JUSINDO, Vol. 8 No. 2, July-December 2026
p-ISSN: 2775-0892, e-ISSN: 2775-3077

Management of Short-Segment Hirschsprung-Associated Enterocolitis in an Infant in East Kalimantan, Indonesia: A Case Report

Megaria Sihombing[1*], Santi Rini[2], Nisa Alyananda Ritonga[3], Sonia Edna Rumondang[4], Nurmasithah[5]
Universitas Mulawarman, Indonesia[1]
RSUD Aji Muhammad Parikesit, Indonesia[2]
RSUD Sultan Syarif Mohamad Alkadrie, Indonesia[3]
RSUD Depati Bahrin, Indonesia[4]
RSUD dr. Abdul Aziz Singkawang, Indonesia[5]
Email: megariashg@gmail.com[1*], dr.santi7@yahoo.co.id[2], dr.nisaritonga@gmail.com[3], soniamanikba@gmail.com[4], nurmasithah.tata@gmail.com[5]

Keywords:
Hirschsprung Disease; Hirschsprung-Associated Enterocolitis; Infant

Abstract

Hirschsprung's disease (HD) is a congenital disorder characterized by the absence of ganglion cells in the Meissner's and Auerbach's plexuses. The incidence of HD has been reported to be higher in Asian infants; nevertheless, HD cases in Indonesia remain underexamined. Delayed diagnosis places neonatal patients with HD at a 13-fold increased risk of developing Hirschsprung-associated enterocolitis (HAEC) preoperatively, which may be a life-threatening condition. A 51-day-old female infant was brought to the emergency room with abdominal distension and vomiting, and had bowel movements only 1–2 times per week, with an estimated volume of approximately 5 cc per episode. Radiographic examination showed a narrowed sigmoid colon and constriction of the rectosigmoid region, accompanied by proximal colonic dilatation. A HAEC score was calculated, yielding a result of 14, consistent with Hirschsprung-associated enterocolitis. Management in this case followed the American Pediatric Surgical Association Board of Governors Guidelines. A transanal Swenson procedure with frozen section biopsy was performed, and intraoperative findings confirmed short-segment Hirschsprung-associated enterocolitis. The diagnosis and patient management were carried out appropriately in accordance with established guidelines and clinical standards.

INTRODUCTION

Neonatal intestinal obstruction is one of the most common causes of neonatal surgical emergencies. Hirschsprung's disease (HD) is a common cause of bowel obstruction in the neonatal population (1). HD is a congenital disorder defined by the absence of ganglion cells in the Meissner's plexus of the submucosa and the Auerbach's plexus of the muscularis in the terminal rectum, extending proximally for a variable distance (2) (Karim et al., 2021). Its prevalence ranges from 1 to 1.63 per 10,000 births and shows an overall male predominance of approximately 4:1 worldwide (2) (Hagens et al., 2022). The incidence of HD has been reported to be higher in Asian infants compared with infants from other ethnic groups (3) (Karim et al., 2021). Nevertheless, HD cases in Indonesia remain underexamined. Such limited awareness often leads to delayed diagnosis and treatment, increasing morbidity and mortality, including Hirschsprung-associated enterocolitis (HAEC). HAEC is the most common and severe complication of HD (Veras & Gosain, 2022). Although the mortality rate of HAEC has declined, it remains a major cause of morbidity and mortality in HD, with reported mortality occurring in up to 50% of severe cases (Trinidad et al., 2022). The number

of HD cases at Aji Muhammad Parikesit Hospital in 2018, 2019, and 2020 were 19, 19, and 21 cases, respectively, with an average male-to-female ratio of 1.7:1.

Although short-segment HD is generally associated with a lower incidence of preoperative HAEC compared with longer-segment disease, delayed diagnosis can significantly negate this advantage and lead to life-threatening enterocolitis (10) (Khorana et al., 2021). In developing countries and geographically challenging regions such as East Kalimantan, Indonesia, systemic barriers—including limited early recognition at primary healthcare facilities and delayed referrals—contribute significantly to late presentations (12) (Trinidad et al., 2022). The clinical management of HAEC in this setting requires strict adherence to both the Indonesian Ministry of Health National Guidelines (PNPK Kemenkes) (13) and international standards, such as the American Pediatric Surgical Association (APSA) guidelines (6). This case report aims to highlight the management of severe HAEC in a 51-day-old infant with delayed diagnosis of short-segment HD at a regional hospital in East Kalimantan, and to evaluate the integration of these guidelines in optimizing clinical outcomes.

