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Clinical Profile of Hirschsprung Disease, Postoperative Complications of Modified Duhamel Procedure and their Management

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Abstract: ***Hirschsprung disease (HD) is a congenital condition marked by the absence of ganglion cells in the distal bowel, causing chronic intestinal obstruction. The modified Duhamel procedure is a widely used surgical approach to treat HD, aiming to restore bowel function by creating a neorectum. This study aimed to evaluate the clinical profile of patients with HD and analyze the postoperative complications associated with the modified Duhamel procedure, as well as their management. A Prospective retrospective review was conducted on pediatric patients diagnosed with HD who underwent the modified Duhamel procedure. Data such as age, gender, and presenting symptoms were collected, along with details of postoperative complications including anastomotic stricture, enterocolitis, and faecal incontinence. Management strategies like reoperation, dilation, and medical therapy were also reviewed. The study included 8 patients with an average age of neonate to 13 years. Out of 8 patients, 5 (62.5%) were males and 3 (37.5%) were females. All patients were analysed for early postoperative complications, with decreased appetite observed in 12.5% of the cases, skin irritation and erythema in 37.5%, postoperative fever in 12.5%, stoma prolapse in 12.5%, and wound infection in 12.5%. Late complications included subacute intestinal obstruction (12.5%), deranged electrolytes (12.5%), and postoperative septicaemia leading to death in 12.5% of the patients. Early intervention with balloon dilatation effectively treated anastomotic strictures, while antibiotic therapy was successfully used for enterocolitis management. A few cases required reoperation due to severe complications. The results highlight that while the modified Duhamel procedure is effective in managing HD, postoperative complications are not uncommon and can significantly impact recovery. Early identification and appropriate management of these complications, particularly through non-surgical interventions, are key to achieving favourable outcomes and improving the quality of life for pediatric patients with HD.***

Keywords: Hirschsprung disease, modified Duhamel procedure, postoperative complications, management

1. Introduction

Hirschsprung disease (HD) is a congenital disorder in which the distal parts of the digestive system lack intrinsic nerves (ganglion cells). Because these aberrant segments do not relax during peristalsis, they induce mechanical blockage. Hirschsprung disease is characterised by a lack of propagation of propulsive waves and aberrant or absent relaxation of the internal anal sphincter as a result of bowel aganglionosis, hypoganglionosis, or dysganglionosis. Hirschsprung disease always begins at the anal verge, but the length of the ganglion-free segment (aganglionic) varies in 75% of cases, it is limited to the rectum and sigmoid; in 8% of cases, it involves the entire colon; and in a tiny percentage of cases, it affects the small bowel. The average age at the time of diagnosis has been decreasing over the years. The diagnosis is established in 15% within the first month of life, 40-50% in the first 3 months, 60% at the end of the first year of age, and 85% by 4 years. Hirschsprung disease is sometimes not diagnosed until a person reaches adulthood. Although most epidemiologic research has been confined to the Caucasian Diaspora, there may be as yet undiscovered interracial differences, demographic studies have demonstrated a fairly stable incidence of HSCR of roughly 1 in 5000 in both hemispheres. The authors of a California survey showed significant racial disparities in the incidence of HSCR: 1:10,000 births in Hispanic subjects, 1:6667 in white subjects, 1:4761 in black subjects, and 1:3571 in Asian individuals. Some of the variances could be explained by varied levels of consanguinity in different populations, but the authors of recent genetic studies concerning frequencies of HSCR-associated mutations point to different frequencies in other ethnic populations.[1] The abnormal colonisation of the enteric nervous system (ENS) neuroblasts during development is thought to be the cause of intestinal aganglionosis. Even though HSCR is primarily thought to be a hereditary disease, research has identified at least five levels where aberrant molecular signalling may play a role in the disease's etiology. In syndromic cases (e.g., Down syndrome), these include a smaller pool of available neuroblasts for migration into the enteric nervous system, early gene-related influences on migrating neuroblasts, abnormally developed neuroblasts migrating into the ENS (e.g., Down syndrome), germline and somatic gene mutations, and changes in the local tissue environment. It would appear that all these contribute to the disruption of normal signalling during enteric neuroblast development.[2] Symptoms in older children range from newborn intestinal blockage to chronic progressive constipation.[3-4] Approximately 80% of patients present with difficult bowel motions, poor eating, and growing abdominal distention in the first few months of life. Hirschsprung disease causes up to 90% of newborns to fail to pass meconium during the first 24 hours of life; however, other causes of this delay should be considered as well. Hirschsprung disease is characterized by infrequent, explosive bowel motions produced by functional intestinal blockage. A tight anal sphincter and explosive stool and flatus discharge may be seen on rectal examination.[3] Although the majority of patients present during childhood and adolescence, others may not show symptoms until later in life.[5-6] Chronic progressive constipation repeated faecal impaction, failure to thrive, and malnutrition are all common symptoms in older children.[7]

