Management and Outcome of Hirschsprung's disease in Children: A Literature Review

Donny Aditia[1], Naisya Balela[2], Hery Poerwosusanta[1]

[1]Departement of Surgery, Faculty of Medicine, Lambung Mangkurat University

[2]Division of Pediatric Surgery, Faculty of Medicine, Public Health and Nursing, Gadjah Mada University,

Indonesia

Correspondence: *drdonnyaditia@gmail.com*

ABSTRACT

Hirschsprung disorder (HD) results from a malfunction in the innervation of the intestine that starts evolving on the inner anal sphincter and extends proximally to the period of the gut that varies, however constantly includes the anus and at the least part of the rectum. The systematical evaluations were culled from full-text English guides posted in the previous ten years (variety 2012-2022). This takes a look at pursuits to evaluate the management of Hirschsprung ailment in children. the primary treatment for HD patients is surgery. some of the surgical strategies used, the Duhamel approach was the maximum used (forty-eight%), followed by the aid of the transanal pull-thru approach (35%). Enterocolitis is a hazard of significant complications for sufferers of Hirschsprung's disease. it could strike at any age, however, is maximum at 2-4 weeks of age, although it can be determined at the age of 1 week. other symptoms encompass persistent constipation, diarrhea, abdominal distension, foul-smelling stools, and fever.

Keywords: Hirschsprung, Children, Surgery

INTRODUCTION

Hirschsprung disease (HD) effects by a malfunction inside the innervation of the intestine that starts evolving at the inner anal sphincter and extends proximally to the period of the gut that varies, however always consists of the anus and at the least a part of the rectum.[1] Hirschsprung disorder takes place in approximately 1 case / 5400-7200 newborns every 12 months. worldwide research has said fees ranging from approximately 1 case according to 1500-7000 newborns even though the exact worldwide occurrence is unknown.[1,2] A hollow muscular tube that stretches from the cecum to the anal canal, the huge gut is lots wider in diameter than the small intestine.it's far approximately 5 feet in period and is made of two sections. the standard anus is around 2.5 inches (about 6. five cm) in circumference, even though the nearer the anus is to the center, the smaller its miles. The

cecum, colon, and rectum are the 3 divisions of the big intestine.[3] A hole muscular tube that stretches from the cecum to the anal canal, the big intestine is an awful lot wider in diameter than the small gut. it's miles about 5 feet (about 1. five m) in length and is made up of sections. the typical anus is around five inches (about 6.5 cm) in circumference, even though the nearer the anus is to the center, the smaller its miles. The cecum, colon, and rectum are the three divisions of the massive intestine.[4]

Hirschsprung disease is a result of a failure of the parasympathetic nerve cells of the myenteric to emigrate from the cephalon to the caudal hemisphere of the brain. This results in ganglion cells no longer being recognized starting from the anus, and their period varies proximally due to this. Down syndrome, neurocristopathy, Waardenburg-Shah syndrome, piebaldism, Goldberg-Shprintzen syndrome, type II a couple of Endocrine Neoplasia, primary relevant hypoventilation (Ondine's curse), and Chagas disorder are amongst situations wherein Hirschsprung disorder can be diagnosed.[4] Abnormalities inside the intestine wall's microenvironment will inhibit neural crest cells from migrating or differentiating. patients with Crohn's sickness have been proven to have higher antigen major histocompatibility complicated (MHC) elegance 2 levels inside the aganglionic region of the gut.[5]

Myenteric and submucosal plexuses are absent in sufferers with HD. most of the time, the anus is implicated, and the aganglionosis might increase to the proximal nerve. The hobby of the cholinergic and adrenergic structures within the intestine is two-3 times more than that of the wholesome gut. because of this condition, clean muscle contractility becomes unbalanced, inflicting disorganized peristalsis and useful blockage. four The extensive majority of HD times are sporadic, with simply 10% of Hirschsprung's sufferers having a circle of relatives and loved ones who are also affected by the circumstance. sufferers with longer segment infection are more likely to look at this sample. Prenatal ultrasonography most effectively detects intestinal blockage in a small percentage of instances, except in instances related to the complete colon.[4]

METHODS

The data for this systematic review were culled from full-text English publications published in the preceding ten years (range 2012-2022). This study aims to assess the management of Hirschsprung disease in children. We utilized the databases to produce this article Pubmed and Google Scholar. In this investigation, the PICO analysis was used to include pediatric patients with Hirschsprung disease, the index was the management of the patient with intussusception, without comparisons and the objective was the patient outcome. The research comprised both clinical trials and randomized clinical trials.