METHOD

This study was conducted as a retrospective case report at Aji Muhammad Parikesit Hospital, Kutai Kartanegara, East Kalimantan, Indonesia. Clinical data, including patient history, physical examination findings, radiological results, surgical interventions, and postoperative follow-up, were extracted from the hospital medical records. The case was reported in accordance with the CARE guidelines to ensure standardized reporting (14). Written informed consent was obtained from the patient's parents for publication of the case report and accompanying clinical data. Management was evaluated based on current national guidelines (PNPK Kemenkes) and the American Pediatric Surgical Association (APSA) guidelines for Hirschsprung-associated enterocolitis (HAEC).

Case Report

1. History

A 51-day-old female infant was admitted with chronic constipation starting from the age of 7-day-old. The patient could only defecate spontaneously 1-2 times per week, with liquid yellow feces about 5 cc per defecation accompanied by explosive defecation. The faces had foully smell, and no rectal bleeding and mucus was recorded. The stomach kept bloating since 7-day-old. Vomiting of breast milk or formula milk that has just been drunk occurred 1 to 3 times a day. The infant was restless and fussy with excessive thirst. There were no intermittent lumps in the abdominal and groin area.

Pregnancy Record: The mother followed routine antenatal care (ANC) once per month from the 5th – 28th week of gestation at the Pusat Kesehatan Masyarakat (Puskesmas) followed by ANC twice per month from 29th week of gestation until birth at the hospital. Iron tablets were taken 1 tablet per day during the 2nd trimester, while tablets of folic acid were taken oncee to twice a week.

Birth record: The patient was a 5th child born through cesarean section due to her breech position and oligohydramnios with a gestational age of 38 weeks and a birth weight of 2950 grams. The baby cried spontaneously after birth. The infant was diagnosed with neonatal jaundice and was taken for phototherapy for 3 days from birth. Meconium was passed on the

2nd day after birth. The baby was fed with formula milk since 7-day-old due to the mother's breastfeeding difficulty. The formula milk was changed three times since 7 day old up until the patient's admission.

Family health history: No similar case was reported in the patient's siblings. Thyrotoxicosis was found in both grandmothers of the infant.

2. Clinical Findings

The patient was admitted on 51-day-old to the ER. Vital signs examination showed compos mentis consciousness (GCS: E4V5M6), temperature 36.2o Celsius, respiratory rate 28 times per minute, and pulse rate 128 times per minute. The patient's weight was 4.0 kg.

Upon examination, the patient was breathing with thoraco-abdominal movement. During abdominal inspection, the patient showed distension without darm contour and darm steifung. On auscultation, bowel sounds were normal with tympanic sound over the entire abdominal region on percussion. On palpation, the abdomen was soft, with an abdominal circumference of 33 cm. No abnormalities were found on other body parts.

First laboratory test showed slight elevated leukocyte count (10.400/mm3), elevated platelet count (617.000/mm3), decreased hemoglobin (8,7 gr/dl) and elevated total bilirubin (1,48 mg/dl; 0,49 conjugated bilirubin, 0,99 unconjugated bilirubin). No other abnormality was found in other laboratory results.

An orogastric tube was placed on the infant for decompression, followed by intravenous fluid resuscitation and PRC transfusion. Antibiotics were given intravenously (Cefotaxime 200 mg/12 hours and Metronidazole 40 mg/8 hours).

The patient was then taken for radiologic examination (Figure 1). Abdominal X-Ray showed increasing amounts of distribution and intestinal air, with dilatation of the structure of the large bowel and visible fecal material. Colon in-loop examination showed reductant of sigmoid colon and narrowing of the anal sphincter region to the rectum accompanied by widening of the intestinal structure in the proximal part and visible fecalite.

The patient was suspected with Hirschsprung-associated enterocolitis, and was admitted to the pediatric ward. The HAEC Score on the infant was 14. Surgery was scheduled after the patient was declared hemodynamically stable. 6 days after admission, the patient underwent a surgery with trans anal pull-through procedure and frozen section biopsy.

