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Use of the Recto-Sigmoid Index to Diagnose Hirschsprung's Disease

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Summary: The recto-sigmoid index on barium enema may aid in the diagnosis of Hirschsprung's disease. However, data on its reliability in different age groups are sparse. The recto-sigmoid index and transitional zone were evaluated blindly in 107 patients with diagnostic rectal suction biopsies. Patients were divided into 3 groups: neonates, infants older than 1 month, and children. The recto-sigmoid index and transitional zone agreed with the histopathologic diagnosis in 79% and 87% of the cases, respectively. Their negative

predictive values reached clinical significance in infants and children but not in neonates. Their positive predictive values were not significant in any age group. The recto-sigmoid index identified 4 patients with recto-sigmoid Hirschsprung's disease whose diagnosis was missed by evaluating the transitional zone alone. *Clin Pediatr.* 2006;00:1-5

Keywords: Hirschsprung's disease; aganglionosis; recto-sigmoid index; barium enema

Hirschsprung's disease (HD) is a congenital disorder of the colon that may present as intestinal obstruction in infants or chronic constipation in children. This condition is characterized by the absence of ganglion cells in the submucosal and myenteric plexus in the intestine.^{1,2} The embryogenesis is thought to be an arrest of the craniocaudal migration of neuroblasts into the intestinal wall. The aganglionic distal segment is hypertonic and does not transmit normal peristaltic contractions.³

In infants and children with constipation, anorectal manometry and radiologic studies have been used to establish the diagnosis of HD.⁴ However, the gold standard for the definitive diagnosis of aganglionosis is the rectal biopsy.

Despite the common use of the barium enema without colonic preparation as a diagnostic test for HD, there is some controversy concerning its use. The reported sensitivity, in the general pediatric population, of the barium enema in HD is 80% to 88%, with a specificity of 76% to 98%.^{4,6}

In 1975, Pochaczewsky and Leonidas published an innovative article concerning the use of the anteroposterior and lateral view of the barium enema to diagnose distal HD. They calculated a recto-sigmoid index as the widest diameter of the rectum divided by the widest diameter of the sigmoid colon (Figure 1).⁷

Initially, the recto-sigmoid index was used for the diagnosis of HD in the first few weeks of life. Some investigators suggest that it can also be used in older infants and children to aid in diagnosis.^{4,8} Other investigators found that a transitional zone was clearly visible in all cases in which the index was positive,^{9,10} suggesting that measuring the index may be superfluous. No systematic study comparing the radiologic sign of a transitional zone with the determination of the recto-sigmoid index on X ray in different pediatric age groups has been published.

