# Contrast Enema of Hirschsprung Disease as an alternative diagnostic tool in rural areas

*By* Lulik Inggarwati

WORD COUNT 4553
TIME SUBMITTED 03-JUN-2024 03:19PM
PAPER ID 109399967

**TYPE OF ARTICLE:** Original Article

# Contrast Enema of Hirschsprung Disease as an alternative diagnostic tool in rural areas

Lulik Inggarwati[1,2],Anni Masrurotun Najiyah Ziha Ul Haq [3], Indrastuti Normahayu[4,5], Solimun[6], Happy Kurnia Permatasari[7], Wisnu Barlianto[8], Respati Suryanto Dradjat[9,10]

[1]Doctoral Program in Medical Science, Faculty of Medicine, Brawijaya University, Malang, East Java, Indonesia

[2] Pediatric Surgery Division, Department of Surgery, Dr. Saiful Anwar General Hospital, Malang, East Java, Indonesia

[3] General Surgery Resident, Faculty of Medicine, Brawijaya University, Malang, East Java, Indonesia

[4] Department of Radiology, Faculty of Medicine, Brawijaya University, Malang, East Java, Indonesia

[5] Department of Radiology, Dr. Saiful Anwar General Hospital, Malang, East Java, Indonesia

[6] Department of Statistic, Brawijaya University, Malang, East Java, Indonesia

[7] Department of Biochemistry and Biomolecular, Faculty of Medicine, Brawijaya University, Malang, East Java, Indonesia

[8] Department of Pediatric, Faculty of Medicine, Brawijaya University, Malang, East Java, Indonesia

[9] Department of Orthopedic and Traumatology, Faculty of Medicine, Brawijaya University, Malang, East Java, Indonesia

[10] Pediatric Division (Department of Orthopedic and Traumatology, Dr. Saiful Anwar General Hospital, Malang, East Java, Indonesia

**CORRESPONDING AUTHOR**

**E-mail**: matahari63@ub.ac.id

## ABSTRACT

**Background and Objectives.** Hirschsprung disease (HSCR) is a disease caused by an absence of ganglion cells in the myenteric plexus and submucosa of the rectum and colon. Rectal biopsy, the gold standard for diagnosing HSCR requires histologic examination which may not be found in rural areas. Meanwhile the holistic diagnostic is essential for ensuring effective HSCR management. We aimed to measure the sensitivity, specificity, and accuracy of contrast enema of HSCR as an alternative diagnostic tool compared to suction rectal biopsy.

**Materials and Methods.** We conducted a cross-sectional study which included 35 suspected HSCR patients in Dr. Saiful Anwar General Hospital Malang Indonesia who met our criteria. We used independent t-test and chi-square to compare the parameter studies. We measured the sensitivity, specificity, and accuracy of contrast enema of HSCR using a confusion matrix. The odds ratio (OR) was also calculated to indicate the diagnosis of HSCR is correct compared to non-HSCR cases.

**Results.** The sensitivity, specificity, and accuracy of contrast enema of HSCR consecutively were 93.3%; 80%; and 91.42%. This present study also showed that the OR of contrast enema of HSCR is 11 (Confident interval (CI) 95%: 1.064—113.731). It means that a positive suction rectal biopsy contrast enema is 11 times more likely to be associated with HSCR than a negative study.

**Conclusions.** Our findings suggest that contrast enema of HSCR can be considered as a tool for diagnosing HSCR in rural areas lacking of histologic examination services.


**Keywords:** Accuracy, Diagnostic, Hirschsprung, Contrast enema, Sensitivity, Specificity

Abbreviations:

AP : Anteroposterior

CI : Confident interval

FN : False negative
FP : False positive
HAEC : Hirschsprung-associated enterocolitis
HSCR : Hirschsprung disease
NPV : Negative predictive value
OR : Odds ratio
PPV : Positive predictive value
RI : Rectosigmoid index
TN : True negative
TP : True positive

## INTRODUCTION

Hirschsprung disease (HSCR) also known as congenital megacolon is a condition caused by the absence of ganglion cells (aganglionosis) in the myenteric plexus and submucosa of rectum and colon [1]. Aganglionosis typically extends to the proximal of transitional zones with varying lengths. This leads to narrowing in the aganglionic segment with the transitional zone taking on a funnel-like shape, as well as dilation and hypertrophy in the proximal segment. Consequently, this condition can impede the peristaltic movement of the intestine, resulting in a clinical condition known as functional obstruction [2].