This analysis followed the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) paradigm, in which the researchers originally entered keywords into each database. The phrases "management" and "hirschsprung disease in children" were used in the search. (("manage"[All Fields] OR "managed"[All Fields] OR "management s"[All Fields] OR "managements"[All Fields] OR "manager"[All Fields] OR "manager s"[All Fields] OR "managers"[All Fields] OR "manages"[All Fields] OR "managing"[All Fields] OR "managment"[All Fields] OR "organization and administration"[MeSH Terms] OR ("organization"[All Fields] AND "administration"[All Fields]) OR "organization and administration"[All Fields] OR "management"[All Fields] OR "disease management"[MeSH Terms] OR ("disease"[All Fields] AND "management"[All Fields]) OR "disease management"[All Fields]) AND ("hirschsprung disease"[MeSH Terms] OR ("hirschsprung"[All Fields] AND

"disease"[All Fields]) OR "hirschsprung disease"[All Fields]) AND ("child"[MeSH Terms] OR "child"[All Fields] OR "children"[All Fields] OR "child s"[All Fields] OR "children s"[All Fields] OR "childrens"[All Fields] OR "childs"[All Fields])) AND ((y_10[Filter]) AND (clinicaltrial[Filter] OR randomizedcontrolledtrial[Filter])). The researchers received five articles, which will be discussed during the discussion **(Table 1)**.
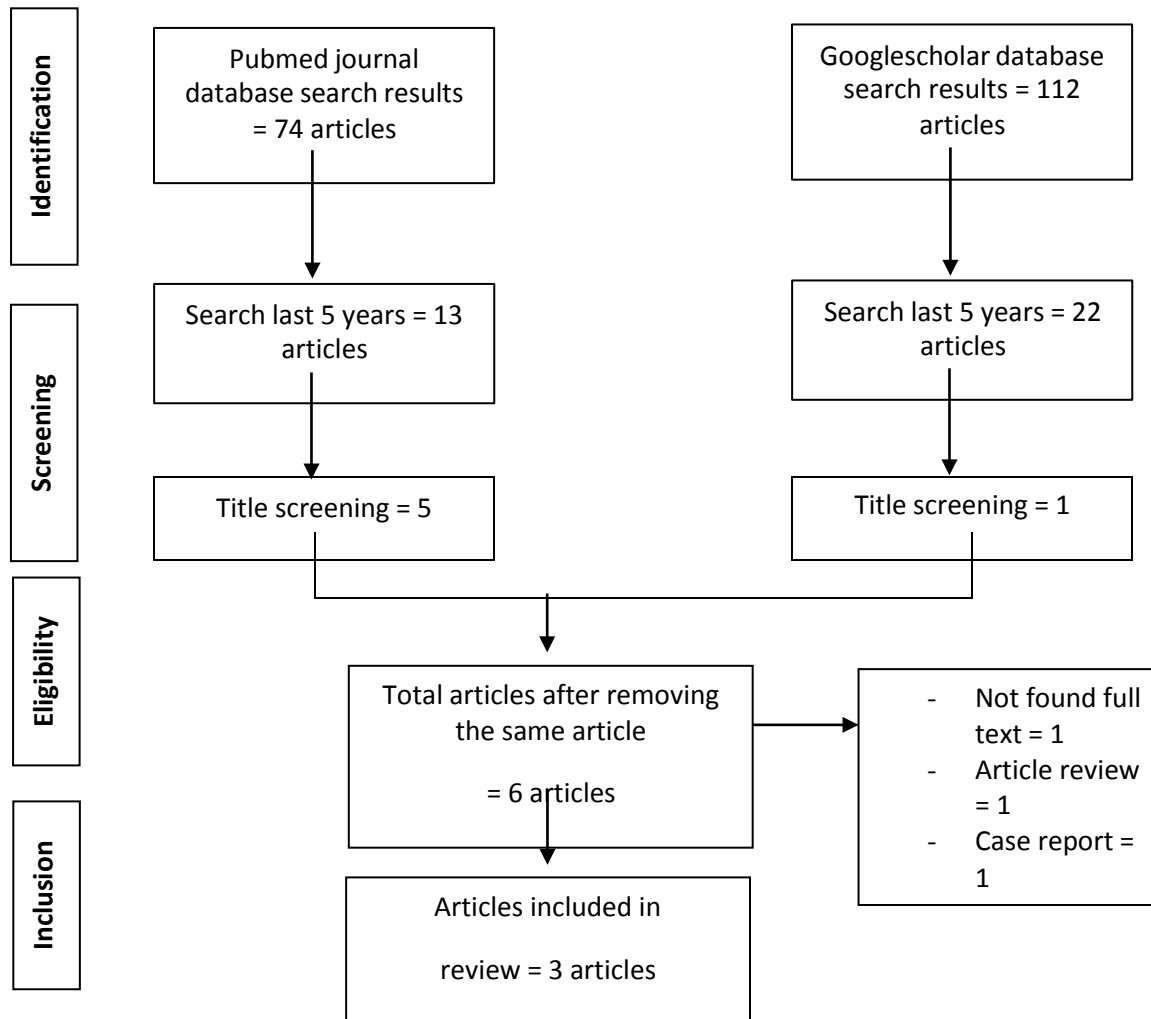

