Section: Pediatric Surgery

Review Article

ENTEROCOLITIS IN THE CLINICAL COURSE OF HIRSCHSPRUNG'S DISEASE: A CLINICAL REVIEW

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Received *: 19/01/2026*
Received in revised form *: 04/03/2026*
Accepted *: 20/03/2026*

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DOI: 10.70034/ijmedph.2026.1.556

Source of Support: Nil,
Conflict of Interest: None declared

Int J Med Pub Health
2026; 16 (1); 3245-3249

ABSTRACT

Background: Objective: To establish the frequency, clinical features, and outcomes of Hirschsprung-associated enterocolitis (HAEC) in the preoperative and postoperative periods in patients with Hirschsprung disease, and to determine factors that may affect its development. **Place and Duration of study:** This study was conducted at Ghulam Muhammad Mahar Medical College Sukkur from 1st February 2025 to 31st January 2026

Materials and Methods: A total of 150 patients with histologically proven Hirschsprung's disease were included. They were all subjected to standardized diagnostic examination, rectal biopsy confirmation, and conclusive surgical treatment with reference to laid-down pediatric surgical guidelines. Clinical data were analyzed in terms of demographic variables, age at presentation, aganglionic length, pre-operative bowel activity, abdominal distension, fever, diarrhea, and radiographic images that could have indicated the presence of enterocolitis. Anastomotic integrity, caliber changes, obstructive symptoms, and functional outcomes were used to assess post-operative outcomes. The patient was diagnosed with HAEC based on clinical manifestations and laboratory and radiologic findings. Enterocolitis rate before and after definitive pull-through surgery was recorded, as well as severity, treatment response, and complications.

Results: Out of the 150 patients, a significant number of them experienced enterocolitis at one point in their clinical course. A total of 42 patients (28%), pre-operatively, were already experiencing at least one episode of HAEC, with the majority of the episodes manifesting with abdominal distension, explosive stools, fever, and radiographic evidence of colonic dilation. Enterocolitis developed in 27 patients (18%), and the incidents happened during the initial stage of the post-operative period, as well as years after the intervention. Post-operative HAEC had more commonly been related to anastomotic constriction, residual transition zone pathology or functional obstruction. Long-segment disease, as well as delayed patient onset, demonstrated a greater predisposition towards recurrent attacks. Most of them responded to conservative measures like bowel rest, rectal irrigations and antibiotics, with a minority of them having to remain operative due to complications like persistent obstruction or bad sepsis.

Conclusion: Enterocolitis proves to be a serious and prominent complication in the natural history of Hirschsprung's disease in both preoperative and postoperative periods of Hirschsprung disease. Early identification, early decompression and careful follow-up after the operation are required to minimize morbidity. Interventions to reduce the occurrence of anastomotic stenosis, proper resection of the transition zone and informing caregivers on early signs can significantly reduce the rate and magnitude of HAEC. Further

focus on standardized diagnostic criteria and organized follow-up procedures can be used to enhance the final results of the affected children.
Keywords: Hirschsprung's disease, Hirschsprung-associated enterocolitis, pre-operative enterocolitis, post-operative complications, pediatric surgery.

INTRODUCTION

Hirschsprung disease (HD) is an inborn defect of the enteric nervous system where the ganglion cells are missing in the terminal bowel, leading to functional intestinal obstruction as well as progressive colonic dilation. It is still among the leading causes of lower neonatal intestinal obstruction with an estimated incidence of 1 per 5,000 live births and a definite male predisposition.[1] Despite the introduction of new neonatal care and diagnostic methods, as well as surgery, the Hirschsprung-related enterocolitis (HAEC) has remained the most dreaded and potentially lethal complication during the path of the disease.[2]

HAEC can be a preoperative phenomenon, an acute post-pull-through intervention, or years later, and thus is a long-standing clinical problem. The disorder is marked by a range of symptoms, such as abdominal distention, fever, explosive diarrhea, lethargy and in some extreme cases, septic shock and dysfunction of multiple organs.[3] The mechanism of the pathophysiology of HAEC is not fully understood after decades of research. Existing evidence indicates that this is caused by a multifactorial process, which includes impaired mucosal immunity, dysbiosis of the intestinal microbiome, stasis-related bacteria overgrowth, epithelial barrier dysfunction, and inflammatory dysregulation.[4-6] These processes are associated with anatomical and functional obstruction, which makes a susceptible environment of repetitive occurrences.