The worldwide incidence of HSCR is estimated to be around 1:5000 births and more in male than female with a ratio 4:1, HSCR also rarely occurs in premature babies [1,3]. In Indonesia, based on the data obtained from the Ministry of Health, HSCR incidence also estimated to be around 1:5000 live births [4]. It is still no published data of HSCR incidence in East Java, especially in Malang city every year. Dr. Saiful Anwar General Hospital Malang, which is one of the East Java referral center only has 2 published studies on HSCR. Study conducted by Inggarwati in 2010 showed that the total of HSCR cases in 2005-2006 is 18 [5]. Then in 2021, Wibowo conducted a research with a total of 8 HSCR cases in 2008 [6].

In general, there are three supporting examinations that can be performed to diagnose HSCR namely contrast enema of HSCR, anorectal manometry, and rectal biopsy [7]. The gold standard for enforcing HSCR is rectal biopsy confirm the absence of ganglion cells in submucosa through

histologic examination [1,8]. Rectal biopsy examination began in 1948, Whitehouse and Kernohan established the diagnosis of HSCR depending on the microscopic evaluation of the rectal biopsy. Initially, a biopsy was performed by excising the entire thickness of the rectal muscle wall to obtain an overview of the myenteric plexus. This procedure is relatively difficult to perform because it requires general anesthesia, it can cause inflammation and connective tissue formation which can complicates further definitive surgery. In addition, this technique can cause complications such as perforation, rectal bleeding, and infection. Later, rectal suction biopsy was introduced as a diagnostic method that can be performed without general anesthesia and with less potential for bleeding or infection [9].

Rectal biopsy and histologic examination can be challenging to perform in rural areas, even the sample of suction rectal biopsy are stable in formalin and can be sent to an adequate center, the delivery process requires time, a long distance, and relatively expensive costs. Still, an accurate diagnosis is essential to determine the appropriate management for HSCR. Contrast enema is a preferred radiological examination of HSCR where the biopsy is not yet available. According to our observation, rectal biopsy has only been available since March 2021 in Dr. Saiful Anwar General Hospital Malang. Contrast enema of HSCR were used before as the most important diagnostic tests. Given the frequency of HSCR cases, the effective utilization of diagnostic options, and the continued use of contrast enema of HSCR in peripheral hospitals and rural areas, this study aims to determine the sensitivity, specificity, and accuracy of contrast enema of HSCR compared to the findings of rectal biopsy which serve as the gold standard.

## MATERIALS AND METHODS

### Patient Sample

This study employed a cross-sectional study design. Research subjects consisted of all suspected HSCR patients aged 0-12 months who presented at Dr. Saiful Anwar General Hospital Malang from January to December 2021. Patients with complications such as Hirschsprung-associated enterocolitis (HAEC) or sepsis, congenital lethal anomalies, and those who had previously

undergone a stoma procedure were excluded from the study. The sampling technique used was total sampling method, namely all existing patients who met the criteria will become research subject. Patients would later classified into HSCR and non-HSCR based on the results of contrast enema of HSCR and suction rectal biopsy to calculate sensitivity, specificity, and accuracy.

**Data Collection**

The data were collected by direct interview with the families of patients. In each patient, heteroanamnesis, physical examination, and diagnostic procedures were conducted, including the contrast enema of HSCR and suction rectal biopsy. In this study, contrast enema performed without fasting and bowel preparation. First, folley catheter inserted into rectum with the tip inside the distal rectum, catheter balloon was not inflated. Second, contrast media was inserted passively using gravitation, followed with fluoroscopy in AP position, until transitional zone identified. Then lateral image of rectum and sigmoid was taken to identify rectosigmoid index. After that, wide field of view abdominal X-ray was taken. Lastly 48 hour photo was taken in AP supine position to check contrast retention. By the result of suction rectal biopsy, HSCR was determined with the absence of ganglion cells in submucosa on histologic examination. In order to reduce the likelihood of bias, the results of each diagnostic tool were read by the same individual.

**Data Analysis**

The collected data was processed first in the form of frequency tables. Data was subsequently analyze with independent t-test and chi-square to compare the study parameters between HSCR and non-HSCR. Furthermore, cross-tabulation was conducted between the two diagnostic tests. Sensitivity, specificity, and accuracy calculations were performed using a confusion matrix (Figure 1). Additionally, the odds ratio (OR) was calculated to indicate the likelihood that the established diagnosis is true for HSCR compared to non-PH cases.