One-third of patients with Hirschsprung disease present with enterocolitis-related diarrhoea rather than constipation.[3] The classical presentation of Hirschsprung disease, supported by a contrast enema showing a transition zone (TZ) [Figure 1], usually makes the diagnosis.

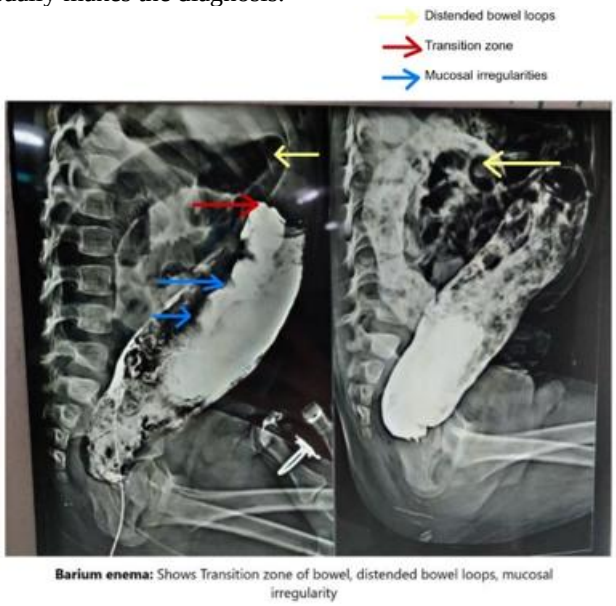

Figure 1: Transition zone of bowel, distended loops, and mucosal irregularity

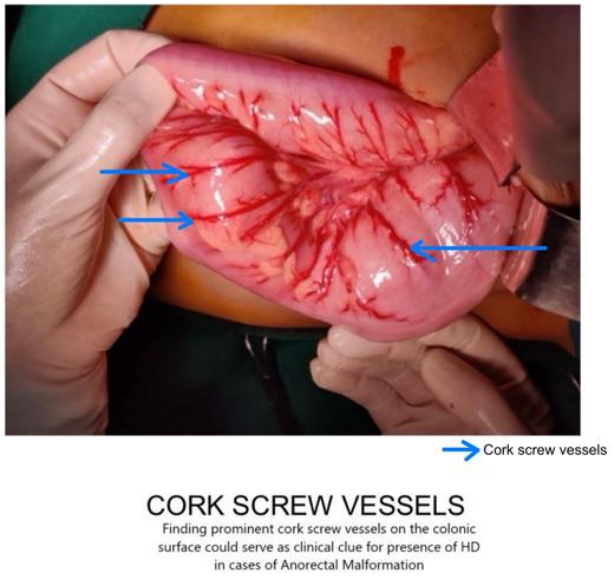

Figure 2: Corkscrew vessels.

