INTRODUCTION

Hirschsprung’s disease is a developmental disorder of the enteric nervous system and is characterized by the absence of ganglion cells in the myenteric plexus and submucosa of the distal intestine. These cells are responsible for intestinal peristalsis. The absence of enteric ganglion cells in the myenteric plexus and submucosa in various segments of the GI tract will cause sustained contraction of the aganglionic bowel segments, leading to intestinal obstruction and distention of its proximal segment (megacolon). Clinical features vary according to age at presentation and the extent of the aganglionic segment. Patients generally present with delayed passage of meconium (>48 hours), feeding intolerance, abdominal distention, and bilious vomiting, symptoms consistent with intestinal obstruction. The worldwide incidence of Hirschsprung’s disease is 1 in 5000 live births, ranging from 1 in 14000 to 1 in 4000 live births. In Indonesia, the incidence of Hirschsprung’s disease is still uncertain, but it is around one in 5000 live births. With a population of 220 million in Indonesia and a birth rate of 35 per million, it is predicted that every year, 1540 babies will be born with Hirschsprung’s disease. Hirschsprung’s disease was conducted in 2017 for 1 year, where an incidence of Hirschsprung’s disease was found to be 1 in 49 live births.

The gold standard for diagnosing Hirschsprung’s disease is a rectal biopsy. Analysis of the biopsy specimens was performed to look for the absence of hypertrophic ganglion cells and nerve trunks. Various methods can make an early diagnosis of Hirschsprung’s disease. However, the first diagnostic procedure often performed is a barium enema or colon in-loop examination. Findings suggestive of Hirschsprung’s disease include the presence of a radiographic transitional zone (TZ) with proximal bowel dilatation.

ABSTRACT

Background: Hirschsprung’s disease is a developmental disorder of the enteric nervous system of the distal intestines. Colon in-loop examination is used as a spearhead in the diagnosis of Hirschsprung’s disease, and according to previous studies, it was found that the presence of transitional zone and rectosigmoid index of less than 1 were correlated with positive pathology results in 79%-87% of cases. The purpose of this study was to provide an overview of the rectosigmoid index of patients suspected of having Hirschsprung’s disease as illustrated by a colon in-loop examination.

Methods: The research method is cross-sectional descriptive from January 2018 to December 2019 at Prof. Dr. dr. I. G. N. G. Ngoerah Hospital Denpasar. Data collected from colon in-loop examination in patients suspected of having Hirschsprung’s disease included gender, age, RSI, and pathology results in patients who underwent biopsy. Data were analyzed using SPSS version 20 for Windows.

Results: The results found from 49 samples were that Hirschsprung was dominant in males (63.27%). Patients aged less than 1 month were the most common, with a percentage of 44.9%. The second most common was at the age of 1 to 12 months at 30.6%, while the incidence rate with the least number was at the age of more than 12 months, namely 24.5%. RSI data found that of the patients suspected of having Hirschsprung’s disease, 40 samples, or 81.6%, had an RSI < 1, and it was found that of the 40 samples with RSI < 1, 82.9% confirmed Hirschsprung’s with an aganglionic pathology result. In contrast, in patients with aganglionic PA, the proportion of patients with RSI < 1 was more than 80%.

Conclusion: The data obtained in this study are in accordance with those described in previous studies and support the role of RSI in strengthening the diagnosis of Hirschsprung’s disease.

Keywords: barium enema; colon in loop; Hirschsprung’s disease; pediatric; radiology; rectosigmoid index.

prof. Dr. dr. I. G. N. G. Ngoerah General Hospital Denpasar is a regional referral hospital that accepts many pediatric surgical cases, one of which is Hirschsprung’s disease. The large number of patients suspected of having Hirschsprung’s disease in Prof. Dr. dr. I. G. N. G. Ngoerah General Hospital has encouraged researchers to examine other new signs that are less invasive as a diagnosis of Hirschsprung’s disease besides biopsy. Departing from this background, the researchers chose to carry out a cross-sectional descriptive study titled regarding characteristics of Rectosigmoid Index of Pediatric Patients with Definitive Hirschsprung’s Disease in Radiology Installation of Prof. Dr. dr. I. G. N. G. Ngoerah General Hospital Denpasar from January 2018 to December 2019.

