more common after Soave's (20.1%) than after Swenson's procedure (8%), which is higher than Kleinhaus et al study [12] (11.3%) after soave's while 5.5% after Soave's procedures). Voiding dysfunction occurred in two patients (8%) who underwent Swenson's pull-through while no patient developed this problem after Soave's procedures and this may be attributed to massive perirectal dissection leading to injury to the pelvic nerves that supply of the urinary bladder in Swenson's procedure, but this result differ from other reported studies.

There was no mortality in our study whereas similar comparative studies (Kleinhaus et al and Ikeda and Goto studies) have shown low mortality after Swenson's (1.8%) and Soave's (2.2%). Although in our study we did three stage pull-through to reduce the incidence of postoperative complications especially Anastomotic leak, recent literatures favor the use of single stage pull-through which eliminates the complications and cost secondary to enterostomy [13-16].

Conclusion

The rate of complications is higher with Swenson's pull-through procedure more than Soave's procedure especially voiding dysfunction, postoperative adhesive intestinal obstruction and fecal incontinence.

References

1. Ashcraft KW, Holocomb GW, Murphy JP. Pediatric Surgery. 5th edition. USA, Philadelphia: Elsevier Saunders, 2010; p. 470-477.
2. Aresman RM, Bambini DA, Almond S. Pediatric Surgery. 2nd edition. USA, Lands Bioscience, 2005, p. 370.
3. O'Neil JA, Micheal Jr, Gauderer WL, Pediatric Surgery. 5th edition. USA, Mosby Year book Inc, 2005; p. 1349.
4. Polly TZ, Coran AG, Wesly JR. A ***t***en year experience with ninety two cases of Hirschsprung's disease. *Ann Surg,* 1998*;* 202: 349-354.

5. O'Donovan A, Harba G, Samers S, Malone DE, Rees A, Winthrop Al: Diagnosis of Hirschsprung's disease. *AM J Roentgenol*, 1996; 167: 517-520.

6. Taxman TL, Yalish BS, Rothestein FC. How useful barium enema in the diagnosis of infantile Hirschsprung's disease? *Am J Dis Child*, 2002; 140: 881-884.

7. Stockman PT, Philippart AI. The Sweson's procedure for Hirschsprung's disease. *Semin Pediatr Surg*, 1998; 7(2): 89-95.

8. Andrassy RJ, Issacs H, Weitzman JJ. Rectal suction biopsy for the diagnosis of Hirschsprung's disease. *Ann Surg*, 1981; 193: 419-424.

9. Ghos A, Griffiths DM. Rectal biopsy in the investigation of constipation. *Ann Dis Child*, 2004; 79: 266-268.

10. Fortuna RS, Weber TR, Tracy TF Jr, Silen Ml. Critical analysis of the operative treatment of Hirschsprung's disease. *Arch Surg*, 1996; 131: 520-525.

11. Ikeda K, Goto S. Diagnosis and treatment of Hirschsprung's disease in Japan: an analysis of 1628 patients. *Ann Surg*, 2000; 199: 400-405.

12. Kleinhaus S, Boley SJ, Sheran M, Sieber WK. Hirschsprung's disease: a survey of the members of surgical section of the American Academy of pedietrics. *J Pediatr Surg*, 1979; 14: 588-597.

13. Bianchi A. One-stage neonatal reconstruction without stoma for Hirschsprung's disease. *Semin Pediatr Surg*, 2002; 7(3): 170-173.

14. Cilly RE, Spitz L, Lazar J, et al: Definitive treatment of Hirschsprung's disease in the newborn with one stage procedure. *J Pediatr Surg*, 1994; 30: 551-556.

15. Wilcox DT, Bruce J, Bowen J, Binanchi A. One-stage neonatal pull-through to treat Hirschsprung's disease. *J Pediatr Surg*, 1997; 32: 243-247.

16. So HB, Becker JM, Schwartz DL. Eighteen years experience with neonatal Hirschsprung's disease treated by endorectal pull-through without colostomy. *J Pediatr Surg*, 2003; 33: 673-675.

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