**Ethic**

This research was conducted after obtaining research permission and ethical approval from the Research Ethics Commission of Dr. Saiful Anwar General Hospital Malang under the serial number 400/131/K.3/102.7/2022. The research subjects were represented by parents or legal family and they also provided their consent to participate in the study.

## RESULTS

### Demographic Characteristics

We analyzed up to 35 suspected HSCR patients who came to Dr. Saiful Anwar General Hospital from January to December 2021. There were included 27 male patients and 8 female patients. The average of age, birth weight, and birth length consecutively is 30.03 days, 3135.14 gram, and 48.54 cm (Table 1). Out of 35 suspected HSCR patients examined using contrast enema, a total of 30 patients showed signs of HSCR, while 5 patients were non-HSCR cases. Based on suction rectal biopsy, there were 29 HSCR patients and 6 non-HSCR patients. The characteristics such as sex, age, birth weight, and birth length had no significant p-values (Table 2). It can be concluded that there was no significant difference characteristic between HSCR and non-HSCR patients. On the other hand, based on the OR calculations (Table 2), male patients had 4.8 times higher risk of HSCR compared to female patients.

Several complaints experienced by patients include delayed meconium passage, enlarged stomach, vomiting, difficulty defecating, and recurrent bloating. The statistical calculations regarding patient complaints based on suction rectal biopsy as a gold standard, consist of vomiting (p=1.000), difficulty defecating (p=0.576), and recurrent bloating (p=0.079) (Table 2) leading to the conclusion that there was no significant difference between HSCR and non-HSCR patients. However, the enlarged stomach indicated a significant difference between HSCR and non-HSCR patients (p=0.003) (Table 2). Patients who present with vomiting complaints are at a 0.96 times higher risk of HSCR compared to patients without vomiting complaints. Additionally, patients experiencing difficulty defecating and recurrent bloating are consecutively at 2.4 and 6.25 times higher risk of HSCR (Table 2).

### *Sensitivity, Specificity, and Accuracy of Contrast Enema*

There were found 28 true positives, 1 false positive, 2 false negatives, and 4 true negatives (Table 4). It showed among 29 (82.86%) patients diagnosed HSCR with suction rectal biopsy results, there

were as many as 96.55% patients with the results of contrast enema also showing a picture as HSCR. Meanwhile, the 6 patients (17.14%) whose suction rectal biopsy results did not show HSCR, only 33.30% patients whose contrast enema showed HSCR.

Based on the confusion matrix formula (Figure 1) and the cross-tabulation results (Table 3), we obtained sensitivity, specificity, and accuracy of contrast enema of HSCR compared to suction rectal biopsy result were consecutively 93.3%, 80%, and 91.42%. The value explained that contrast enema of HSCR capable to show which patient suffer from HSCR among the patients who were truly HSCR by 93.3%. Contrast enema of HSCR also had an ability of 80% to show non-HSCR patients and it was true that these patients are non-HSCR based on suction rectal biopsy result. From the results, it was obtained that contrast enema of HSCR can accurately predict whether a patient had HSCR or not with a 91.42% accuracy.

Furthermore, a positive predictive value (PPV) of 96.55% was obtained. This means that contrast enema of HSCR was able to correctly predict HSCR with a rate of 96.55%. Additionally, the 66.66% negative predictive value (NPV) was indicating that contrast enema of HSCR can correctly predict non-HSCR. The OR of contrast enema of HSCR was 11 (95% Confident interval (CI): 1.064–113.731) (Table 4). If the result of contrast enema of HSCR indicated HSCR, the likelihood of the patient also being diagnosed with HSCR from biopsy result is 11 times greater compared to non-HSCR cases.