A rectal biopsy showing absent ganglionic cells, nerve hypertrophy, and increased acetylcholinesterase activity on an immune histochemical study is confirmatory. Acetylcholinesterase histochemistry is a useful adjunct for the diagnosis of Hirschsprung disease and is only performed with frozen sections. [8] Several surgical procedures have been described to treat the disease[8—14]. The aganglionic part of the colon is surgically resected, with a normal piece pulled through and anastomosis to the anus. Despite the multiple ways for proper aganglionic segment resection, pre- and postoperative problems are common, and total mortality in low- and middle-income nations is currently 18%.[15] Postoperative problems including faecal incontinence, dehiscence, retraction, and constipation have a significant negative impact on these children's quality of life. [16,17] In separate studies, the incidence of one of the most serious consequences, enterocolitis, ranged from 10% to 30%.[18] The standard surgical treatment of Hirschsprung disease was done in three stages: the creation of a proximal diverting stoma, resection of the aganglionic bowel segment and restoration of bowel continuity. At present, surgery is the most common treatment method. The most popular procedures for definitive treatment of Hirschsprung disease include Swenson's procedure, Soave's endorectal pull-through and Modified Duhamel procedure. Nowadays, multistage surgery has progressed to open or laparoscopically assisted one-stage repair. The rationale of single-stage surgery has been that it provides a cure without multiple operations, decreases surgical scars and hospital stays and avoids a stoma. Previous reports have mentioned that multistage and single-stage repair results are comparable. The advantages of the Modified Duhamel procedure include ease of performance, reduction of anastomotic leaks and strictures, retention of anal sensory receptors and preservation of Nervi erigentes. The present study aims to generate scientific data revealing early and late postoperative complications of the Modified Duhamel Procedure for the treatment of Hirschsprung disease and comparing them with other surgical modalities to prevent morbidities and mortalities associated with Hirschsprung disease.

Aims and Objectives

1) To study the pathophysiology and clinical presentation of Hirschsprung disease
2) To study the Modified Duhamel Procedure
3) To study the early postoperative complications of the Modified Duhamel Procedure
4) To evaluate the long-term or late post-operative complication profile of the Modified Duhamel Procedure
5) Management of complications of Modified Duhamel Procedure

2. Methodology

2.1 Study Design, Settings, Area, and Population

This prospective retrospective study aims to generate scientific data on early and late postoperative complications of the Modified Duhamel Procedure in treating Hirschsprung disease and to compare these outcomes with other surgical modalities. All histopathological proved cases of Hirschsprung disease admitted to the Department of Surgery in Paediartic surgical wards in a Tertiary Care Center from July 2019 to December 2021 were included in the study. In our study, we were able to recruit 8 cases in the study. Parental psychological counselling was provided to address the early and late postoperative complications, as well as the long term management aspects, associated with the Modified Duhamel Procedure in patients with Hirschsprung disease.

2.2 Ethical Considerations

The study was ethically approved by the Institutional Ethics Committee (IEC). Written informed consent was obtained from all the patients. Informed consent was provided in different languages understandable by patients.

2.3 Data Collection and Analysis

Data for this study were collected through a thorough clinical assessment of patients, beginning with a detailed history to understand symptoms, previous treatments, and relevant background. Each patient then underwent a series of investigations to confirm the diagnosis of Hirschsprung disease. These investigations included clinical examination, radiological imaging, and histopathological confirmation to ensure diagnostic accuracy. Patients were included in the study if they had a confirmed diagnosis of Hirschsprung disease, established through clinical, radiological, and histopathological evidence, and were within the age range of neonatal to thirteen years. Cases that lacked histopathological confirmation were excluded from the study to maintain diagnostic rigor and ensure the reliability of the collected data. SPSS software version 24 was used for the analysis of data.

3. Results

3.1 Gender Distribution

The present prospective retrospective study was conducted in the Department of Surgery among 8 cases of histopathologically proved Hirschsprung disease admitted in the Department of Surgery in Paediartic surgical wards in a Tertiary Care Center from July 2019- to December 2021. Out of 8 subjects; 5 (62.5%) were males and 3 (37.5%) were females as shown in Table 1, Figure 3.