The perineal dissection began with the placement of circumferential 2-0 silk traction sutures from the dentate line to the perineum 2 cm from the anus. A needle-tipped electrocautery was used to incise the rectal mucosa approximately 0,5 - 1 cm circumferentially from proximal to the anal columns. Fine silk traction sutures were then placed in the rectal mucosa to help retract the mucosa during circumferential dissection. The muscular cuff of the rectum had been divided, and the ganglionic colon had been exteriorized through the anal canal. A frozen section biopsy was performed with the following results:

a. Preparation I: adequate plexus, 1 immature ganglion
b. Preparation II: adequate plexus, 1 immature ganglion
c. Preparation III: adequate plexus, 2-3 mature ganglions
d. Preparation IV: adequate plexus, > 3 mature ganglions

The anastomosis was performed 11 cm proximally from the anal column (Figure 2). The pull-through colon was completely transected above the biopsy site and was made ready

for the coloanal anastomosis. A rectal tube and tampon were inserted, and the surgery was completed after 2 hours and 15 minutes.

After the procedure, the patient was admitted to the Pediatric Intensive Care Unit (PICU). The patient. The patient was given broad spectrum antibiotics (Cefotaxime 200 mg/12 hours IV and Metronidazole 40 mg/8 hours IV) Paracetamol 40 mg/8 hours IV, and Gentamicin cream on anal skin / 12 hours. Nasogastric tube (NGT) and urinary catheter were inserted for evaluation. After 5 days in PICU, the rectal tube, NGT, and urine catheter were taken out and the patient was admitted back to the pediatric ward and the intravenous medications were replaced into oral medications.

Observation was conducted for 2 days in the pediatric ward, and the patient was discharged. The patient was scheduled for a follow-up and first rectal dilatation 2 weeks post-surgery. The first rectal dilatation procedure was done using general anesthesia. Anal dilation was conducted using dilator no. 6, 7, 8, 9, 10, 11, 12, and 13 with gel lubricant. The evaluation of the coloanal anastomosis was carried out for 10 minutes at no. 13 dilator. It was then removed and followed by bleeding evaluation. The procedure lasted for 30 minutes with 0.1 cc bleeding. 12 hours after the anal dilation, the patient was discharged and scheduled for a routine follow-up every 30 days up until 3 years old.

RESULT AND DISCUSSION

HD is a disorder of distal intestinal motility that is caused by congenital absence of ganglion cells in the myenteric plexuses of the rectum and the distal colon. It usually presents in infancy but can present at any age (4). Late diagnosis of HD is life-threatening. The infant's breathing will be disturbed due to the fact that abdominal distention will inhibit the diaphragm muscle in the infant, which is the main respiratory muscle in infant (5) (Veras & Gosain, 2022). The patient in this case was presented with chronic constipation, fouly stool, breast milk reflux since the patient was 7 day old, and abdominal distention. The patient was brought to the hospital and was treated for an emergency case. The patient was decompressed with OGT No 8 for maintaining the diaphragm muscle compression. Moreover, fluid balance monitoring every 3 hours and abdominal circumference monitoring every 8 hours was done in this case.

Infants with HD will have corrective surgery depending on their overall health and degree of colon distension. The goal of pre-operative management is to prevent enterocolitis which is an inflammatory disorder of the bowel because of the fact that HAEC represents the leading cause of serious morbidity and mortality (Hagens et al., 2022). The classic manifestations of HAEC include abdominal distention, fever, and diarrhea (Veras & Gosain, 2022). However, there is a broad clinical spectrum with which children present, and other signs or symptoms may include vomiting, rectal bleeding, lethargy, loose stools, and obstipation (6). Most HAEC cases occur during the preoperative phase (7) (Hagens et al., 2022). In the present study, the estimated overall prevalence of preoperatively diagnosed HAEC is 18,5% (Hagens et al., 2022). Developed countries with good knowledge of the disease allows them for an early diagnosis and management, but in developing countries the diagnosis is often late (8) (Trinidad et al., 2022). Late diagnosis of HD itself is a risk factor to develop HAEC.

Studies showed that the incidence of HAEC is lower in newborns identified with HD within the first week of life (11%) than in those diagnosed later in life (9) (Khorana et al., 2021). In another study, compared to diagnosis made within 7 days after birth, delayed diagnosis put neonatal patients with HD at a 13-fold increased risk for HAEC before surgery (10) (Ali et al., 2021). The prevalence for preoperative HAEC is lowest in short segment HD (15,2%) compared to long segment HD and total segment HD (7) (Hagens et al., 2022). Long-segment HD that is aganglionosis proximal to the splenic flexure, may increase the risk of HAEC by causing luminal stasis due to dysmotility of the residual segment. A study found that patients with long-segment disease had much greater rates of HAEC than those with short-aganglionic segment disease (11). In this case, the patient had a short-aganglionic segment with HAEC. Based on the clinical manifestations of this patient, the presence of HD should have been suspected during the first 7 days, but because of the diagnosis delay, HAEC still occurred even though the patient had short segment HD.