Figure 1. Article search flowchart

RESULT

Tang, 2020 conducted a study on 73 and 75 patients assigned to the traditional and enhanced recovery after surgery (ERAS) groups, respectively. Length of stay (LOS) decreased from 9.5 days in the traditional group to 7.9 days in the ERAS group (P < 0.001). White blood cell (WBC) count on a postoperative day (POD) 1 showed no significant difference between the two groups. C-reactive protein (CRP) on POD 1 in the ERAS group was significantly lower (P < 0.001). In the ERAS group, the blood glucose was higher during anesthesia compared to the TRAD group (P < 0.001). The blood glucose at 24 h after surgery was significantly lower in the ERAS group (P < 0.001).[6]

Intraoperative fluid volume was lower in the EARS group (P < 0.001). ERAS could also reduce the time to first defecation (P < 0.001), discontinuation of intravenous infusion (P < 0.001) and regular diet (P < 0.001). In the ERAS group, the concentrations of albumin and retinol conjugated protein on POD 5 were higher than those in the TRAD group (P < 0.001, P < 0.001, respectively). The plasma atrium had no difference in the two groups on POD 5 (P > 0.05). The rate of complications (P > 0.05) and 30-day re-admission (P > 0.05) were not significantly different between the two groups.[6]

Hospitalization costs were also reduced (P <0.001). ERAS group has a higher parental satisfaction rate, although there was no statistical difference (96% vs 89%). There was no difference in growth between the ERAS and the TRAD groups from admission to 6 months after the surgery (weight for age z score: P > 0.05, weight for length z score: P > 0.05). We also found that the shortening of LOS by the application of ERAS protocol was more obvious in the OPPT group (-2.5 ± 1.0) than that in the transanal endorectal pull-through (TEPT) (-1.9 ± 1.3) and laparoscopic-assisted pull-through (LAPT) (-1.3 ± 0.4) groups.[6]

Zhang et al (2020) showed outcomes within 1 month after surgery indicated a lower incidence of abdominal distention (4% vs 15.5%, P < 0.05) and postoperative HAEC (1.2% vs 6.8%, P < 0.05) in group A compared to group B. Beyond 1 month after surgery, the overall incidence of HAEC was

Table 1. The litelature include in this study

Author	Origin	Method	Sample Size	Period	Result
Garcia, 2015[7]	Spain	Case-crossover	95 cases	2006 to 2009	76.6 percent were less than the age of 2 years out of a total of 95 cases chosen; 65.3% were male and 34.7% were female. The link with intussusception and past drug use varied by exposure window: odds ratio (OR) = 1.45 (95% CI = 0.86–2.43); OR = 1.46 (95% CI = 0.80–2.67); and OR = 2.26. (95% CI = 1.10-4.64). These relationships were more pronounced in children less than 2 years and were often attributable to the recent administration (within the previous 2 days) of antibiotics (OR = 8.00; 95% CI = 1.47-43.7).
Jamshidi, 2022[8]	Iran	RCT	52 cases	March 2015 to February 2018	They had 52 patients who needed surgical exploration (26 in each group). There were four (15%) and seven (27%) patients with self-reduced intussusception in LS and OS groups, respectively. The conversion rate was 31% (eight cases). Five cases (19%) in the LS group and four cases (15%) in the OS group needed bowel resections. Operating time was longer in the LS group (P $\leq$ 0.006), and the postoperative complication rate was higher in the OS group (P $\leq$ 0.021).
Wei, 2015[9]	Taiwan	Clinical trial	23 and 35 patients in LAP and OPEN group	January 2007 and July 2013	No significant difference was found on age, operative indication, surgical procedure, type of intussusception, level of intussusceptum, and presence of spontaneously reduced intussusception between both groups. In LAP group, mean OP time was significantly longer; mean PO time and LOS were significantly shorter. One surgical recurrence occurred in each group (p = 0.76). In comparison of LAP-IP (n = 15) and LAP-NIP (n = 8), OP time, PO time, and LOS were similar in both subgroups. One recurrence was noted in LAP-IP (p