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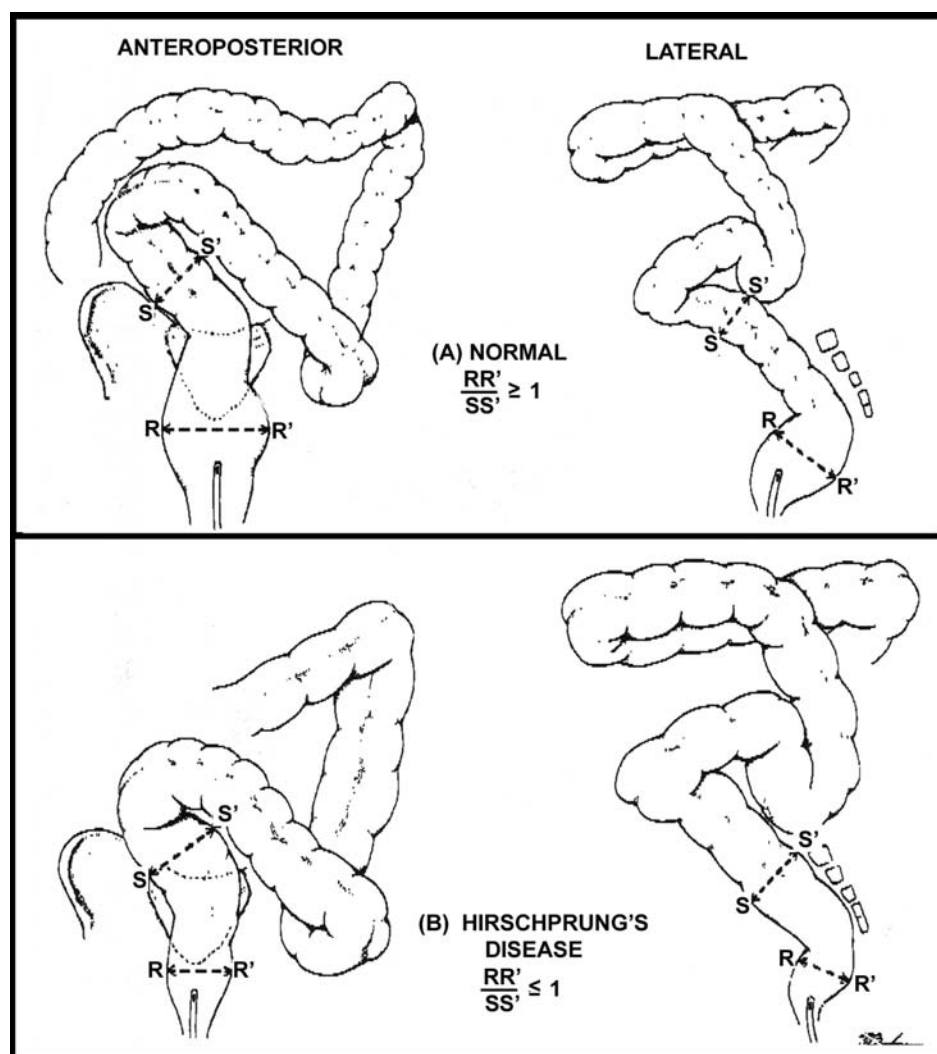


Figure 1. A, Normal recto-sigmoid index. The ratio between the widest diameter of the rectum (RR') and the widest diameter of the sigmoid (SS') is larger or equal to 1. B, Abnormal recto-sigmoid index: Hirschsprung's disease. The ratio is smaller than 1. Adapted with permission from Pochaczewsky R, Leonidas JC. The "recto-sigmoid index": a measurement for the early diagnosis of Hirschsprung's disease. *Am J Roentgenol Radium Ther Nucl Med.* 1975;123:770-777.

The purpose of our study was to evaluate the reliability of the recto-sigmoid index compared with the transitional zone in the diagnosis of HD in 3 age groups.

Materials and Methods

Unprepared barium enemas on patients who had rectal suction biopsies at Children's Hospital of New Orleans, Louisiana, from January 1999 to June 2004 were evaluated. We included only those with an adequate rectal suction biopsy or full-thickness biopsy. Only biopsies in which the submucosa was

represented were accepted as adequate. Patients with ganglion cells on the biopsy were considered to be negative for HD. Those with both absent ganglion cells and the presence of acetylcholinesterase-positive nerve fibers were considered positive for HD.^{11,12} In all these cases, the diagnosis was confirmed with a second biopsy or by intraoperative biopsies. Patients were divided into 3 age groups: neonates (first month of life), infants (1-12 months of age) and children (older than 1 year).

All the barium enemas were reviewed retrospectively and independently by 3 experienced pediatric radiologists. The radiologists were unaware of the pathology report and other clinical information. In

Table 1. Study Population Analysis and Distribution

	Neonates	Infants	Children	Total
Number	15	29	63	107
Age (range)	0.04 (1-29 d)	0.38 (35 d-10 mo)	5.26 (1-19 y)	(1 d-19 y)
male:female	3:2	1.2:1	5:4	5:4
HD (%)	6 (40)	5 (17.2)	5 (7.9)	16 (15)
male:female HD	2:1	4:1	4:1	3:1
Recto-sigmoid HD	4/6	5/5	4/5	13/16

HD = Hirschsprung's disease.

each barium enema, the recto-sigmoid index was calculated, and the presence or absence of a transitional zone was evaluated.

