Management of Hirschsprung's Associated Enterocolitis as a Post-Operative Complication: Single-Center Experience

Ahmed Oshiba[1*], Isooba Safiyo ayub[2], Ahmed khairi gabr[1], Mohamed abdelazim abouheba[1]

[1]Pediatric Surgery Department, Faculty of Medicine, Alexandria University, Alexandria, Egypt.

[2]General Surgery Departement, Hoima Regional Referral Hospital, Ughanda.

***Address for Corresponder:** Dr Ahmed Oshiba, Pediatric Surgery Department, Faculty of Medicine, Alexandria University, Alexandria, Egypt (email: ahmed_eshiba2012@yahoo.com)

How to cite this article:
Oshiba A, Ayoub I, Khairi A, Abouheba M**.** Management of Hirschsprung's Associated Enterocolitis as a Post-Operative Complication: Single-Center Experience. Iranian Journal of Pediatric Surgery 2024; 10 (2): 179-192.

DOI: https://doi.org/10.22037/irjps.v10i2.45743

Abstract

Introduction: Enterocolitis is a potentially fatal consequence of Hirschsprung's disease, it is characterized by fever, abdominal pain, foul-smelling diarrhea, and sepsis. Most neonatal morbidity and mortality are caused by Hirschsprung-associated enterocolitis (HAEC). Harald Hirschsprung initially discovered Hirschsprung's related enterocolitis in the 19th century and included it in his iconic description of megacolon. Trisomy 21, illness, familial history, and prior bouts of HAEC are risk factors for the condition. (3,4) According to Engun et al. patients who presented with a history of Hirschsprung's disease had a 35% incidence of HAEC, but those without such a history only had a 16% incidence. Similarly, HSD and trisomy 21 have a known association with 2.9 - 8.2% of HSD patients also having trisomy 21**.**

received: 8 July 2024
accepted: 27 August 2024
Published online: 15 October 2024

This study aimed to evaluate HAEC as a postoperative complication in children who were admitted after pull-through surgery as regards the type of surgery, incidence, clinical presentation, sepsis workup, and management in Alexandria University Children's Hospital.

Materials and Methods: This retrospective study included 30 patients with Hirschsprung disease who developed post-operative Hirschsprung-associated enterocolitis and were scheduled for surgical intervention from January 2021 to 2022.

Results: In the studied group, the most commonly used pull-through procedure was the Soave (53.33%), followed by the Duhamel procedure (20%). Fewer subjects were operated using the Swenson and Rehbein methods, at 16.67% and 10% respectively. Most of our studied patients (26 patients) presented with signs and symptoms of Grade I HAEC, while 3 patients with Grade II and 1 patient with Grade III.

Conclusion: Post-operative HAEC is a serious complication and needs to proper management to save the child either conservative treatment, repeated anal dilatation or surgical intervention in severe cases.

Keywords

- Hirschsprung-Associated Enterocolitis
- Contrast Enema
- Pull-Through
- Megacolon

Introduction

Enterocolitis is a potentially fatal consequence of Hirschsprung's disease, it is characterized by fever, abdominal pain, foul-smelling diarrhea, and sepsis. Most neonatal morbidity and mortality are caused by Hirschsprung's associated enterocolitis (HAEC). Harald Hirschsprung initially discovered Hirschsprung's related enterocolitis in the

19th century and included it in his iconic description of megacolon. [1]

Researchers have found preoperative enterocolitis risk factors such as delayed diagnosis, long-segment disease, female gender, family history, and trisomy 21. The proximal bowel's physical enlargement, and changes in the mucin's synthesis and component composition, some of the hypotheses proposed to explain its occurrence include rotavirus, Clostridium difficile, elevated prostaglandin E1 activity, mucosal immunity abnormalities, a Schwartzman-type reaction, disordered motility connected to protein, sensitization, and sucrase isomaltose shortage. [2]

Infants are more likely to develop HAEC if Hirschsprung disease is not diagnosed during the perinatal period. According to a study, the incidence of HAEC increased from 11% to 24% among newborns with HSD who received their diagnosis after the first week of life.[1]

Trisomy 21, illness, familial history, and prior bouts of HAEC are risk factors for the condition.[3-4] According to Engun et al.[5] patients who presented with a history of HSD had a 35% incidence of HAEC, but those without such a history only had a 16% incidence. Similarly, HSD and trisomy 21 have a known association with 2.9 - 8.2% of HSD patients also having trisomy 21. Children with Down syndrome and HSD are more likely to develop HAEC, with a 50% incidence.[6-7] Patients with HSD who have trisomy 21 have an HAEC incidence as high as 48%, compared to a 25% rate in those who do not. In 1995, El Halaby et al. found that individuals with long-segment diseases had significantly greater rates of HAEC about 49%.[8]

Since HAEC's cause is unknown, treatment remains important and directed towards combating severe symptoms as well as managing the contributing factors to its pathogenesis. Children with HAEC are treated acutely with Intravenous fluid resuscitation and antibiotics. These patients are put on close hemodynamic monitoring and started on gentamicin, ampicillin, or metronidazole.