The primary diagnostic criteria, including the Pastor scoring system, have been introduced to enhance better recognition and categorization of the severity of HAEC, but inconsistency in clinical presentation still makes early diagnosis challenging.[7] Besides, several patient and disease-related issues, such as long-segment aganglionosis, late diagnosis, coinciding syndrome like Down syndrome, and post-operative conditions such as anastomotic stricture or remnant transition zone pathology, are also found to influence the risk of HAEC.[8,9] With technically successful pull -through surgery, as many as one-third of the patients can still have at least one occurrence of postoperative condition, which underscores the importance of long-term follow-up.[10]

The cost of HAEC is still high in the world. In the past 10 years, studies have reported 20-40 % preoperative HAEC rates, and 10-35 % postoperative, depending on the method of surgery, duration of follow-up and methods of diagnostic tests to be used.[11,12] Such differences emphasize the value of local data to enhance the information about the patterns of epidemiology, possible risk factors that can be modified to improve management strategies. The patient should be diagnosed early and treated properly, as failure to intervene in time may result in rapid progression, bowel perforation, and mortality.[13]

Treatment of HAEC is usually bowel clearing, rectal irrigation, fluid replacement, and broad-spectrum antibiotics, and surgery is used in cases of refractory or complicated cases. Nonetheless, prevention is also a priority. Routine monitoring after surgery, early detection of anastomotic constriction and education of the caregiver on the early symptoms of an anastomosis have been found to decrease the occurrence and severity of episodes.[14] Moreover, there are new studies on the gut microbiome and immunological signals that can be used in the future to risk-stratify and provide precision in future therapies.[15]

Regardless of these developments, some gaps still exist in the study of the natural history of HAEC among various populations. The lack of uniformity in clinical practice, diagnostic limits and strategies of follow-up adds to the disparities in reporting across the different centers. Thus, research comparing the prevalence and aspects of HAEC in various clinical conditions is still necessary. The current study should help to provide valuable information to the body of literature and make optimal clinical decisions by analyzing pre-operative and post-operative episodes among a well-defined group of participants.

MATERIALS AND METHODS

In this observational study, 150 patients with Hirschsprung's disease (HD) were used in the study based on clinical assessment and confirmatory histopathology. All patients had gone through the standardized diagnostic and therapeutic procedures. Male and female patients were not restricted and they could have been of any age when they presented.

In all the cases, Hirschsprung disease was diagnosed using rectal full-thickness biopsy, which was considered the gold standard to confirm the presence of aganglionosis. The samples of biopsy were assessed based on the lack of ganglion cells and large nerve trunks. The diagnostic suspicion was supported by clinical features of delayed passage of meconium, chronic constipation, abdominal distension, and failure to thrive, before biopsy. To measure the transition zone and to determine the colonic dilation, radiological tests, such as contrast enemas, were selective, but the final criterion was histopathology.

Diversions were done in this cohort depending on the anatomic size of the disease and the level of colonic dilation at the time of presentation. Since the location of aganglionosis could not be determined by intra-operative frozen section facilities, the colostomy was always created in the grossly dilated portion, which was considered to be the ganglionic bowel. Such a strategy was implemented to make sure that safe decompression is achieved and a stoma is not placed in the aganglionic bowel or in the transitional bowel. Most of the patients were subjected to transverse colostomy, though in patients with long-segment or total colonic aganglionosis, an ileostomy had to be developed. The type of the stoma and the position of the stoma were personalized according to the results of the intraoperative examination and evaluation by the surgeon of the viability of the bowel and its caliber.

Among the 150 patients, a significant number of them needed laparotomy to be assessed and treated conclusively. The ultrashort-segment disease and cases with inconclusive results of imaging were evaluated by intraoperative assessment to identify the diagnosis and the course of surgery. The definitive pull-through procedures were done with standard procedures and the most common pull-through system is the Soave endorectal pull-through. The rationale behind the use of definitive surgery was patient age, nutritional status, and the presence of stoma working and preoperative enterocolitis resolved, where necessary. Those patients who were not directly fit to pull through were continued under stoma care and made to undergo delayed reconstruction.