					= 0.46). The overall conversion rate was 13.0 % (6.8 vs. 25 %, p = 0.21). Compared to patients with intussusceptum to ascending colon, the conversion rate was significantly higher in patients with intussusceptum to transverse and descending colon. With the exclusion of conversion, OP time was significantly shorter in LAP-NIP (p = 0.01).
Vazquez, 2012[10]	Spain	Prospective study	19 patients	August 2009 till January 2012	Complete reduction exclusively by external manual reduction was accomplished on 12 occasions (80%). In the remaining three procedures, partial reduction to the cecum was obtained. Subsequent enema achieved complete reduction in two. Overall non-surgical reduction rate was 93%.
Zhang, 2015[11]	China	Retrospective study	234 children	January 1, 2011 and December 30, 2013	They determined that the overall antibiotic use rate following successful air enema reduction was 41% (97/234), which decreased from 99% (67/68) in phase I to 18% (30/166) in phase II. In phase I, prophylactic antibiotic usage reached up to 84% (56/67). The quantity of aztreonam for injection accounted for 63% (45/71), and cefamandole nafate for injection accounted for 25% (18/71). In phases II, prophylactic antibiotic usage were reduced to 13% (4/30). The quantity of aztreonam for injection was decreased to 12% (4/33) and cefamandole nafate for injection was 3% (1/33).

due to the recent administration (preceding 2 days) of antibiotics (OR, 8.00; 95% CI, 1.47-43.7).[7]

Jamshidi, *et al* conducted a study with 52 patients who needed surgical exploration (26 in each group). There were four (15%) and seven (27%) patients with self-reduced intussusception in LS and OS groups, respectively. The conversion rate was 31% (eight cases). Five cases (19%) in the LS group and four cases (15%) in the OS group needed bowel resections. Operating time was longer in the LS group (P ≤ 0.006), and the postoperative complication rate was higher in the OS group (P ≤ 0.021).[8]

Wei, *et al* showed mean PO time and LOS were significantly shorter in laparoscopy group. One surgical recurrence occurred in each group (p = 0.76). OP time, PO time, and LOS were similar in both subgroups. One

recurrence was noted in LAP-IP (p = 0.46). The overall conversion rate was 13.0 % (6.8 vs. 25 %, p = 0.21) compared to patients with intussusceptum to ascending colon, the conversion rate was significantly higher in patients with intussusceptum to transverse and descending colon. With the exclusion of conversion, OP time was significantly shorter in LAP-NIP (p = 0.01).[9]

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DISCUSSION

Intussusception was first described by Paul Barbette in Amsterdam (1674). Jonathan Hutchinson was the first to perform a successful intussusception operation on a 2-year-old child in 1873. The word intussusception comes from the Latin *intus* (in) and *suscipere* (to receive). Intussusception is the invagination of one part of the intestine into another. Three cylinders in the intestinal wall are involved. The inner cylinder and the middle cylinder are the invaded intestine (intussusceptum), and the outer cylinder is the recipient of the intestinal invagination (intussuscipiens).[12]

Most etiologies of primary invagination in children are unknown. Viruses and intestinal cancers may cause invagination in children. In the past, invagination was linked to rotavirus vaccination. Rotavirus causes diarrhoea, vomiting, fever, and dehydration. Tumors of the gastrointestinal system, intestinal adhesives, surgical wounds of the small intestine and colon, IBS, and Hirschsprung's may induce invagination. Payer's patch hypertrophy in the ileum may cause invagination by stimulating intestinal peristalsis. Invagination occurs often following upper respiratory infections and gastroenteritis, leading lymphoid tissue enlargement.[13–16]

Adenovirus is present in 50% of invaders. Because of their high viral vulnerability, children aged 6-36 months are most susceptible. Intussusception causes include an inverted appendix, Meckel's diverticulum, intestinal polyps, duplication, or lymphosarcoma. Intussusception may develop in dehydrated cystic fibrosis patients. Males had more reported instances of intussusception than women, generally 2:1 or 3:1, usually 78 percent of men at 9 months.[17]

Intussusception was shown to be more prevalent in males less than 2 years of age. When medicines were delivered 2 to 7 days before the development of symptoms in children younger than the age of 2 years.[7] Prescription medication use was related with intussusception, particularly in the shorter time range. The outcomes were larger for children under 2 years. The relationship between past prescription medication usage and intussusception was obviously positive for boys in the gender-specific analysis. The associations were not evident or substantial for girls.[18]

