

THE "RECTO-SIGMOID INDEX"*

A MEASUREMENT FOR THE EARLY DIAGNOSIS OF HIRSCHSPRUNG'S DISEASE

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THE roentgen diagnosis of Hirschsprung's disease may be established relatively easily when the classic colonic transition segment is identified by contrast enema examination. It may also be suspected if there is prolonged colonic retention of contrast medium. However, a typical transition and/or prolonged colonic stasis may not be apparent during the first days of life.^{2,4,5,10,13} Moreover, even in those cases where the presence of transition segments may be subjectively suspected on fluoroscopy, spot film documentation may be incomplete or inconclusive.

Early diagnosis of Hirschsprung's disease, nevertheless, remains a desirable objective, since immediate surgical intervention is of the utmost importance. An ob-

jective definition of the colonic transition segment associated with Hirschsprung's disease in the form of a measured range of recto-sigmoid relationships would, therefore, be desirable as an ancillary diagnostic aid. It would be particularly useful in questionable cases, where transition segments are equivocal or not readily demonstrable.

A condition which often mimics Hirschsprung's disease is the meconium plug syndrome.^{6,7,10,14-16} Despite several recently outlined roentgen criteria,¹⁰ which were found to be helpful in distinguishing the meconium plug syndrome from Hirschsprung's disease and other pathologic states, it was emphasized¹⁰ that Hirschsprung's disease remained the most difficult

TABLE I
CONTRAST ENEMA EXAMINATIONS IN NEWBORNS WITH HIRSCHSPRUNG'S DISEASE
VS. MECONIUM PLUG SYNDROME

Hirschsprung's Disease with No Apparent Transition Segment		Meconium Plug Syndrome	
Patient	"Recto-Sigmoid Index"	Patient	"Recto-Sigmoid Index"
I	0.7	1	1.3
II	0.8	2	1.6
III	0.8	3	1
IV	0.8	4	1.1
V	0.9	5	1.5
VI	0.8	6	1
VII	0.9	7	1.7
VIII	0.7	8	1
IX	0.7	9	1.8
X	0.7	10	1.4
Mean Value	0.8		1.3

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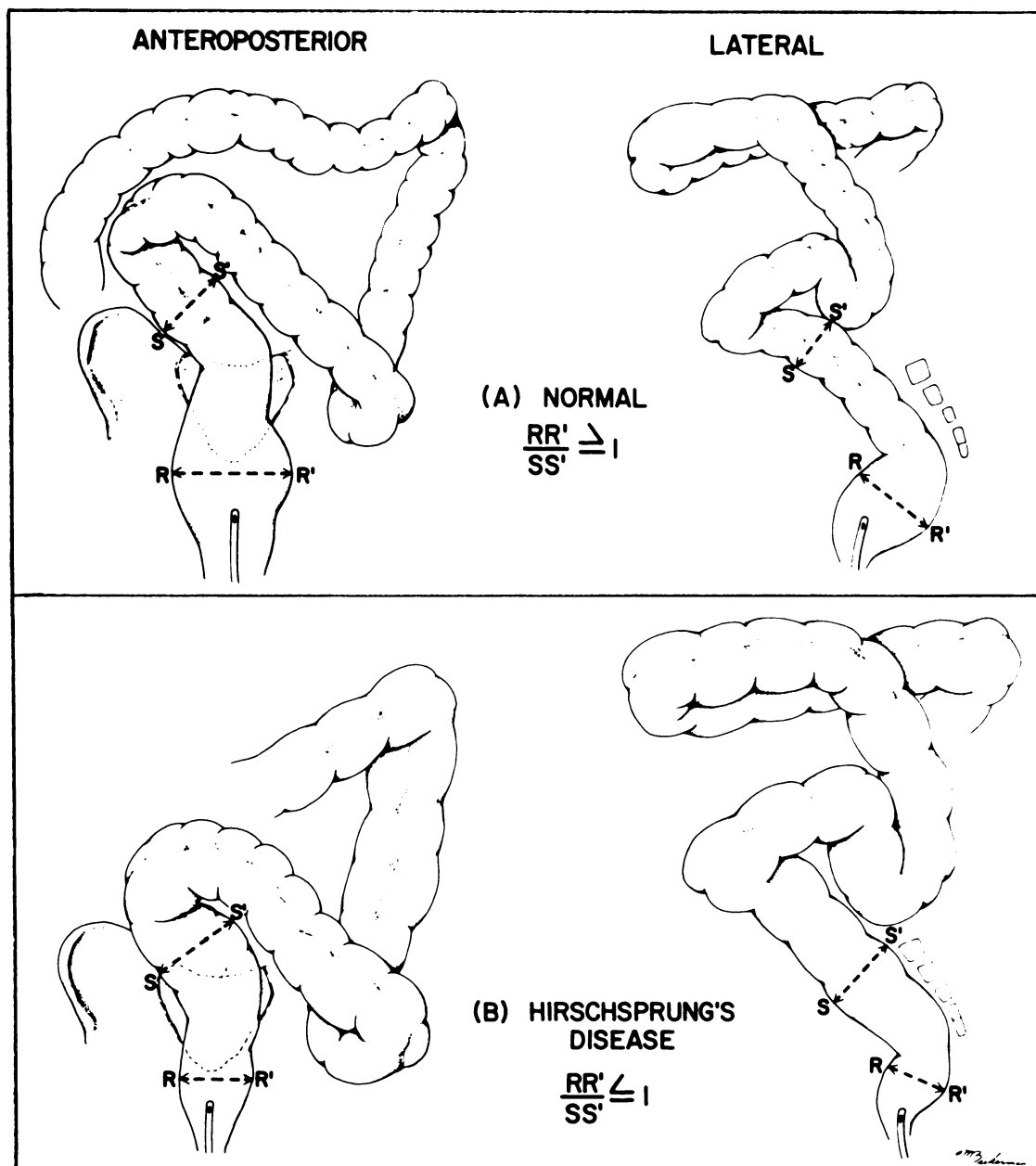