## DISCUSSION

Based on characteristics of patients with HSCR, the majority (constituting 80%) are male. This is consistent with other studies that also state that male predominates among the majority of HSCR patients [10-12]. Although there is no significant difference in sex distribution of HSCR and non-HSCR patients in this study, the value of OR suggests that the risk of developing HSCR is 3.2 times greater in male patients than female patients. As it is acknowledged that male are more commonly affected than female in the case of HSCR [13]. Out of the 35 suspected HSCR patients, the average age of baby at the first time examination is 30.03 days. The age distribution was more dominant within the 0-4 week range following birth (36.4%) [14]. This is supported by a study at

Ulin General Hospital in South Kalimantan, which stated that the average age of patients arriving at the hospital was 30 days (74.2%) [15]. No significant difference are found in the birth weight and birth length between HSCR and non-HSCR patients. It means that there is a lack of disparity observed in the birth weight and birth length among patients. This statement is supported by research conducted at Dr. Mohammad Hosein General Hospital in Palembang, which stated that Hirschsprung's Disease is not associated with a low percentage of neonatal birth weight [16].

A clinical indicator pointing towards HSCR is delayed meconium passage. Delayed meconium passage beyond the first 24 hours occurs in around 90% of HSCR cases [1]. This study reports that all patients experienced delayed meconium passage and exhibited a significance value of 0.001. Other story indicated that the proportion of HSCR patients with a history of delayed meconium passage at 48 hours is significantly higher than non-HSCR patients ($p<0.001$) [17]. Delay in the passage of meconium is the primary indication of HSCR in newborn. More than 90% of individuals are affected by this condition do not excrete meconium within the initial 24 hours of their life [18]. Patients usually present with delayed meconium passage (>24 hours), vomiting, abdominal distension, and constipation. All term babies who fail to pass meconium within 24–48 h of life and with abdominal distension should subjected to rectal biopsy. Constipation is a common childhood problem. Only a subset of children with constipation has significant problems, and of this subset has HSCR [19].

In this study, the majority of HSCR patients diagnosed with suction rectal biopsy experienced enlarged stomach with a significant p-value ($p=0.003$) compared to non-HSCR cases. Vomiting ($p=1.000$), difficulty defecating ($p=0.576$), and recurrent bloating ($p=0.079$) indicates there are no significant difference between HSCR and non-HSCR patients. In children over 1 month of age, the strongest factors associated with HSCR are male gender, developmental failure, abdominal distension, and vomiting [20]. Additionally, patients often have a history of recurrent bloating or experience severe chronic constipation since infancy [1].

This present study states that contrast enema of HSCR has 93.30% sensitivity, 80.00% specificity, and 91.24% accuracy. Additionally, PPV of 96.55% and NPV of 66.66% were also obtained. Considering the false negative value of 33.33% and the potential for harm by delayed or over

treatment, a suction rectal biopsy still needs to be carried out and the sample is sent using formalin because the delivery process takes time and distance. The patient can be treated with a saline laxative enema first. For the center that do not have equipment for biopsy, it is better to refer patients to the center with more adequate equipment.

The values obtained exhibit a sufficiently high magnitude to differentiate and diagnose HSCR and non-HSCR. A study conducted at Sanglah Hospital in Denpasar mentioned that barium enema had a sensitivity of 95.50%, specificity 87.50%, PPV 97.60%, and NPV 77.8% for diagnosing HSCR [21]. Radiological observations such as transition zone and irregular rectosigmoid index (RI) during contrast enema examination prove to be highly accurate for the diagnosis of HSCR with the reported contrasting values for sensitivity, specificity, accuracy, PPV, and NPV at 83.78%, 80.00%, 83.33%, 96.88%, and 40.00% respectively [22]. Research in Pakistan, with higher results reported that barium contrast enema had a sensitivity of 95.24%, specificity of 90.40%, PPV of 90.91%, NPV of 94.96%, and accuracy of 92.83% [23]. Another study reported the accuracy of barium enema as a diagnostic tool for HSCR to be around 72% [24].

Contrast enema of HSCR according to the theory is a radiographic examination of the colon using contrast media which aims to show the anatomy of the colon. It used preliminary anteroposterior (AP) photo projections and images with contrast media, as well as AP, lateral and AP post-evacuation projections [25]. It helps confirm the diagnosis or rule out a differential diagnosis of HSCR by determines the transition zone [26]. Meanwhile, rectal biopsy is the gold standard for diagnosing HSCR which is determined by the absence of ganglion cells in the distal intestine, submucosal/Meissner plexus, and intermuscular plexus/Auerbach [1]. Comprehensive categorization of irregularities in colonic mucosa detected through a contrast enema shows a high level of sensitivity and moderate specificity for HSCR [27]. Colon area examination will be more visible using contrast media. Other studies in the case of Hirschsprung's were not carried out with rectography examination, only using colon in-loop examination with several projections, namely abdominal plain AP, AP, and lateral post-contrast [28]. Barium contrast enema is considered effective for diagnosing lower gastrointestinal obstructions. It can be used for diagnosing HSCR when suction rectal biopsy equipment is not available [29,30]. Another study examined 24-hour