Severe HAEC and sepsis patients will require to be admitted to the intensive care unit (ICU), vigorous fluid resuscitation, and, in certain circumstances, these patients need vasopressors drugs and ventilation support. A continuous irrigation system or rectal irrigation using a warm saline solution should be implemented as soon as possible. For those patients who come with sepsis and severe HAEC,

immediate diversion is strongly recommended.[9]

After transanal pull-through, Gao and colleagues[10] reported routine anal dilatation for three months to prevent stricture. According to a different study, medical professionals' weekly anal dilatations reduce the detrimental psychological and social impacts that routine anal dilatations have on families.[10] [relax] the internal sphincter using a less invasive procedure called "chemical sphincterotomy" which involves applying isosorbide dinitrate or nitroglycerine to the anal canal. As a defense against post-pull-through HAEC, these authors promote the repetitive use of topical nitrates. It has been demonstrated that planned rectal irrigations reduce the prevalence of postoperative HAEC.[11]

This study aimed to evaluate HAEC as a postoperative complication in children who were admitted after pull-through surgery regarding the type of surgery, incidence, clinical presentation, sepsis workup, and management in Alexandria University Children's Hospital.

Materials and Methods

Thirty pediatric patients who had undergone surgery for HSD in the Pediatric Surgery Department, at Alexandria University Children's Hospital were reviewed from January 2021 to 2022. History was taken from parents, and patient medical records were reviewed to identify: the type of surgery, the approach used, whether frozen biopsies were taken, if a rectal tube was used after the pull-through, and management of HAEC.

All information was obtained from parents, discharge charts, and investigation results. The information obtained included: demographic data of the patients including age at first surgery, age at presentation and gender, surgical history including the type and approach used, signs and symptoms including abdominal distension, pain, explosive watery diarrhea, fever, vomiting, lethargy, rectal bleeding, pneumoperitoneum, hypotension and shock, clinical grade classification by signs and symptoms, time lapse between pull-through and first attack of HAEC, frequency of attacks, contrast enema, preoperative transanal rectal biopsies, fresh frozen-section biopsies during surgical procedures.

During the period of postoperative Hirschsprung-associated enterocolitis, the following information was obtained: how it was managed, the use of rectal tubes after surgery and duration of use, post-operative dilatation and its schedule.

Result

In the studied group, there were 24 male patients (80%), and 6 female patients (20%). They were presented at 1 – 52 weeks old with a mean age of 18.8 ± 14.74 weeks.

Those patients underwent surgery at 1 – 14 months old with a mean of 7.10 ± 3.45 months.

In the studied group, the most commonly used surgical pull-through was the Soave (53.33%), followed by the Duhamel procedure (20%). Fewer subjects were operated using the Swenson and Rehbein methods, at 16.67% and 10% respectively as shown in **Figure 1.** Similarly, the main surgical approach used was the laparoscopic-assisted approach in 15 patients (50%), followed by the abdominal approach in 10 patients (33.3%) and 5 patients (16.7%) were operated transanally as shown in **Figure 2.**

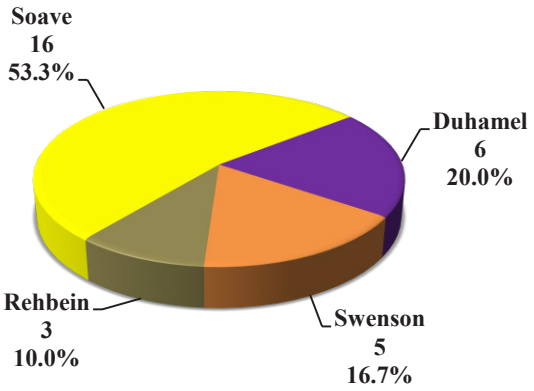

Figure 1: Distribution of the studied cases according to Surgical procedure (n=30)