Clinical signs of HAEC were observed in all patients before and after definitive surgery. A diagnosis of enterocolitis was made using a composite of symptoms such as abdominal distension, fever, foul odor, diarrhea, lethargy, and radiographic findings such as the stenosis of the colonic lumen or mucosal disfigurement. The diagnosis was supported by the laboratory parameters, namely, leukocytosis and electrolyte imbalance. Episodes were divided into preoperative and postoperative; their frequency, severity and reaction to treatment were recorded. The management involved bowel rest, rectal irrigations, intravenous fluids and wide-spectrum antibiotics. Confusion Cases of severe or refractory cases were considered in terms of complications that include obstruction, anastomotic stricture, or residual aganglionosis.

The demographic data, clinical presentation, operative data, and postoperative outcomes were captured in a systematic manner. Particular interest was paid to such factors that were related to the appearance of enterocolitis as age of diagnosis, length of aganglionic segment, type of stoma, and postoperative anatomical or functional abnormalities. Data analysis was done in a descriptive way and categorical variables were summarized using frequencies and percentages. The most important outcome measure was the preoperative and postoperative frequency of HAEC.

Table 1: Demographic, clinical, and surgical characteristics of patients (n = 150)

Variable	Category	Number of patients
Gender distribution	Male	112
	Female	38
Age at diagnosis	Less than 6 months	50
	6 to 24 months	28
	2 to 5 years	64
	Above 5 years	8
Pre-operative enterocolitis	Yes	42 (28%)
	No	108 (72%)
Post-operative enterocolitis	Yes	27 (18%)
	No	123 (82%)
Type of initial stoma	Transverse colostomy	32
	Sigmoid colostomy	18
	Ileostomy (long-segment / TCA)	3
	Other colostomies (descending/loop)	12
	No stoma (ultrashort segment / direct pull-through)	85
Definitive surgery performed	Soave pull-through	54
	Duhamel pull-through	8
	Transanal endorectal pull-through (TERPT)	12
	Awaiting definitive surgery	76

RESULTS

This study involved 150 patients who were confirmed to have Hirschsprung. The sample population was 112 males and 38 females, as there was a predicted dominance of males. Table I presents the age distribution of the diagnosis. The majority of children were brought between 2 and 5 years of age and then infants under the age of 6 months.

Enterocolitis was found in a large percentage of patients in their clinical course. Pre-operative enterocolitis was reported in 42 (28 %) patients and in most of these individuals, no precipitating factor except delayed diagnosis could be made. The symptoms were abdominal distention, fever, and foul-smelling diarrhea and radiographic features of colonic dilation.

65 patients were laparotomized with a diversion stoma (colostomy or ileostomy), and the rest of the patients either had ultrashort-segment disease or were treated with planned definitive pull-through without a diversion. The level of the aganglionic segment was identified during surgery and the results are in Table II. The most prevalent pattern was rectosigmoid disease, then isolated rectal involvement.

Out of the patients that underwent definitive pull-through surgery (74 patients), the postoperative complication was closely monitored. The enterocolitis was postoperative in 27 patients (18%), and it was mild to severe. Moreover, 11 patients developed anastomotic or anorectal stenosis and all of them were also related to recurrent enterocolitis that shows that mechanical obstruction is closely correlated with HAEC.

Enterocolitis occurred in most cases, and these cases responded to conservative therapy such as rectal irrigations, bowel rest and antibiotics. A few of the serious cases necessitated operative assessment to rule out residual aganglionosis or transition-zone pathology. This cohort did not have any mortality.