delayed barium enema film on suspected HSCR patients revealed that the significance of barium retention levels persists in predicting aganglionic bowel segments, playing a role in the decision-making process for surgical physicians. While the correlation rate may fluctuate based on the age and clinical type of HSCR patients, the retained barium level serves as a valuable predictor for the actual extent of disease involvement. Nevertheless, histopathology remains essential for precisely determining the aganglionic bowel segment in individuals with HSCR [31].

In this cross-sectional study, high values of sensitivity, specificity, and accuracy were obtained. This statement is supported by previous research findings and the obtained of OR. The value of OR for the diagnosis of contrast enema of HSCR is 11 (95% CI: 1,064 – 113,731) (OR> 1). It can be concluded that the diagnosis of contrast enema of HSCR can be one of the factors for predicting a HSCR diagnosis. Where if the results of contrast enema show HSCR, the probability of patient is correctly diagnosed with HSCR from the results of the suction rectal biopsy is 11 times greater than non-HSCR. This study has a limitation including a limited sample size as the research was only conducted at a single hospital center within one-year period.

## CONCLUSION

The results of contrast enema of HSCR performed on patients suspected of HSCR had a 93.3% sensitivity, 80.00% specificity, 91.4% accuracy, and obtained OR of 11.0 (95% CI: 1.064-113.731). It can be concluded that contrast enema of HSCR can be considered as a tool for diagnosing HSCR in rural areas where the suction rectal biopsy as a gold standard diagnosis and histologic examination services are not available.

## CONFLICT OF INTEREST

All authors undersign and certificate that we do not have any financial or personal relationships that might bias the content of this work.

## AUTHOR'S CONTRIBUTIONS

Conceptualization, Lulik Inggarwati.; methodology, Lulik Inggarwati., Anni Masrurotun Najiyah Ziha Ul Haq., Indrastuti Normahayu.; software, Anni Masrurotun Najiyah Ziha Ul Haq.; validation, Lulik Inggarwati., Indrastuti Normahayu., Solimun.; formal analysis, Lulik Inggarwati., Indrastuti Normahayu., Solimun.; investigation, Anni Masrurotun Najiyah Ziha Ul Haq.; resources, Lulik Inggarwati., Indrastuti Normahayu.; data curation, Anni Masrurotun Najiyah Ziha Ul Haq.; writing—original draft preparation, Lulik Inggarwati., Anni Masrurotun Najiyah Ziha Ul Haq.; writing—review and editing, Lulik Inggarwati., Indrastuti Normahayu.; visualization, Anni Masrurotun Najiyah Ziha Ul Haq.; supervision, Happy Kurnia Permatasari., Wisnu Barlianto., Respati Suryanto Dradjat.; project administration, Lulik Inggarwati., Anni Masrurotun Najiyah Ziha Ul Haq.; funding acquisition, Lulik Inggarwati., Anni Masrurotun Najiyah Ziha Ul Haq. All authors have read and agreed to the published version of the manuscript.

## ACKNOWLEDGEMENTS

None.