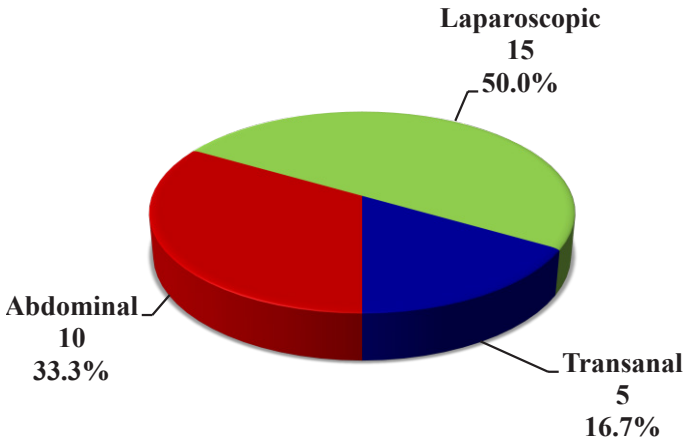

Figure 2: Distribution of the studied cases according to surgical approach (n=30)

Only 3 patients had post-pull-through contrast enema of whom 2 revealed a missed segment as in **Figure 4** and **Figure 1** patient had a stricture as shown in **Figure 5.** A total of 27 patients underwent a post-pull through rectal biopsy after recurrent HAEC, out of whom 25 yielded sufficient ganglia, while 2 revealed aganglionosis, and the last 3 patients didn't do the biopsy as shown in **Figure 3.**

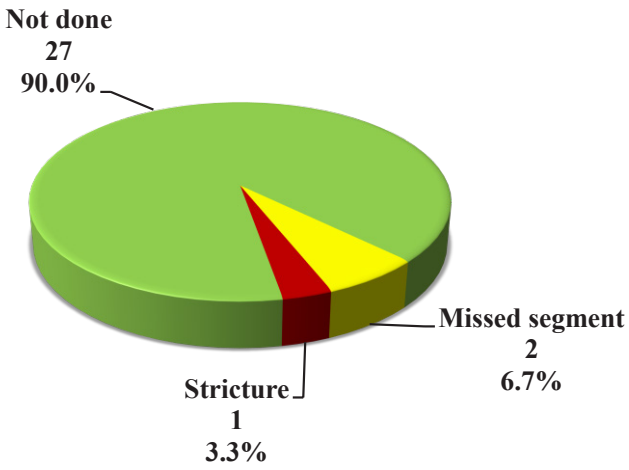

Figure 3: Distribution of the studied cases according to contrast enema findings

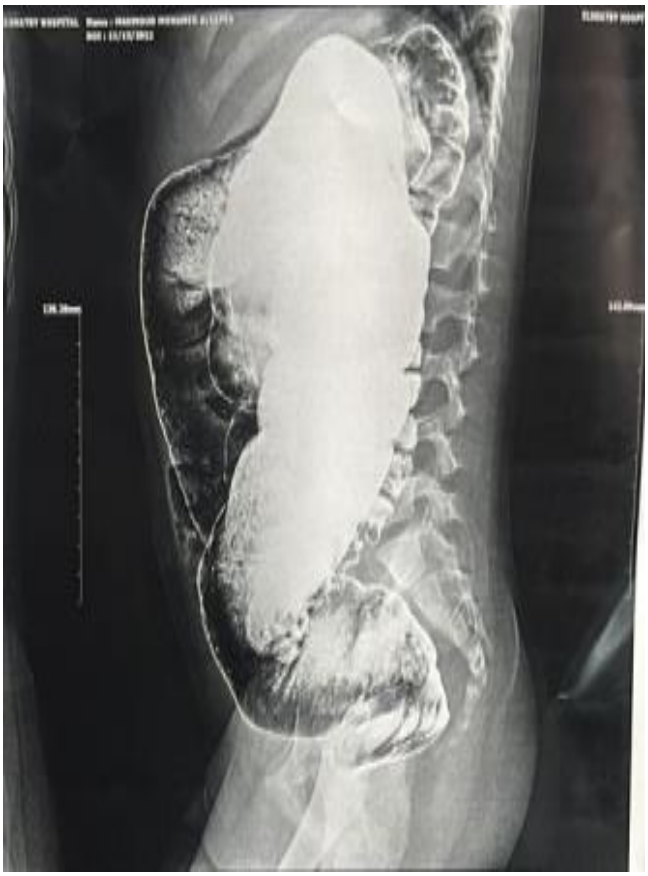

Figure 4: Lateral view of barium enema with diffuse dilation of the colon with no definite transition zone, flow of barium all over the colon and rectum. Rectal biopsy showed aganglionic rectal fragment.