METHODS

This study uses a cross-sectional descriptive research method at the Radiology Department of Prof. Dr. dr. I. G. N. G. Ngoerah General Hospital Denpasar involved all patients suspected of Hirschsprung’s disease who underwent colon in-loop examinations from January 2018 to December 2019. The patients underwent colon in-loop examinations using barium or soluble contrast water. For barium, it was diluted with a ratio of 1:8 using 0.9% NaCl; water-soluble contrast was diluted with a ratio of 1:3 using 0.9% NaCl. A marker is fixed on the anal dimple and approximately 50-100 cc of contrast is inserted into the anus using a Foley catheter without inflating the balloon. After the contrast was administered, X-rays were taken with anterior-posterior, lateral, and oblique projections. After that, demographic data such as age and gender, RSI data, and anatomic pathology results data were collected. This study categorizes age into 3 groups: age less than 1 month, age 1 to 12 months, and age over 12 months. Gender is divided into 2 groups, namely men and women. RSI data is divided into RSI <1 and RSI >1, and anatomic pathology results are categorized into aganglionic and ganglionic. Data were analyzed using SPSS version 20 for Windows.

RESULTS

Colon in loop contrast examination has been carried out on 49 children suspected of having Hirschsprung’s disease at Prof. Dr. dr. I. G. N. G. Ngoerah General Hospital from August 2018 to December 2019. From a total of 49 patients, there were 31 male patients (63.27%) and 18 female patients (36.73%). Based on the age group, the incidence of Hirschsprung’s disease was the highest in patients under 1 month, with 22 patients (44.9%). At the age of 1 to 12 months, 15 patients (30.6%) were found, while the lowest number of cases was at the age of > 12 months, namely 12 patients (24.5%). Based on the results of the entire colon in loop examinations of patients with suspected Hirschsprung’s disease, 40 patients (81.6%) were found with a rectosigmoid index (RSI) < 1, and 9 patients (18.4%) with RSI > 1. Of all the anatomic pathology examinations in patients with Hirschsprung’s disease, it was found that there are 41 patients (83.7%) with aganglionic segments, while there are 8 patients (16.3%) with ganglionic segments. Detailed subject characteristics showed in Table 1. Based on anatomical pathology results with aganglionic segments, RSI <1 was found in 34 patients (82.9%) and RSI > 1 in 7 patients (17.1%), as seen in Table 2. Based on anatomical pathology results with ganglionic segments, RSI <1 was found in 6 patients (75%) and RSI > 1 in 2 patients (25%), as seen in Table 3.

From the total examination data of patients with Rectosigmoid Index below 1 (RSI <1), a total of 40 patients were found, with aganglionic segments found in 34 patients (85%) and ganglionic segments found in 6 patients (15%) as shown in Table 4.

DISCUSSION

Hirschsprung’s disease is a congenital disease characterized by the absence of Auerbach’s and Meissner’s plexus in the colon. The most common predilection is at the rectosigmoid, found in as many as 90% but can also affect the entire colon and all parts of the intestine (Total Colonic Aganglionosis).15-17

A study conducted at Chittagong Hospital Bangladesh by Rahman Z et al., found that the incidence of Hirschsprung’s
disease in 2005 to 2009 was higher in boys, with 122 out of 181 cases, compared to girls with a ratio of 2.08:1.\(^\text{15}\) Research regarding the same subject was also carried out by Izadi M et al. at Poursina Hospital Iran in 2009, where 67% of Hirschsprung's disease patients were boys with a ratio of 2:1.\(^\text{16}\) A study by Henna N et al. researching the incidence of Hirschsprung's disease in Pakistan on March 2009 to October 2009 revealed a higher male incidence, with 41 patients out of 51 cases and a ratio of 4:1.\(^\text{17}\)

Furthermore, a similar study conducted in the Indian region in 2012 by Abbas M et al. revealed higher incidence of males more than females with a ratio of 3.28:1.\(^\text{18}\) In 2014 research by Ishaq M at Nishtar Hospital and Ibn-e-Siena, Multan found a higher incidence of Hirschsprung's disease in males than in females with a ratio of 5:1.\(^\text{19}\) In Indonesia, Kartono in 2004 at Cipto Mangunkusumo Hospital also found more male patients than females with a ratio of 3:1.\(^\text{20}\) A similar distribution of data was also found in this study, where the number of male patients suspected of having Hirschsprung's disease was higher than that of female patients, namely 31 male patients (63.27%) compared to 18 female patients (36.73%). The ratio found in our study is 1.72:1.