Table 1: Gender distribution among the study subjects

Gender	N	%
Male	5	62.5
Female	3	37.5
Total	8	100

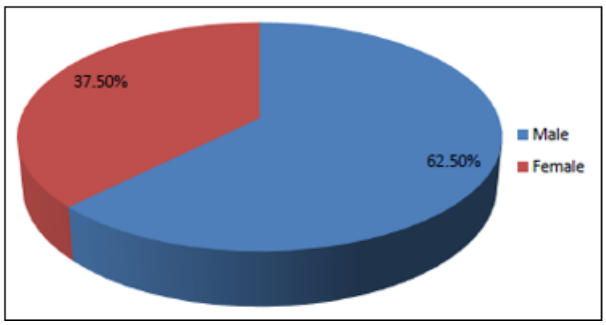

Figure 3: Gender distribution among the study subjects

3.2 Age Distribution

The age distribution among the study subjects ranged from less than 1 to more than 5 years of age (Figure 4). 50%, 37.5% and 12.5% of the subjects were of age ≤1, >1-5 and >5 years respectively.

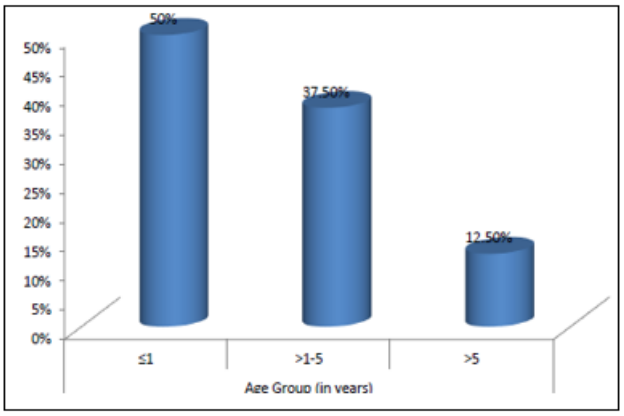

Figure 4: Age Distribution

3.3 Chief Complaint in the Study Subjects

Figure 5 shows the chief complaints among the study subjects. All the subjects complained of constipation as well as abdominal distension. Vomiting and feeding intolerance were revealed in 25% and 12.5% of the subjects respectively.

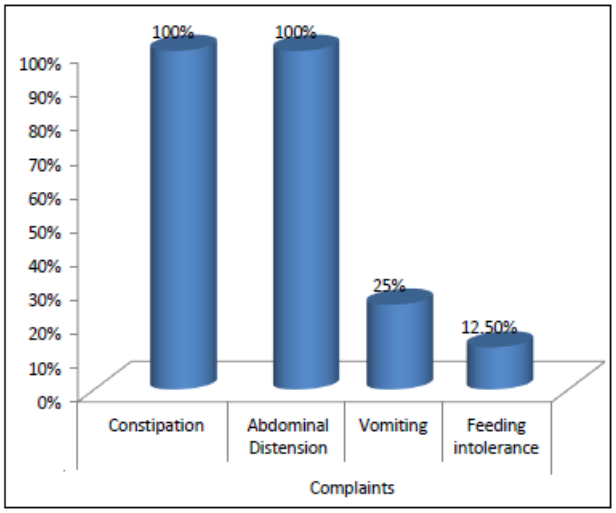

Figure 5: Chief complaints among the study subjects.

3.4 Treatment Given in The Study Subjects

Table 2 shows the various treatments given among the study subjects. Laparotomy with multiple colonic biopsies alongside colostomy was performed in 6 subjects. Colostomy closure and emergency laparotomy alongside colostomy with rectal biopsy were done on 3 and 2 subjects respectively. All the subjects underwent the Modified Duhamel procedure.

Table 2: Treatment given among the study subjects

Treatment	N	%
Laparotomy with multiple colonic biopsies with colostomy	6	75
Modified Duhamel procedure	8	100
Colostomy closure	3	37.5
Emergency Laparotomy with colostomy with rectal biopsy	2	25

3.5 Early and Late Complications Among the Study Subject

Figures 6 and 7 show the early and late complications among the study subjects. The most common complication was skin

irritation and erythema (37.5%). The rest of all the complications viz. decreased appetite, post-op fever, stoma prolapse, wound infection, deranged electrolytes and post-op septicemia & death were revealed in 1 subject each.