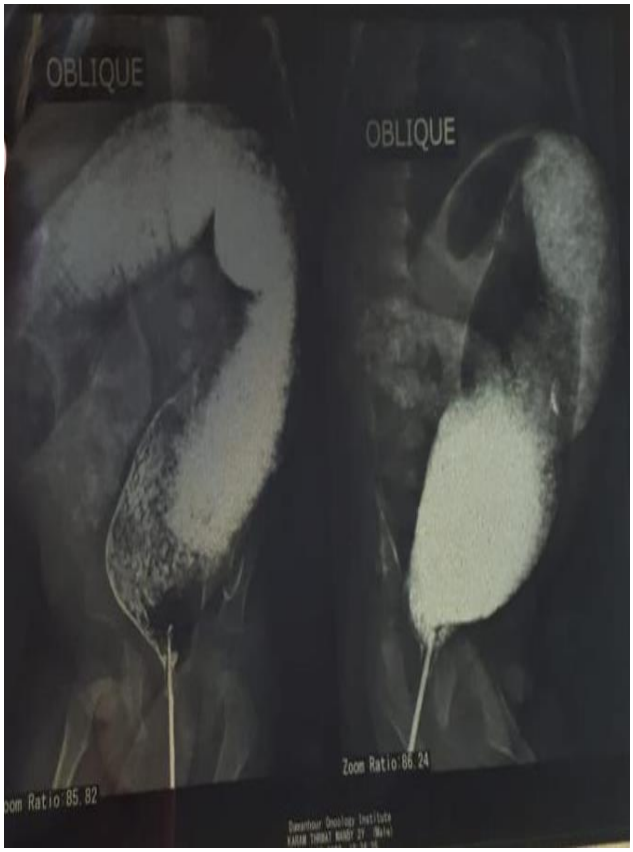

Figure 5: Post-operative barium enema: Markedly dilated rectum with abrupt narrowing at the level of anorectal junction. Significant residual barium in post evacuation and post 24 hours

Most of our studied patients (26 patients) presented with signs and symptoms of Grade I HAEC as shown in **Table 1**, 3 patients with Grade II and 1 patient with Grade III.

Table 1: Distribution of the studied cases according to HAEC grade (N=30)

Grade	No.	%
I	26	86.7
II	3	10.0
III	1	3.3

Grade I: Diarrhea, abdominal distension, and anorexia.

Grade II: Fever, explosive diarrhea, abdominal distension and lethargy.

Grade III: Hypotension, pneumoperitoneum, altered mentation, peritonitis and abdominal distension.

A rectal tube was inserted from 4 to 7 days post-operative to decrease the incidence of enterocolitis.

In our study, the majority of patients (26) were treated conservatively with repeated anal dilatation for 8 weeks which proved helpful, while 2 patients with missed a ganglionic segment (post laparoscopic Soave pull-through), so a redo laparoscopy was done, 1 patient with anal stricture underwent a temporary stoma diversion followed by a later pull through surgery, and only 1 patient needed laparotomy due to peritonitis and severe sepsis**.**

Discussion

Hirschsprung-associated enterocolitis has been a challenge to both pediatricians and pediatric surgeons. Since the Danish pediatrician Harald Hirschsprung presented 2 cases of constipation in newborns caused by colon dilatation and hypertrophy to the International Congress for Children Disease in Berlin in 1886,[12] a great deal has been learned about the pathophysiology, etiology, and best practices for medical and surgical management, and follow-up care of children with the disease that bears his name.

This current study aimed to evaluate HAEC as a postoperative complication in children who underwent pull-through surgeries, regarding the type of surgery, incidence, clinical presentation, sepsis workup, and management.

It has been recognized that males are more commonly affected than females with a male-to-female ratio of 4:1. Badner et al demonstrated that recurrence risk to siblings is dependent upon the sex of the person affected, that if the index patient is female, the proportion of affected siblings is higher.[13] In our study, there was no statistical significance between sex and post-operative HAEC but there was male preponderance in terms of absolute numbers.

In our study, it was discovered that HAEC can be present neonatally or during infancy. The minimum age at presentation was 0 weeks and the maximum at 52 weeks. The previous literature showed that the incidence of HAEC was 24% for infants diagnosed with HSD after the first week of life compared with 11% diagnosed within the first week.[14] Paradoxically children who are diagnosed with HSD after the neonatal period may be resistant to the development of HAEC.[15] In our study there was no correlation between the age of presentation with the postoperative HAEC.