Invagination is an emergency case, so that urgent action is needed in the form of improving the patient's general condition, inserting a gastric sonde to decompress and prevent aspiration, rehydration, sedatives for pain relief. After the general condition is good, surgery is done, if there are clear signs of intestinal obstruction. Or do repositioning if there are no contraindications. The basis of treatment for invagination is repositioning of the intestine into the lumen of the other intestine. Repositioning can be achieved by barium enema, pneumostatic repositioning or surgically.[19]

Indications for manual reduction are in patients who are unstable, have an increase in temperature and leukocyte count, experience prolonged symptoms or are found to have advanced disease characterized by abdominal distension, bloody stools, severe intestinal system disorders until shock or peritonitis occurs. The patient is immediately prepared for a Laparotomy operation with a transverse interspinal incision. If an abnormality is found to have necroses, reduction does not need to be performed and resection is performed immediately.[8,9]

Initially, the use of laparoscopy in intussusception was used only for diagnosis also in cases with doubtful radiological examinations or the presence of suspected pathological lesions. Once the diagnosis was confirmed, the operation was changed to a laparotomy. Recent studies have shown variable success in laparoscopic reduction of intussusception. Various techniques have been reported, but the majority of minimally invasive approaches describe the use of three abdominal ports: one in the infraumbilical region with the other two ports along the left side of the abdomen.[5,12]

Laparoscopic reduction is completed by applying gentle pressure distal to the intussusceptum using atraumatic graspers. Although counterintuitive to conventional open methods, traction is usually required proximal to the intussuscipiens to complete reduction. A careful examination is then performed to evaluate for signs of ischemia, necrosis, or perforation. If a resection is required, this can sometimes be accomplished by removing the bowel through a periumbilical incision. If this cannot be completed safely, the operation should be changed to an open lalarotomy.[5,12]

Laparoscopy as a screening technique confirmed intussusception and excluded many instances requiring OS. Only a simple laparoscopic evaluation might obviate the need for more extensive surgery. Laparoscopy also helped minimise intussusceptions and pull the ilium out of the cecum using the Chinese fan method, with no recurrences. Laparoscopy eliminates the necessity for OS in all but tight intussusception instances. Laparoscopy reduces the incidence of OS and its complications. Less stress to the abdominal wall tissues and viscera means less problems such postoperative paralytic ileus, adhesion bands, and surgical site infection.[8,9]

Non-surgical therapy of paediatric intussusception has a lengthy history, dating back to the 18th century. Hirschsprung, Ravitch, and a host of other contemporary writers trace their roots back to Hippocrates and the beginnings of modern systematic medicine. Vazquez showed complete reduction exclusively by external manual reduction was accomplished on 12 occasions (80%). In the remaining three procedures, partial reduction to the cecum was obtained. Subsequent enema achieved complete reduction in two. Overall non-surgical reduction rate was 93%.[10]

The administration of an image-guided (fluoroscopic or sonographic) enema (air, water, saline, water-soluble contrast agent, or barium) is the usual non-surgical therapy for intussusception. Invasiveness is minimised, the time of hospitalisation is reduced, and expenditures are saved by nonoperative reduction. There are less surgical and post-surgical hazards, such as the post-surgery sticky small intestinal blockage that occurs in 3–6 percent of patients following operational reduction, which has not been documented after non-operative reduction.[20]

Bowel perforation is a substantial enema reduction risk. Perforation by different procedures is rare, usually less than 1%. Most perforations are non-necrotic and do not necrosis. Pressures too strong or too fast to change are technical issues. Slowing down the enema and lowering the pressure may help prevent perforation. Infants under 6 months and symptoms lasting more than 36 hours are at risk for perforation.[21] Postoperative complications following laparotomy and laparoscopic intussusception include wound infection, fascial dehiscence, and small intestinal blockage.[5,22]

Complications (4%) would be fewer if no enterotomy or intestinal resection were required (26%). Adhesions in the small bowel may occur after surgery. Up to 20% of patients have recurrent intussusception. Most reoccur between 24 hours to 6 months. It frequently occurs after surgery and is idiopathic. Parents will recognise the signs of repeated intussusception and rush to the hospital. Multiple recurrences in the same kid indicate additional reasons, such as cancer. Ultrasound imaging is advised. When enema reduction fails or clinical problems continue after the operation, exploratory surgery is necessary.[5,22]

CONCLUSION

The patient's overall health must be improved, a stomach sonde placed to decompress and avoid aspiration, fluids and sedatives administered for pain management. Then, if there are evidence of intestinal blockage, surgery is performed. Up to 20% of patients have recurrent intussusception. Most reoccur between 24 hours to 6 months.

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