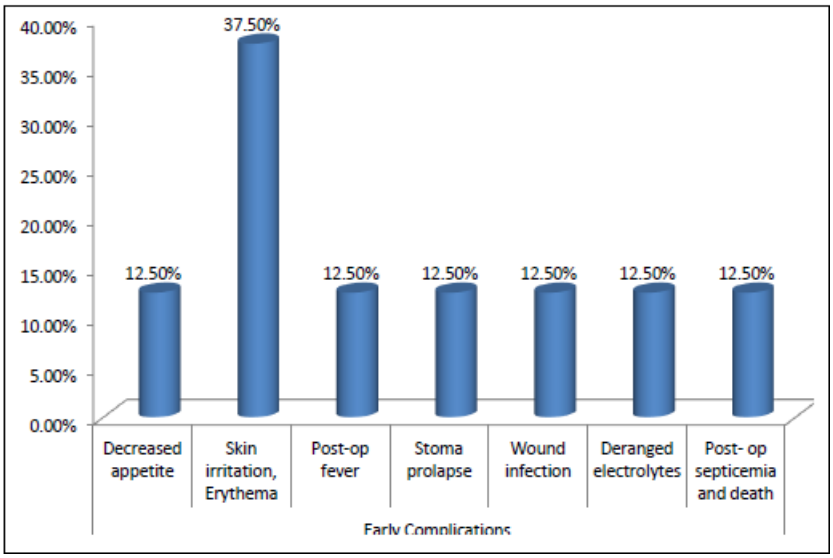

Figure 6: Early complications among the study subjects

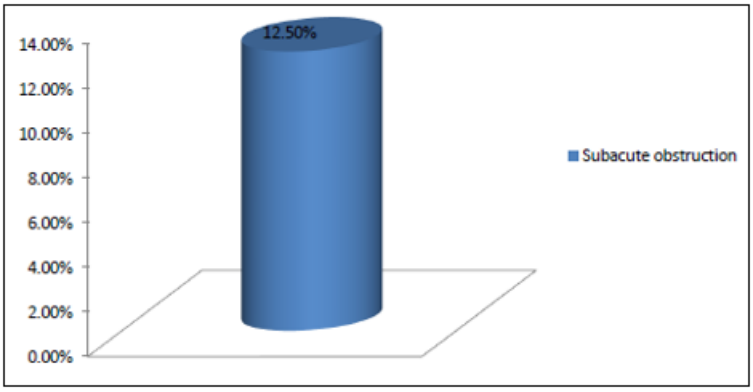

Figure 7: Late Complications of Study Subject

4. Discussion

Hirschsprung disease is one of the "classics" of pediatric surgery and the development of Hirschsprung disease management closely portrays the overall development and quality of pediatric surgery. Regardless of approach, the basics of surgical repair remain the removal of the aganglionic bowel, pushing through of ganglionated bowel, and preservation of the anal canal and sphincter mechanism. [19] Despite tremendous advances in our understanding of the pathologic anatomy and physiology of Hirschsprung disease, surgical treatment results are still far from flawless.[20] The three traditional surgeries (Soave, Swenson, and Duhamel Procedures) and their adaptations to single-staged techniques, total endorectal techniques, and laparoscopy-assisted procedures have advanced surgical care of Hirschsprung disease. A committed histopathology team with expertise in frozen section methods is a prerequisite for all single-staged procedures.[21,22] Staged procedures for Hirschsprung disease can be performed safely with acceptable functional outcomes even in the absence of a frozen section facility. As the Duhamel Procedure require minimal pelvic dissection and can be easily performed with fewer procedure-related complications, it is a reasonably good option for the management of Hirschsprung disease.[23]

The original Duhamel procedure implied complete division of the internal anal sphincter[24], but this caused a high incidence of incontinence. This prompted a series of modifications to partially preserve the internal anal sphincter[25-29]. Martin's modification was introduced to take care of the blind rectal pouch, and the problems associated with it. Martin's modification of the Duhamel procedure (MDP) was used in our patients.[28] Hirschsprung disease can affect neonates most commonly. In a study by K L Aravind *et al.* [23], the most common age at presentation was a neonatal period, and among them, the majority presented within the 1st week, accounting for 11 (64.7%) cases. Four (23.52%) of the patients presented between 1 and 12 months of age. Late presenters accounted for 2 (11.76%) of cases, who presented beyond 1 year of age. The current study revealed that 50%, 37.5% and 12.5% of the subjects were aged ≤1, >1-5 and >5 years respectively in this study. According to a study by Peters et al[37] fifty-seven, (82.6%) of patients had a history of delayed passage of meconium and > 98.6% presented with constipation (98.6%). In the present study, the subjects complained of constipation as well as abdominal distension. Vomiting and feeding intolerance were revealed in 25% and 12.5% of the subjects respectively in our study.