The majority (86.7%) of the studied patients presented with clinical grade 1 HAEC with the following signs and symptoms, anorexia, diarrhea, and abdominal distension. Three patients (10%) presented with grade II HAEC signs and symptoms which included, fever, explosive diarrhea, abdominal distension and tenderness, and lethargy. Only one patient (3.3%) presented with grade III HAEC signs and symptoms including pneumoperitoneum, hypotension, altered mentation, abdominal distension, and peritonitis.

Frozen section biopsy is one of the investigation criteria used to identify aganglionic segments during the intra-operative procedure. In our study, it was done in only 7 patients who had an abdominal surgical approach out of the 30 patients**.** Whereas during postoperative HAEC, contrast enema was used in 3 patients of whom 2 showed a missed segment and 1 showed stricture.

The correlation between post-operative HAEC and contrast enema was found to be statistically significant, with the enema findings of missed segment and stricture. In the literature, it's reported that

histopathological confirmation is the gold standard for the diagnosis of HAEC.[16] This agrees with our findings that rectal biopsy was able to diagnose (missed segment), one of the causes of post-operative HAEC in the patients who had conservative treatment failure.

In the current study, 3 of the 30 patients (10%) had a Rehbein procedure, 16 (53.33%) had a Soave procedure, 5 (16.67%) had Swenson and 6 (20%) had Duhamel. In the literature, it was found that HAEC frequency was significantly higher in the Duhamel than in Soave[17] when a comparison was done between the two procedures, in contrast to the literature, there was no statistical significance between the surgical procedure and the post-operative HAEC in our study.

In our study, 10 of the 30 study patients (33.3%) underwent an abdominal approach, 5 patients (16.7%) had a trans-anal approach, and 15 patients (50%) had a laparoscopic-assisted approach. With the advent of less invasive surgery, classic pull-through procedures have been modified and refined via laparoscopy with consequent less pain and better cosmesis.

Such benefits led to the introduction of the single-stage trans anal endorectal pull-through by de la Torre-Mondragon and Ortega-Salgado.[18] In our study, the transanal, abdominal, and laparoscopic-assisted approaches were reported. There were more laparoscopic approaches than any other approach used. This could be explained by the increasingly available laparoscopically skilled surgeons.

In the literature, it was reported that approximately one-third of patients had long-term gastrointestinal issues, such as constipation, soiling, and recurring enterocolitis following the laparoscopic-assisted approach.[18] In correlation to post-operative HAEC, there was no statistical significance between the surgical approach used with post-operative HAEC. Large, randomized studies with longer follow-ups are needed to compare the outcomes of laparoscopic-assisted to completely trans-anal pull-through operations.[19]

In our study, twenty-three patients had a rectal tube inserted for 5 days, four patients for 7 days, and three patients for 4 days**.** Our data suggests that the minimum duration of rectal tube insertion after pull-through was 4 days and the maximum duration was 7 days. Similarly, post-operative rectal dilatation was 1 day and a maximum of 90 days.

In our study, the majority of patients (26) were treated conservatively with repeated

anal dilatation for 8 weeks which proved helpful, while 2 patients with missed aganglionic segment post laparoscopic Soave pull-through had a redo laparoscopy done, 1 patient with anal stricture needed a temporary stoma diversion before a definitive pull-through surgery, and only 1 patient needed laparotomy due to peritonitis and severe sepsis.

According to earlier research, patients' clinical outcomes are significantly affected by moderate rectal washouts using 30–50 ml of normal saline and recurrent tube decompression.[13] The few cases of redo surgeries in the current study could be attributed to the success of this conservative approach. A small number of patients who developed recurrent episodes of attacks were due to anatomical or pathological causes. Redo pull-through procedures, when necessary, seem to be as successful as primary procedures in terms of continence and frequency of stools, and they can reduce HAEC occurrences.[13] Finally, in most cases, patients who undergo pull-through procedures do well, but a small number develop persistent symptoms. In our study, redo surgeries were very few as many patients were treated conservatively. This is consistent with literature that claims that after a standard pull-through procedure, retained or acquired aganglionosis, intractable constipation and distension with or without enterocolitis, and anastomotic strictures that don't respond to dilatation call for repeat surgery.[20]

Conclusion

Postoperative HAEC is a serious complication and needs proper diagnosis and treatment. Contrast enema plays a significant role in the diagnosis of the stricture and missed segment which are some of the causes of recurrent post-operative HAEC.