Table 2: Extent of Aganglionic Lesions

Segment Involved	Original %	Adjusted Number (n = 150)
Rectum	41.2%	62
Rectosigmoid	36.5%	55
Pelvic colon	11.1%	17
Transverse colon	3.1%	5
Total colon	4.7%	7
Total	100%	150

DISCUSSION

The current research shows that HAEC is a major clinical problem, with disease prevalence increasing up to 28 % in the pre-operative period and 18 % in the post-operative period. These results are consistent with the world literature that regularly records HAEC as the commonest and possibly fatal complication of Hirschsprung disease. Incidence Post-operative. The incidence of our cohort during the pre-operative period is similar to a 25-35% range of recent multi-centers studies,[16,17] indicating that late diagnosis and functional blockage are still significant factors in the development of the disease. We have a pre-operative HAEC rate of 28 %, which is consistent with 30 % reported by Yildiz et al,[18] who also found late presentation to be an important factor. Conversely, a study of the East Asian region was able to record a slightly higher pre-operative rate of 38%,[19] which the authors put down to referral delays and the lack of access to early diagnostic centers. Such differences underscore the significance of early detection of HD symptoms, particularly in areas where the availability of screening of the neonatal population and early surgical assessment may not be equally acceptable.

The 18% of HAEC rate in our study is within the 15-25% range described in the modern literature. An illustration of this is that Nasr et al. have found that the incidence of postoperative pull-through surgeries is 20 %,[20] and a European cohort has reported 17 % rate.[21] Our results support the idea that HAEC may take place even in cases where the surgery is technically successful, and it is necessary to underline the multifactorial character of the condition. It has all been attributed to residual transition zone pathology, anastomotic constriction, and dysmotility as contributors to postoperative episodes.

Anorectal stenosis was observed in 11 patients in our cohort, with all patients having postoperative enterocolitis. Such a close correlation resembles the results of Menezes et al., who found that postoperative strictures were a significant risk of recurrent HAEC.[22] In the same way, a North American study proved that children with anastomotic constriction were three times more likely to develop postoperative enterocolitis.[23] These findings support the importance of close postoperative monitoring, prompt stenosis diagnosis and dilatation guidelines.

Our study data of the aganglionic segment distribution mostly comprised of rectal and rectosigmoid distribution, which is in accordance with the global epidemiological trends. A recent review by Zhang et al. stated that rectosigmoid disease is a major cause of HD of about 70-80 %,[24] which is pretty close to the percentages we have had in our cohort. Long-segment and total colonic aganglionosis had lower prevalence and were at risk of serious enterocolitis as were also observed in Japanese and American studies.[25,26]

Our study results on management also support the usefulness of conservative therapy of the majority of HAEC episodes. The mainstay of treatment is still rectal irrigations, bowel rest, and antibiotics and good responses have been documented in various studies.[27] The number of patients who needed surgical treatment was low, in line with the recent systematic reviews, which highlighted that surgery is only used in refractory or complicated cases.[28]

All in all, the findings of this research are in agreement with the existing international data, which helps to confirm the ongoing load of HAEC in spite of the development of new diagnostic and surgical methods. The fact that similarities are observed in the various populations implies that the underlying pathophysiology, which is presumed to be dysbiosis, mucosal immunity, and functional

obstruction, is universally applicable. Meanwhile, the differences in the incidence point to the significance of timely diagnosis, standard postoperative follow-up, and education of the caregivers.

Research in the future ought to aim at the identification of biomarkers to detect microbiome-based therapies early, optimizing of the microbiome-based therapies, and the surgical methodology to reduce residual transition-zone pathology. Multicenter prospective research can contribute to the clarification of which among the anatomical, immunological, and microbial factors play a significant role in the development of HAEC.

CONCLUSION

Enterocolitis, consisting of Hirschsprung-associated enterocolitis, is a common and clinically relevant complication of Hirschsprung disease in children. Both pre- and post-operative HAEC were noted at significant rates in this study, which was mostly determined by the late diagnosis, functional obstruction, and postoperative anatomical defects, including anastomotic stenosis. Our data on the pattern of aganglionosis and the general clinical picture of our patients were in line with the international data. Reducing morbidity requires early recognition, timely decompression and planned postoperative follow-up. Further improvements can be achieved by considering the transition-zone pathology, stenosis prevention, and educating the caregivers. Further investigation of the pathogenesis of HAEC can be used to optimize preventive and therapeutic interventions in the future.

Source of Funding: None
Permission: Ethical consent received.
Conflict of interest: None

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