Hirschsprung-associated enterocolitis (HAEC) represents the primary cause of high morbidity and mortality in Hirschsprung disease patients. There are several hypotheses for the cause of Hirschsprung-associated enterocolitis involving dysbiosis of the intestinal microbiome, compromised mucosal barrier function, changed innate immune responses, and translocation of bacteria.[30] In the present study, the most common complication was skin irritation and erythema (37.5%). The rest of all the complications viz. decreased appetite, post-op fever, stoma prolapse, wound infection, deranged electrolytes and post-op septicemia & death were revealed in 1 subject each. In a study by K L Aravind *et al*[23], post-operative complications included wound infection (11.76%), perianal excoriation (17.64%), enterocolitis (11.76%), stricture formation (11.76%), constipation (11.76%), fecal soiling (29.41%), and adhesive bowel obstruction (5.89%). These complications have been variedly reported in several large series (Pontarelli EM *et al.*[31], Seo S. *et al.*[32], Pratoa AP *et al.*[33]). The success of a procedure to correct Hirschsprung disease is judged by the continence achieved by the patient. Continence is the ability to control the passage of faeces; to hold on if necessary, and to pass completely when the individual wants at a socially convenient time. Voluntary bowel movements were established in 7 (87.5%) children. By voluntary bowel movement, we mean the feeling of urge, and capacity to verbalize and hold the bowel movement. In their study, S. Agarwala et al [34] found that the incidence of early obstruction was 11.7% in the Modified Duhamel Procedure. According to Peters et al[35], enterocolitis occurred in 4 (5.8%) after surgery, resolved with medical management. None of them had a history of preoperative enterocolitis. Bourdelat *et al*[36] reviewed 2, 430 patients who underwent a Duhamel operation. The majority of patients had outstanding anal function after 15–30 years of follow-up. Constipation affected only 8.07 percent of patients, whereas soiling affected 5.3 percent. However, lingering postoperative issues such as soiling, constipation, enterocolitis, and anastomotic strictures were reported by Baillie et al[37] in 10%–80% of patients. Other evidence found a 32 to 35 percent incidence of perineal excoriations and soiling[38-41]. Long-term perineal excoriation is not only an indicator of persistent soiling and incontinence but also may be an indicator of anastomotic stenosis[42]. In our study, mortality was observed in 1 subject (12.5%). In contrast, Peters et al.[35] reported no mortality in their study, and K. L. Aravind et al.[23] also found no instances of mortality. Most studies on postoperative- complications in the Modified Duhamel Procedure, such as those by Parahita IG *et al*.[30] and Prato AP et al.[33], have reported low mortality rates. Hirschsprung-associated enterocolitis remains the leading cause of morbidity and mortality in patients with Hirschsprung disease. Before the development of advanced diagnostic and surgical options for Hirschsprung disease, toxic megacolon was a significant cause of death; however, none of the patients in our series experienced this condition. In the present study, parents were educated on the importance of early detection and intervention, including bowel decompression, antibiotics, and fluid resuscitation, to prevent morbidity and mortality in the event of enterocolitis symptoms.

5. Conclusion

The Modified Duhamel Procedure (MDP) is a safe procedure that has a shorter learning curve for new surgeons and can treat a wide range of Hirschsprung diseases. The use of the Modified Duhamel Procedure in conjunction with an initial levelling colostomy helps to compensate for the lack of a frozen section facility. To some extent, stapled anastomosis helps to overcome variability in hand-stitched anastomosis. Other series of the Modified Duhamel Procedure and other procedures have similar complications. Following the Modified Duhamel Procedure, children with Hirschsprung disease can attain an excellent quality of life with proper follow-up.

Supplementary information: Informed consent and ethical committee approval was obtained for the original research paper.

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