

A rare case of porokeratosis mibelli in 3-year-old boy



Harry Galuh Nugraha^{1*}, Pramesti Indri Miranti¹

ABSTRACT

Background: Hirschsprung's disease is a congenital gastrointestinal disorder that is the most common cause of intestinal obstruction in children. One of the factors that influence gastrointestinal disorders is the sacrum bone. The sacral index can review the evaluation of the sacral bone, an instrument that can evaluate the development of the sacral bone in different clinical conditions. This study assessed the value of the sacral ratio in children with Hirschsprung's disease and non-Hirschsprung's disease at Dr. Hospital. Hasan Sadikin Bandung.

Methods: This study was a descriptive-analytic study with a case-control design. Sampling was done retrospectively by collecting medical record data and the enema barium readings from October 2019 to July 2020. The study was conducted at the Department of Radiology Hospital Dr. Hasan Sadikin Bandung from September to October 2020. Data processing uses an unpaired T-test.

Results: The total sample was 58 objects, consisting of 29 with Hirschsprung's disease and 29 non-Hirschsprung, with an average age of Hirschsprung's disease was 3.14 ± 3.905 years. In the Hirschsprung's disease group, the mean sacral ratio value was 0,62 and, and the mean sacral ratio in the non-Hirschsprung group was 0,74. Statistical test results with unpaired T-test showed significant mean differences in the case and control groups ($p=0.0001$).

Conclusion: The sacral ratio value in children with Hirschsprung's disease compared to non-Hirschsprung were different significantly.

Keywords: Hirschsprung disease, Sacral ratio, Pelvic Rontgen.

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¹Department of Radiology, Universitas Padjadjaran, Dr. Hasan Sadikin General Hospital, Bandung

*Corresponding to:
Harry Galuh Nugraha. Department of Radiology, Universitas Padjadjaran, Dr. Hasan Sadikin General Hospital, Bandung. Address:
hg.nugraha@gmail.com

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INTRODUCTION

Hirschsprung's disease is one of the most common causes of intestinal obstruction in children, among other congenital disorders of the gastrointestinal tract. One of the leading sign and symptoms of Hirschsprung's disease is constipation, usually appears a moment after birth. Older children also have prominent clinical symptoms is chronic constipation. The sacrum bone is one of the structures that most often affects gastrointestinal disorders. Abnormalities in the formation of the sacrum bones directly correlate with intestinal function, leading to pathological conditions such as constipation. The sacral ratio can be considered a method to evaluate the development of the sacrum in various clinical conditions, one of which is constipation.^{1,2}

Child constipation is defined as the decrease in the frequency of deposition (less than two per week) and the presence

of hard consistency stool that can provoke pain. It is considered that four entities are the leading causes of chronic constipation in children: idiopathic chronic constipation, Hirschsprung's disease, anorectal malformation and chronic intestinal pseudo-obstruction syndrome.^{2,3}

The sacral index was described over a decade ago and is considered a helpful tool for evaluating sacral development in different clinical conditions, including anorectal malformations. Partial or complete sacral agenesis belongs to sacral bone disorders. Sacral agenesis can lead to urinary and fecal disorders such as incontinence, repetitive urinary infections, urinary reflux, and constipation.³⁻⁵ Despite this, no studies have been performed in the Indonesian population that evaluate this index's relationship in patients with Hirschsprung's disease, which would serve as a reference point. Regarding the

importance of this issue, we assessed the sacral ratio between chronic constipation children in Hirschsprung's disease and normal children. We also determined whether sacral dysplasia is more common in children with Hirschsprung's disease.

METHOD

Study Design

This study was a case-control study on 58 children. The study group included 29 cases with Hirschsprung's disease, and the control group consisted of 29 children with non-Hirschsprung. The study was performed from October 2019 to July 2020 at the radiology department of DR. Hasan Sadikin Hospital Bandung.

Case and Control criteria

We included children between the ages of 0 and 18 who performed pelvic radiography with findings of Hirschsprung's disease into case group; and a group of patients

without this disease, which had pelvic radiography in frontal projections into the control group. We excluded children with any structural anomaly in the defecation system, such as anorectal malformation and cloaca exstrophy. Also, children with neurologic problems (spina bifida, myelomeningocele, meningocele or history of spinal cord or spine surgery) were excluded.

Procedure and Analysis

Measuring the sacral index was done in 58 patients who were part of the sample taken between October 2019 and July 2020. Estimation of the sacral index of each of the radiographs taken was done independently. The variables age, sex and sacral index value were taken into account. The following procedure was carried out: The research subjects were obtained from medical record data and Digital Imaging and Communication in Medicine (DICOM) data who did a barium examination with the results of Hirschsprung's disease, AP pelvic radiography was performed on non-Hirschsprung patients who were included in the criteria and measurement of sacral

ratio on AP pelvic radiographs was performed.

The sacral index was measured by the following method (as shown in **Figure 1**):

1. We are drawing a line that joins the highest points of the iliac crests (line A).
2. Then draw the second line by joining the lowest points of the posterior and inferior iliac spines (line B).
3. The third line runs parallel to the two previous ones and touches the lowest visible point of the coccyx (line C).

The sacral index resulted from the ratio distance between the two lowest lines (distance BC) over the two superior lines (distance AB). It can be measured both in the frontal projection (anteroposterior/AP) and the lateral projection.^{2,3,6}

The data was then calculated with statistical analysis using the SPSS software vers. 23. The sacral ratio was tested with a normality test to define the parametric or non-parametric test. We used an independent T-test if the data were distributed normally and a Mann-Whitney test for non-parametric. P-value <0.05 was considered significant.