To calculate the recto-sigmoid index, the widest diameter of the rectum on the barium enema was obtained at any level below the third sacral vertebra. The sigmoid loop was measured at 3 points along its course (proximal, peak of the loop, and distal sigmoid colon). The largest measurement was selected as the sigmoid diameter. All measurements were obtained along a transverse axis, vertical to the longitudinal axis of the colon at that point.⁷

When the majority of the radiologists (at least 2 of 3) agreed with a recto-sigmoid index equal to or less than 1, the test was considered suggestive of HD (sigmoid colon wider than rectum). If the radiologists agreed that the recto-sigmoid index was greater than 1, the test was interpreted as normal (rectum wider than sigmoid colon) (Figure 1).

The subjective finding of a relative narrowed aganglionic segment distal to a dilated normal colon on an unprepared barium enema was considered a true transitional zone. The presence of the transitional zone was recorded if at least 2 of the 3 radiologists agreed in the interpretation.

Agreement between radiologic and pathologic findings was used to estimate the efficacy of the recto-sigmoid index and the transitional zone to diagnose HD. The sensitivity, specificity, positive predictive value, and negative predictive value of the recto-sigmoid index and the transitional zone were calculated in each age group using the exact binomial test, and 95 exact confidence intervals (CIs) were estimated using the SAS 9.1 (SAS Institute, Cary, NC). Bonferroni adjustment for multiple testing was done considering 8 test-confidence intervals with an overall significance of .05. These indicators were considered clinically significant if the lower limit of the corresponding confidence interval was greater than 0.5.

Results

A total of 107 patients (47 females and 60 males) with rectal suction biopsies and barium enemas were evaluated (Table 1). They ranged in age from 1 day to 19 years at the time of the barium enema, with a mean age of 3 years and a male:female ratio of 5:4.

Sixteen patients (15%) had HD. They were younger (mean, 1 year of age; range, 3 days-4 years) than the total patient population, with a male:female ratio of 3:1. Hirschsprung's disease was more frequently diagnosed in the neonatal group compared with the older patients (Table 1).

Of the 107 barium enemas, the recto-sigmoid index agreed with the biopsy report (true positives + true negatives) in 85 cases (79%; CI, 0.70-0.88). For the transitional zone, the radiologists agreed with the pathology report (true positives + true negatives) in 93 cases (87%; CI, 0.79-0.95). Sensitivity, specificity, and negative predictive values were clinically significant for both recto-sigmoid index and transitional zone in the total patient population (Table 2). However, differences among the 3 age groups were noted: In neonates, these values generally did not reach clinical significance. In contrast, in infants and children, specificity and negative predictive values of both recto-sigmoid index and transitional zone were clinically significant. Positive predictive values were not clinically significant in any age group.

Among the 16 patients diagnosed with HD, 3 had long-segment aganglionosis (Figure 2). In these 3 patients, the recto-sigmoid index was negative (>1). However, a transitional zone was detected in 2 of them (1 in the transverse colon and 1 at the splenic flexure). The diagnosis was missed in the third patient (total colonic aganglionosis).

Among the remaining 13 patients with recto-sigmoid HD, the recto-sigmoid index was positive (≤ 1) in 12 (92%; CI, 0.60-1.00; Figure 2). In contrast, a