**REFERENCES**


1. Langer JC. Hirschsprung disease. In: Peter SDS, editor. *Holcomb and Ashcraft's Pediatric Surgery*. 7th ed. New York: Elsevier; 2020. pp. 557–76.
2. O'Donnell AM, Montedonico S, Puri P. Pathophysiology of Hirschsprung's Disease. In: Puri P, editor. *Hirschsprung's Disease and Allied Disorders*. 4th ed. Springer Cham; 2019. pp. 153–66.
3. Chen H. Hirschsprung Disease. [Internet]. Los Angeles: Society of American gastrointestinal and endoscopic surgeon; 2019 [Retrieved 2024 February 11]. Available at: https://www.sages.org/wiki/hirschsprung-disease-2/.
4. Ministry of Health. *Pedoman nasional pelayanan kedoteran tata laksana penyakit Hirschsprung*. Jakarta: Kementrian Kesehatan Republik Indonesia; 2017.
5. Inggarwati L, Triambodo B. Colon irrigation as colostomy replacement on Hirschsprung disease surgery. *J Kedokt Brawijaya*. 2010; 26(2): 117–119. https://doi.org/10.21776/ub.jkb.2010.026.02.10.
6. Wibowo H. Duhamel Procedure untuk Hirschsprung's Disease Anak di RS Syaiful Anwar Malang. *KELUWIH J Kesehat dan Kedokt*. 2021; 2(2): 91–94. https://doi.org/10.24123/kesdok.V2i2.2532.
7. Puri P. *Newborn Surgery*. 4th ed. London: CRC Press; 2017.
8. Coran AG, Adzick NS, Krummel TM, Laberge J-M, Shamberger R, Caldamone A. *Pediatric surgery*. 7th ed. New York: Mosby; 2012.
9. Ambartsumyan L, Smith C, Kapur RP. Diagnosis of Hirschsprung Disease. *Pediatr Dev Pathol*. 2020; 23(1): 8–22. https://doi.org/10.1177/1093526619892351
10. Gunadi, Karina SM, Dwihantoro A. Outcomes in patients with hirschsprung disease following definitive surgery. *BMC Res Notes*. 2018; 11(1): 1–5. https://doi.org/10.1186/s13104-018-3751-5.
11. Rizky M, Isa MM, Kamarlis RK. Comparison of barium enema and frozen section results in the diagnosis of hirschsprung's disease in a tertiary care hospital at Aceh, Indonesia. *Med J Malaysia*. 2020; 75 (Suppl 1): 37–40. https://www.e-mjm.org/2020/v75s1/barium-enema.pdf.

12. Balela N, Fauzi AR, Nugroho N, Dwihantoro A, Gunadi. Prognostic factors for persistent obstructive symptoms in patients with Hirschsprung disease following pull-through. *PLoS One*. 2023; 18 (9):1–9. http://dx.doi.org/10.1371/journal.pone.0290430.
13. Puri P, Nakamura H. Epidemiology and clinical characteristics of Hirschsprung's disease. In: Puri P, editor. *Hirschsprung's Disease and Allied Disorders*. 4th ed. Switzerland: Springer Cham; 2019. pp. 167–74.
14. Anandasari PPY, Tjan A, Widyasari NN. Radiology Perspective One-Year Study of Hirschsprung Disease. *Folia Medica Indones*. 2021; 57(1): 41-45. https://doi.org/10.20473/fmi.v57i1.9857.
15. Poerwosusanta EE, Hariastawa IA, Setiawan A, Sulistiawati S, Ary Wibowo A. Correlation Between the Initial-Surgery Age With the Hirschsprung'S Disease Patient Stooling Patterns in Ulin Hospital Banjarmasin, South Kalimantan, Indonesia in 2012-2015. *Maj Biomorfologi*. 2021; 31(2): 66-69. https://doi.org/10.20473/mbiom.v31i2.2021.66-69.
16. Aswadi T, Maritska Z. Risk Factors of Congenital Anomalies in South Sumatera Indonesia. *Biosci Med J Biomed Transl Res*. 2020; 4(4): 60–69. https://doi.org/10.32539/bsm.v4i4.165.
17. Khorana J, Phiromkanchanasak P, Kumsattra J, Klinoun S, Aksorn S, Chantakhow S, et al. Development of clinical referral score model for early diagnosis of hirschsprung's disease in suspected pediatric patients. *Healthc*. 2021; 9(6): 1-13. https://doi.org/10.3390/healthcare9060678.
18. Heuckeroth RO. Hirschsprung disease - Integrating basic science and clinical medicine to improve outcomes. *Nat Rev Gastroenterol Hepatol*. 2018; 15(3): 152–67. https://doi.org/10.1038/nrgastro.2017.149.
19. Redkar R, Chigicherla S, Tewari S, Sharma RD. Comparison between suction rectal biopsy and full-thickness rectal biopsy in the diagnosis of hirschsprung's disease. *J Indian Assoc Pediatr Surg*. 2021; 26(3): 144-147. 10.4103/jiaps.JIAPS_47_20.
20. Jaroy EG, Emblem R, Reims HM, Mai TT, Risa GT, Ougland R. Evaluation of diagnostic factors used to refer children with constipation for rectal biopsies. *Int J Colorectal Dis*. 2022;37(3):597–605. https://doi.org/10.1007/s00384-021-04069-4.