HAEC suspicion on this patient is based on HAEC Score. A score of 10 or higher was associated with a positive diagnosis by an international panel of experts (1) (Luzman et al., 2021). In this case, the patient's Score was 14 (Table 1).

The American Pediatric Surgical Association Board of Governors has categorized the clinical suspicion and severity of HAEC into three grades based on history, physical examination, and imaging study. This category is not intended as a scoring system, but rather a decision-support tool to ensure that all of the relevant history, examination and radiographic findings are considered. In general, presence of higher-grade findings should prompt providers to assign the higher grade (6). The patient in this case was categorized into grade II.

Treatment was based on The American Pediatric Surgical Association Board of Governors Guidelines, as detailed in Table 2 (6). Management of HAEC on this guideline is based on the severity of the clinical presentation. In this case, the patient was given Metronidazole 40 mg IV and Cefotaxime 200 mg IV, a combination of antibiotics. Both were given immediately in the ER and were maintained until day 7 post-surgery.

Once the diagnosis of HD is suspected, it must be confirmed by rectal biopsy (1). In this case, the frozen section biopsy was performed during the surgery and diagnosis of HD is made (Gunadi et al., 2020). The goals of surgical management for HD are to remove the aganglionic bowel and to reconstruct the intestinal tract by bringing the normally innervated bowel down to the anus while preserving normal sphincter function. The most commonly performed operations are the Swenson, Duhamel, and Soave procedures, although the Rehbein and State procedures are still performed in some centers (1,2).

The patient underwent a Swenson procedure. The goal of the Swenson pull-through is to remove the entire aganglionic colon, with an end-to-end anastomosis above the anal sphincter. The operation was originally performed via a laparotomy, with the anastomosis being performed from the perineum after severing the aganglionic rectum. It is important to keep the dissection in the correct plane along the rectal wall to avoid injury to the deep pelvic nerves, vessels, and other structures such as the vagina, prostate, vas deferens, and seminal vesicles. Despite the theoretical risks inherent in the deep pelvic dissection, long-term functional outcomes after the Swenson procedure are excellent (1) (Pham et al., 2023; Zhang et al., 2023).

Most patients who underwent transanal pull-through can be fed immediately and discharged within 24-48 hours. However, the patient in this case was transferred to PICU for 5 days post-surgery to reduce the risk of the rectal tube being displaced, with strict fluid balance monitoring until the patient was stable and NGT evaluation.

The anastomosis is calibrated with an appropriately sized dilator or finger 1–2 weeks after the procedure. Although most surgeons instruct the parents to perform daily dilatations, a program of weekly calibration by the surgeon is less traumatic and is associated with similar outcomes. In this case, the combination of both was performed. The first dilatation was done in the OR 2 weeks post-surgery to avoid neurogenic shock. The patient's family were also taught to do daily anal dilatation on this patient, with monthly dilator's size calibration on the outward clinic. Long-term problems in children with HD include ongoing obstructive symptoms, soiling, and enterocolitis (Davidson et al., 2021; Dai et al., 2020). The patient in this case will be monitored monthly at least until they've passed the toilet training period at 3-year-old (1).

CONCLUSION

The diagnosis and management of this patient were effectively carried out by integrating national guidelines and American Pediatric Surgical Association (APSA) recommendations, despite the limitations of a regional hospital setting. This case underscores that although short-segment Hirschsprung's disease (HD) is generally associated with a lower risk of preoperative Hirschsprung-associated enterocolitis (HAEC), delayed diagnosis—often driven by systemic referral barriers and limited early detection in developing regions—can still result in life-threatening complications. Early recognition of chronic neonatal constipation at the primary healthcare level is essential. Prompt bowel decompression, appropriate broad-spectrum antibiotic therapy, and fluid resuscitation, followed by definitive pull-through surgery, remain the cornerstone of management to reduce mortality and achieve favorable long-term functional outcomes in delayed presentations of HD.