Ethical Consideration

Approval was obtained from the Ethics Committee of the Faculty of Medicine – Alexandria, Egypt. (EC serial Protocol Number: 16-0106815- 17/06/2021)

Acknowledgment

Not applicable

Funding/Support

Not applicable

Conflict of interests

There is no conflict of interest

References

1. Scholfield DW, Ram AD: The importance of recording the first passage of meconium in neonates. BMJ. 2017 Mar 2;356.
2. Moore SW, Zaahl MG: A review of genetic mutation in familial Hirschsprung's disease in South Africa: towards genetic counseling. Journal of pediatric surgery. 2008 Feb 1;43(2):325-9.
3. Levin DN, Marcon MA, Rintala RJ, et al: Inflammatory bowel disease manifesting after surgical treatment for Hirschsprung disease. Journal of pediatric gastroenterology and nutrition. 2012 Sep 1;55(3):272-7
4. Suita S, Taguchi T, Ieiri S, et al: Hirschsprung's disease in Japan: analysis of 3852 patients based on a nationwide survey in 30 years. Journal of pediatric surgery. 2005 Jan 1;40(1):197-202.
5. Engum SA, Petrites M, Rescorla FJ, et al: Familial Hirschsprung's disease: 20 cases in 12 kindreds. Journal of pediatric surgery. 1993 Oct 1;28(10):1286-90.
6. Morabito A, Lall A, Gull S, et al: The impact of Down's syndrome on the immediate and long-term outcomes of children with Hirschsprung's disease. Pediatric surgery international. 2006 Feb; 22:179-81.
7. Heuckeroth RO: Hirschsprung disease—integrating basic science and clinical medicine to improve outcomes. Nature reviews Gastroenterology & hepatology. 2018 Mar;15(3):152-67.
8. Almetaher HA, Hassan HS, Elhalaby EA: Current management of Hirschsprung's disease in Egypt: a survey of members of the Egyptian Pediatric Surgical Association. Annals of Pediatric Surgery. 2015;11(4):207-11.
9. Stockmann PT, Philippart AI: The Duhamel procedure for Hirschsprung's disease. In Seminars in Pediatric Surgery 1998 May 1 (Vol. 7, No. 2, pp. 89-95). WB Saunders.
10. Gao Y, Li G, Zhang X, et al: Primary transanal rectosigmoidectomy for Hirschsprung's disease: preliminary results in the initial 33 cases. Journal of pediatric surgery. 2001 Dec 1;36(12):1816-9.
11. Tiryaki T, Demirbag S, Atayurt H, et al: Topical nitric oxide treatment after pull through operations for Hirschsprung disease. Journal of pediatric gastroenterology and nutrition. 2005 Mar 1;40(3):390-2.

12. Gosain A, Brinkman AS: Hirschsprung's associated enterocolitis. Current opinion in pediatrics. 2015 Jun;27(3):364.
13. Holschneider AM, Puri P, editors: Hirschsprung's disease and allied disorders. Berlin: Springer; 2008.;115-123
14. Surana R, Quinn FM, Puri P: Evaluation of risk factors in the development of enterocolitis complicating Hirschsprung's disease. Pediatric surgery international. 1994 Apr; 9:234-6.
15. Haricharan RN, Seo JM, Kelly DR, et al: Older age at diagnosis of Hirschsprung disease decreases risk of postoperative enterocolitis, but resection of additional ganglionated bowel does not. Journal of pediatric surgery. 2008 Jun 1;43(6):1115-23.
16. De Lorijn F, Kremer LC, Reitsma JB, et al: Diagnostic tests in Hirschsprung disease: a systematic review. Journal of pediatric gastroenterology and nutrition. 2006 May 1;42(5):496-505.
17. Parahita IG, Makhmudi A: Comparison of Hirschsprung-associated enterocolitis following Soave and Duhamel procedures. Journal of pediatric surgery. 2018 Jul 1;53(7):1351-4.
18. Tomuschat C, Zimmer J, Puri P: Laparoscopic-assisted pull-through operation for Hirschsprung's disease: a systematic review and meta-analysis. Pediatric surgery international. 2016 Aug; 32:751-7.
19. Ambartsumyan L, Smith C, Kapur RP: Diagnosis of Hirschsprung disease. Pediatric and Developmental Pathology. 2020 Jan;23(1):8-22.
20. Green HL, Rizzolo D, Austin M: Surgical management for Hirschsprung disease: a review for primary care providers. JAAPA. 2016 Apr 1;29(4):24-9.