21. Darmajaya IM, Subhawa IK. Validity of Barium Enema as Hirschsprung's Disease Diagnostic Tools for Infant in Sanglah Hospital Denpasar. *JBN (Jurnal Bedah Nasional)*. 2021; 5(1): 16-21. https://doi.org/10.24843/JBN.2021.v05.i01.p03.

22. Irsan M, Gunadi, Dwihantoro A. Accuracy of Contrast Enema for Diagnosing Hirschsprung Disease in Dr. Sardjito Hospital Yogyakarta. [Internet]. Yogyakarta: Universitas Gadjah Mada; 2019 [Retrieved 2024 February 11]. Available at: https://etd.repository.ugm.ac.id/penelitian/detail/168809.

23. Anwar Z, Ahmed N, Rehman S, Afzal T, Ahmed I. Diagnostic accuracy of barium enema for the diagnosis of Hirschsprung disease in children. *Pakistan Armed Forcec Med J*. 2023; 73: 219–222. https://doi.org/10.51253/pafmj.v73i1.5818.

24. Yan J, Sun J, Wu R, Tan SS, Chen Y, Peng Y, et al. Barium enema findings in total colonic aganglionosis: a single-center, retrospective study. *BMC Pediatr*. 2020; 20(1): 1–8. https://doi.org/10.1186/s12887-020-02403-3.

25. Lampignano J, Kendrick LE. *Bontrager's Textbook of Radiographic Positioning and Related Anatomy*. 9th ed. London: Mosby; 2017.

26. Peyvasteh M, Askarpour S, Ostadian N, Moghimi M-R, Javaherizadeh H. Diagnostic accuracy pf barium enema findings in Hirschsprung's disease. *Arq Bras Cir Dig*. 2016; 29(3): 155–158. 10.1590/0102-6720201600030007.

27. Vlok SSC, Moore SW, Schubert PT, Pitcher RD. Accuracy of colonic mucosal patterns at contrast enema for diagnosis of Hirschsprung disease. *Pediatr Radiol*. 2020; 50(6): 810–816. https://doi.org/10.1007/s00247-020-04631-2.

28. Nasokha MIM, Anggresti, N. Charges Detection of Ultrashort Hirschsprung's Disease (Congenital) with Additional Rectography Examinations. *Nusantara Science and Technology Proceedings*. 2022; 59-62. https://doi.org/10.11594/nstp.2022.2909.

29. Frongia G, Günther P, Schenk JP, Strube K, Kessler M, Mehrabi A, et al. Contrast Enema for Hirschsprung Disease Investigation: Diagnostic Accuracy and Validity for Subsequent Diagnostic and Surgical Planning. *Eur J Pediatr Surg*. 2016; 26(2): 207–214. http://dx.doi.org/10.1055/s-0035-1546755.

30. Manzoor A, Talat N, Adnan HM, Zia MW, Aziz MA, Ahmed E. Contrast Enema: Solving Diagnostic Dilemmas in Neonates With Lower Intestinal Obstruction. *Cureus*. 2022; 14(3): 6–10. 10.7759/cureus.23458.

31. Zhou B, Wang D, Chen K, Niu Y, Jiao C, Zhu T, et al. The utility of the 24-h delayed film of barium enema for detecting the dysganglionic bowel segment in Hirschsprung's disease. *Front Pediatr*. 2022; 10: 1–9. 10.3389/fped.2022.979149.

## TABLES

**Table 1.** Demographic Characteristics

| Demographic | Frequency | Percentage (%) |
|---|---|---|
| Sex | | |
| Male | 27 | 77.14% |
| Female | 8 | 22.86% |
| Patient age (Mean±SD) | 30.03±65.10 | |
| Birth weight (gram) (Mean±SD) | 3135.14±332.18 | |
| Birth length (cm) (Mean±SD) | 48.54±2.13 | |
| Mother's age (Mean±SD) | 29.74±6.10 | |
| Gestational age | | |
| <37 week (Preterm) | 2 | 5.7% |
| 37-42 week (Normal) | 33 | 94.3% |
| >42 week (Postterm) | 0 | 0% |
| How to give birth | | |
| Normal | 26 | 74.29% |
| Sectio caesaria | 9 | 25.71% |
| Meconium | | |
| >24 hours | 35 | 100% |
| <24 hours | 0 | 0.0% |
| Rectal suction biopsy results | | |
| Hirschsprung | 29 | 82.86% |
| Non Hirschsprung | 6 | 17.14% |
| Contrast enema of HSCR results | | |
| Hirschsprung | 30 | 85.71% |
| Non Hirschsprung | 5 | 14.29% |
| Operation action | | |
| There isn't any | 24 | 68.57% |
| Duhamel Procedure | 4 | 11.43% |
| Sigmoidostomy | 4 | 11.43% |
| TEPT | 3 | 8.57% |