Rahman Z et al. found that the distribution of Hirschsprung's disease was more common in patients within 1 day to 3 years of age.\(^\text{21}\) In Indonesia, Kartono's study in 2004 revealed that Hirschsprung's disease patients are more common at 0-1 months.\(^\text{22}\) Furthermore, research conducted at Prof. Dr. R. D. Kandaou Hospital Manado from January 2010 to September 2014 also revealed similar results with the dominance of patients ages 0-1 month.\(^\text{23}\) Likewise, in this study, similar results were obtained where the group of patients within the age range of 0-1 month was the largest number of the samples, 22 patients out of 49 (44.9%).\(^\text{24}\)

Definitive diagnosis requires a histopathologic examination of an intestine wall biopsy that shows the absence of enteric ganglion cells in that particular portion of the intestine. Patients suspected of having Hirschsprung's disease should undergo a diagnostic evaluation first. The two screening tests for the diagnostic workup of Hirschsprung's disease are contrasted enema or colon in the loop and anorectal manometry. Patients with findings consistent with Hirschsprung's will undergo a biopsy to confirm the diagnosis. This study obtained anatomic pathology results with aganglionic segments in 41 (83.7%) of 49 patients suspected of having Hirschsprung's disease.\(^\text{25,26}\)

RSI <1 is one of the radiological signs that suggest the presence of Hirschsprung's disease.\(^\text{25}\) To calculate the rectosigmoid index, the largest rectal diameter on a barium enema examination is obtained at any level below the third sacral vertebra. Meanwhile, the sigmoid colon is measured at 3 points, namely at the proximal, loop apex, and distal sigmoid colon, and the largest diameter is determined as the sigmoid diameter. All measurements were obtained along the transverse axis to the longitudinal axis of the colon at that point.\(^\text{27}\) In this study, there were 9 patients (18.4%) with RSI >1 and 40 patients (81.6%) with RSI <1. In addition, in this study, the results of anatomic pathology with aganglionic and ganglionic segments were also differentiated based on the rectosigmoid index, which in patients with aganglionic segments, 34 patients (82.9%) had RSI <1 and 7 patients (17.1%) had RSI >1. Whereas in patients with ganglionic segments, 6 patients (75%) had RSI <1, and 2 patients (25%) had RSI >1.

Next, out of 40 patients with RSI < 1.34, patients (85%) had aganglionic segments, and 6 patients (15%) had ganglionic segments. This finding is in accordance with the theory and findings of previous studies.

**CONCLUSION**

The number of cases of Hirschsprung's disease patients from January 2018 to December 2019 was 49. The proportion of boys suspected of having Hirschsprung's disease is higher than girls, with a ratio of 1.72:1. Based on age, there were more Hirschsprung's disease patients examined at neonatal age. Generally, the initial examination is a radiological colon in loop. The definitive diagnosis is made using an anatomic pathological examination of the intestinal wall biopsy. This study found that in accordance with the theory, in patients suspected of having Hirschsprung's disease, the proportion of RSI <1 was higher than RSI >1. In patients with RSI <1, it was found that the proportion with aganglionic segment results was higher than ganglionic segment results, and vice versa. In patients with aganglionic segment, the proportion of patients with RSI <1 was greater than 80%.

**CONFLICT OF INTEREST**

The authors declare no conflict of interest regarding the publication of the research report.

**ETHICAL CONSIDERATIONS**

The ethics committee of Udayana University approved this study with Ethical Clearance Register Number 2173/UN14.2.2.VII.14/LT/2022.

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AUTHOR CONTRIBUTIONS
All authors have made the same contribution in the preparation of this research, from preparing a conceptual framework, data collection, and data analysis to preparing a publication manuscript in the form of a research report.

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