Table 2. Recto-Sigmoid Index and Transitional Zone in Different Age Groups

	Neonates		Infants		Children		Total	
	RSI	TZ	RSI	TZ	RSI	TZ	RSI	TZ
Sensitivity,	33	50	100	80	80	60	68	63
% (CI)	(0.03-0.67)	(0.17-0.82)	(0.3-1)	(0.62-0.99)*	(0.68-0.93)*	(0.45-0.75)	(0.58-0.8)*	(0.51-0.74)*
Specificity,	56	78	79	92	86	93	81	91
% (CI)	(0.24-0.88)	(0.51-1)*	(0.60-0.97)*	(0.79-1)*	(0.75-0.97)*	(0.85-1)*	(0.72-1)*	(0.84-0.98)*
PPV,	33	60	50	67	33	43	39	56
% (CI)	(0.03-0.94)	(0.29-0.92)	(0.27-0.73)	(0.45-0.89)	(0.19-0.48)	(0.27-0.58)	(0.28-0.51)	(0.44-0.68)
NPV,	56	70	100	96	98	96	94	93
% (CI)	(0.24-0.88)	(0.41-0.99)	(0.66-1)*	(0.86-1)*	(0.94-1)*	(0.91-1)*	(0.88-0.99)*	(0.87-0.99)*

RSI = recto-sigmoid index; TZ = transitional zone; PPV = positive predictive value; NPV = negative predictive value; CI = confidence interval

*Clinically significant (lower limit of CI > 0.5).

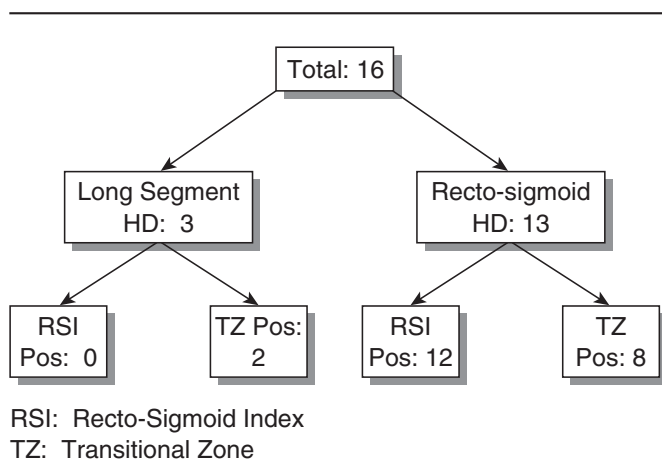


Figure 2. Radiologic findings in patients with biopsy-proven Hirschsprung's disease (HD). RSI = recto-sigmoid index; TZ = transitional zone; Pos = positive.

transitional zone was detected in only 8 of the 13 patients (62%; CI, 0.28-0.95). In 4 of the remaining 5 patients with recto-sigmoid HD, the diagnosis was made solely on the basis of measuring the recto-sigmoid index. In one patient, the diagnosis was missed by the radiologists.

Discussion

Our study shows that the reliability of the barium enema in the diagnosis of HD is age dependent. Whereas a negative result ruled out HD with a high probability (high negative predictive value) in children older than 1 month, the reliability of this test was less in the neonatal period.

Previous studies found a wide range of sensitivity and negative predictive values of the transitional zone on barium enema.^{1,5,13} Some of these studies focused on the patients' age, and in agreement with our findings, the authors reported a lower sensitivity of the transitional zone for the diagnosis of HD in the neonatal period compared with later in life (65% vs 75%).^{7,9,14} This finding suggests that in the neonate, additional tests such as rectal suction biopsy and/or anorectal manometry should be used to rule out HD.

In contrast, Reid et al,⁴ like us, found a high negative predictive value of the barium enema in patients older than 1 month (98%), suggesting that in this age group a normal barium enema reliably excludes HD.

On the other hand, our results showed generally low positive predictive values of barium enemas for the diagnosis of HD in all age groups. This finding suggests that barium studies are frequently over-interpreted and that clinical judgment should be used in the decision of whether additional tests are needed after an abnormal barium enema.

In general, we did not find any significant differences in the reliability of the recto-sigmoid index versus the transitional zone in the diagnosis of HD. However, certain observations are worth mentioning: As expected,¹⁰ the recto-sigmoid index cannot be used for the diagnosis of long-segment HD. However, the measurement of the recto-sigmoid index on X ray helped identify 4 additional patients with HD whose diagnosis would have been missed by looking at the transitional zone alone. These observations suggest that in the radiologic diagnosis of HD, both the transitional zone (a more subjective

parameter) and the recto-sigmoid index (a more objective measurement) should be evaluated.

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