HSCR = Hirschsprung disease; TEPT = transanal endorectal pull-through

**Table 2.** Bivariate Analysis of Patient Characteristics Based on Contrast Enema of HSCR Result

| Characteristics | HSCR (n) | Non-HSCR (n) | P-Value | Odds Ratio |
|---|---|---|---|---|
| Sex | | | | |
| Male | 24 | 3 | 0.110[a] | 3.2 (CI 95%: 0.75 – 13.66) |
| Female | 6 | 2 | | |
| Age (Median) | 7 | 19 | 0.380[b] | |
| Birth weight (Mean) | 3123.48 | 3157.5 | 0.778[a] | |
| Birth length (Median) | 50 | 49.0 | 0.481[a] | |
| Meconium | | | | |
| >24 hours | 30 | 5 | 0.001[a] | |
| <24 hours | 0 | 0 | | |
| Complaints of enlarged stomach | | | | |
| Yes | 30 | 2 | 0.044[a] | |
| No | 0 | 3 | | |
| Complaints of vomiting | | | | |
| Yes | 24 | 5 | 0.000[a] | 23.75 (CI 95%: 3.69 – 152.89) |
| No | 6 | 0 | | |
| Complaints of difficulty defecating | | | | |
| Yes | 24 | 4 | 0.002[a] | 10.8 (CI 95%: 2.09-55.67) |
| No | 6 | 1 | | |
| Complaints of recurrent bloating | | | | |
| Yes | 25 | 3 | 0.002[a] | |
| No | 5 | 2 | | |

HSCR = Hirschsprung disease; CI = confident interval; (a) Analysis with chi-square test (b) Analysis with independent t-test

**Table 3.** Cross-tabulation on Contrast Enema of HSCR and Suction Rectal Biopsy

| | Suction Rectal Biopsy | | |
|---|---|---|---|
| | **HSCR** | **Non-HSCR** | **Total** |

| | | | | | |
|---|---|---|---|---|---|
| Contrast enema of HSCR disease | HSCR | Count | 28[a] | 2[b] | 30 |
| | | % within output of biopsy | 96,55% | 33,33% | 85,71% |
| | Non-HSCR | Count | 1[c] | 4[d] | 5 |
| | | % within output of biopsy | 3,45% | 66,66% | 14,29% |
| Total | | Count | 29 | 6 | 35 |
| | | % within output of biopsy | 100.0% | 100.0% | 100.0% |

HSCR= Hirschsprung disease; (A) true positive; (B) false negative; (C) false positive; (D) true negative

**Tabel 4.** Risk Estimation on Contrast Enema of HSCR and Suction Rectal Biopsy

| | Value | 95% Confident Interval | |
|---|---|---|---|
| | | Lower | Upper |
| OR Colon in Loop Result HSCR/ Non-HSCR | 11[a] | 1.064[b] | 113.731[b] |
| Cohort Biopsy Result HSCR | 1.435 | 0.953 | 2.161 |

| | | | |
|---|---|---|---|
| Cohort Biopsy Result non-HSCR | 0.130 | 0.016 | 1.041 |
| Valid Cases | 35 | | |

HSCR = Hirschsprung disease

**FIGURES**

| | | Suction Rectal Biopsy (Gold Standard) | | |
|---|---|---|---|---|
| | | Positive | Negative | |
| Contrast Enema of Hirschsprung Disease | Positive | True Positive (TP) | False Negative (FN) | **Sensitivity** TP/(TP+FN) |
| | Negative | False Positive (FP) | True Negative (TN) | **Specificity** TN/(TN+FP) |
| | | **Positive Predictive Value (PPV)** TP/(TP+FP) | **Negative Predictive Value (NPV)** TN/(TN+FN) | **Accuracy** (TP+TN)/(TP+TN+FP+FN) |

**Figure 1.** Confusion Matrix