REFERENCES

Ali, A., Haider, F., & Alhindi, S. (2021). The prevalence and clinical profile of Hirschsprung's disease at a tertiary hospital in Bahrain. *Cureus, 13*(1), e12480. https://doi.org/10.7759/cureus.12480

Davidson, J. R., Kyrklund, K., & Eaton, S. (2021). Long-term surgical and patient-reported outcomes of Hirschsprung disease. *Journal of Pediatric Surgery, 56*(7), 1502–1511. https://doi.org/10.1016/j.jpedsurg.2021.01.054

Dai, Y., Deng, Y., Lin, Y., Ouyang, R., & Li, L. (2020). Long-term outcomes and quality of life of patients with Hirschsprung disease: A systematic review and meta-analysis. *BMC Gastroenterology, 20*(1), 64. https://doi.org/10.1186/s12876-020-01208-z

Frykman, P. K., & Short, S. S. (2012). Hirschsprung-associated enterocolitis: Prevention and therapy. *Seminars in Pediatric Surgery, 21*(4), 328–335.

Gunadi, Sukarelawanto, A. V. R., Ritana, A., Balela, N., Putri, W. J. K., & Sirait, D. N. (2020). Postoperative enterocolitis assessment using two different cut-off values in the HAEC score in Hirschsprung patients undergoing Duhamel and Soave pull-through. *BMC Pediatrics, 20*(1), 457. https://doi.org/10.1186/s12888-020-02825-4

Hagens, J., Reinshagen, K., & Tomuschat, C. (2022). Prevalence of Hirschsprung-associated enterocolitis in patients with Hirschsprung disease. *Pediatric Surgery International, 38*(1), 3–15. https://doi.org/10.1007/s00383-021-05020-y

Karim, A., Tang, C. S., & Tam, P. K. H. (2021). The emerging genetic landscape of Hirschsprung disease and its potential clinical applications. *Frontiers in Pediatrics, 9*, 638093. https://doi.org/10.3389/fped.2021.638093

Khorana, J., Phiromkanchanasak, P., Kumsattra, J., Klinoun, S., Aksorn, S., Chantakhow, S., Tepmalai, K., & Singhavejsakul, J. (2021). Development of clinical referral score model for early diagnosis of Hirschsprung's disease in suspected pediatric patients. *Healthcare, 9*(6), 678. https://doi.org/10.3390/healthcare9060678

Lee, C. C., Lien, R., Chiang, M. C., Yang, P. H., Chu, S. M., Fu, J. H., et al. (2012). Clinical impacts of delayed diagnosis of Hirschsprung's disease in newborn infants. *Pediatrics and Neonatology, 53*(2), 133–137.

Luzman, R. A., Kencana, S. M. S., Arthana, B. D., Ahmad, F., Sulaksmono, G., Rastaputra, A. S., Arini, G. P., Pitaka, R. T., Dwihantoro, A., & Makhmudi, A. (2021). Comparison of two different cut-off values of scoring system for diagnosis of Hirschsprung-associated enterocolitis after transanal endorectal pull-through. *Frontiers in Pediatrics, 9*, 705663. https://doi.org/10.3389/fped.2021.705663

Pham, T. H. N., Dang, Q. V., Nguyen, T. P. T., Le, H. T. M., Trinh, N. T., Trung, N. T., Nguyen, T. D., & Nguyen, H. N. (2023). Adopting the Swenson-like technique for patients with Hirschsprung disease in Vietnam. *Pediatric Surgery International, 39*(1), 110. https://doi.org/10.1007/s00383-023-05415-z

Trinidad, S., Kayima, P., Kotecha, V., Massenga, A., Rymeski, B., Frischer, J. S., Situma, M., & Kotagal, M. (2022). Hirschsprung's disease in low- and middle-income countries. *Seminars in Pediatric Surgery, 31*(2), 151163. https://doi.org/10.1016/j.sempedsurg.2022.151163

Veras, L. V., & Gosain, A. (2022). Current understanding of Hirschsprung-associated enterocolitis: Pathogenesis, diagnosis and treatment. *Seminars in Pediatric Surgery, 31*(2), 151164. https://doi.org/10.1016/j.sempedsurg.2022.151164

Xie, C. M., & Yao, Y. T. (2021). Anesthesia management for pediatrics with congenital heart diseases who undergo cardiac catheterization in China. *Journal of Interventional Cardiology, 2021*, 8861461. https://doi.org/10.1155/2021/8861461

Zhang, Z., Li, Q., Li, B., Alganabi, M., & Li, L. (2023). Long-term bowel function and pediatric health-related quality of life after transanal rectal mucosectomy and partial internal anal sphincterectomy pull-through for Hirschsprung disease. *Frontiers in Pediatrics, 11*, 1099606. https://doi.org/10.3389/fped.2023.1099606