RESULT

Of the 58 patients, 38 were male (65,5%), and 20 were female (34,5%). The research subjects were between ≤ 18 years old with pelvic radiographic and frontal projections classified according to this investigation's objective study group. The average age of Hirschsprung's disease was 3.14 ± 3.905 years. The average age of non-Hirschsprung's disease was 7.78 ± 5.594 years. Out of the total, 29 patients (50%) corresponded to the control group; 29 patients (50%) to the case group. All of the samples had frontal projection.

The sacral ratio for both groups was distributed normally. The sacral ratio mean in the non-Hirschsprung group was $0,74 \pm 0,058$ (0,62 - 0,83). The sacral ratio mean in the Hirschsprung group was $0,62 \pm 0,122$ (0,45 - 0,88). Unpaired T-test showed a statistically significant difference in the sacral ratio value between case and control ($p = 0.0001$) (**Table 1**).

DISCUSSION

The sacrum and spinal medulla are the bony structures that are most frequently affected in children with urinary, anorectal and intestinal pathology. The degree to which alterations of the sacrum are present is directly related to the child's functional prediction. Because of this, Pena developed the sacral index as standardised and objective measurement that tries to establish the presence and severity of sacral dysplasia, with a normality standard parameter of 0,74 in the frontal projection and 0,77 on the lateral projection.^{2-4,7}

Several studies have been made to evaluate the measurement in different clinical conditions. Caicedo et al. reported that the sacral index in patients with vesicoureteral reflux and constipation did not show significant variation compared to the values obtained in the normal pediatric population. However, the sacral ratio values were below the average in anorectal malformations, especially in high subtypes. Kajbafzade et al. reported the sacral index in healthy patients, patients with recurring infections of the urinary system and patients with fecal incontinuity. They found that the average value was higher in the healthy group (0,71) than in those with urinary

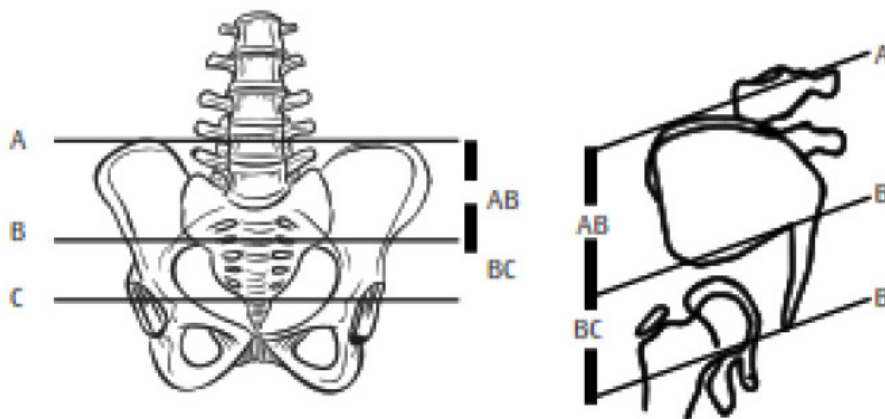


Figure 1. The measuring of the sacral index.³

Table 1. Comparison sacral ratio value between group Hirschsprung and non-Hirschsprung

Variable	Group		P-Value
	Non-Hirschsprung N=29	Hirschsprung N=29	
Sacral ratio value			0.0001
Mean±Std	0.74±0.058	0.62±0.122	
Median	0.74	0.62	
Range (min-max)	0.62-0.83	0.45-0.88	

affection (0,55) and constipation (0,491). Torre et al. studied the sacral development in anorectal malformations and normal populations. They observed complete bowel continence in 48% of children with normal sacral ratio, while only 17% of cases with low sacral ratio showed bowel continence.^{3,4,8,9}

In the normal population mean sacral ratio was 0,74, similar to those reported in the literature. Based on the comparison of the sacral ratio value between the non-Hirschsprung and Hirschsprung groups, the average sacral ratio value in the non-Hirschsprung group is higher than the average sacral ratio value in the Hirschsprung group. This result indicates a significant or statistically significant difference in mean. However, the average sacral ratio value in the Hirschsprung group was still greater than the pathological sacral ratio value, which was greater than 0.52. We have not found any data in the literature that could explain sacral ratio value in patients with Hirschsprung's disease.

The study is a case-control study that only compares SR values in children with Hirschsprung and non-Hirschsprung patients but does not look at the etiology of the difference in the mean SR values of Hirschsprung patients compared to non-Hirschsprung patients.

CONCLUSION

The non-Hirschsprung group has a higher sacral ratio value than Hirschsprung

group. We found that the sacral ratio value between both groups was significantly different. The sacral ratio can be considered as a reliable index to evaluate Hirschsprung's disease in children. Therefore, further research is needed to see the value of the sacral ratio to the duration of constipation and the mild severity of Hirschsprung's disease.

DISCLOSURE

Conflict of Interest

The authors have no potential conflicts of interest to disclose.

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Ethics Consideration

The Health Ethics Commission of Dr. Hasan Sadikin General Hospital has approved this study's ethical feasibility, which was appropriated according to ethics requirements with letter number LB.02.01/X.6.5/253/2020.

Author Contribution

All authors contributed equally regarding conducting the study and preparing the manuscript for publication.

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