FIG. 1. (A) Normal "Recto-Sigmoid Index:" The ratio between the widest diameter of the rectum (RR') and the widest diameter of the sigmoid (SS') is larger or equal to 1; (B) abnormal "Recto-Sigmoid Index:" Hirschsprung's disease. The ratio is smaller than 1.

problem in differential diagnosis in view of occasional, but almost identical, clinical and roentgen findings.

THE "RECTO-SIGMOID INDEX"

In an attempt to strengthen current roentgen diagnostic criteria as they apply

to Hirschsprung's disease, our material of Hirschsprung's disease was reviewed. It has been our experience that the widest rectal diameter is usually smaller than the widest diameter of the sigmoid loop in Hirschsprung's disease, even in the absence of a roentgenographically definite transi-

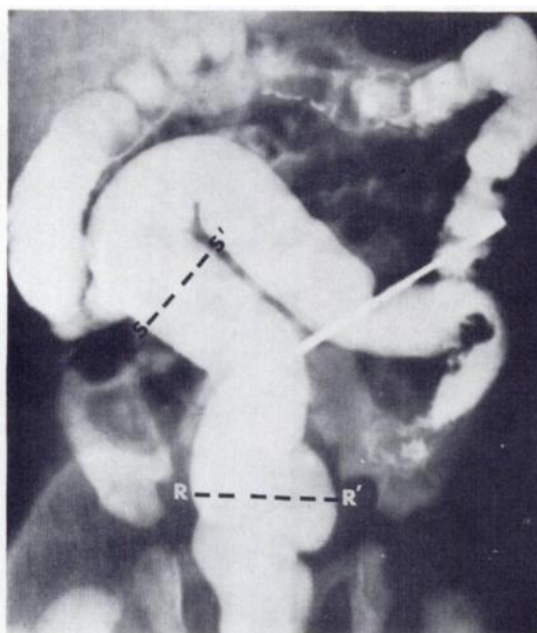


FIG. 2. Normal barium enema in a 2 day old female with a history of a mild episode of rectal bleeding. Long term follow-up did not disclose any clinical evidence of Hirschsprung's disease. Note that the rectum is wider than the sigmoid loop. The "Recto-Sigmoid Index" (RR'/SS') is 1.4.



FIG. 3. Normal barium enema in a newborn female. The contrast study was performed to determine cause of a single occurrence of rectal bleeding. The infant remained well for over 2 years with no clinical evidence of Hirschsprung's disease. The "Recto-Sigmoid Index" in this lateral view is 1.3.

tion segment. In normal newborn infants, as well as in cases of the meconium plug syndrome and other pathologic states, the

rectum was commonly found to be wider than the sigmoid loop. A numerical index was obtained in our review by dividing the widest diameter of the rectum by the widest diameter of the sigmoid loop, the "Recto-Sigmoid Index."

TABLE II
NORMAL CONTRAST ENEMA EXAMINATIONS
IN 12 NEWBORNS

Patient	History	"Recto-Sigmoid Index"
I	Mild diarrhea	1.9
II	Poor feeding, vomiting	1.5
III	Rectal bleeding	1.4
IV	Poor feeding, vomiting	1.5
V	Rectal bleeding	1.0
VI	Rectal bleeding	1.4
VII	Vomiting	1.1
VIII	Vomiting	1.5
IX	Vomiting	1.4
X	Vomiting and diarrhea	1.7
XI	Abdominal distention	1.0
XII	Abdominal distention	1.2
Mean Value		1.4

TABLE III
CONTRAST ENEMA EXAMINATIONS IN NEWBORNS
WITH MISCELLANEOUS PATHOLOGY OF THE
GASTROINTESTINAL TRACT

Patient	Diagnosis	"Recto-Sigmoid Index"
I	Meconium ileus	1.5
II	Midgut malrotation	1.4
III	Gastroschisis, post-operative	1.3
IV	Necrotizing enterocolitis of infancy	1.3
V	Jejunal atresia	1.3
Mean Value		1.4

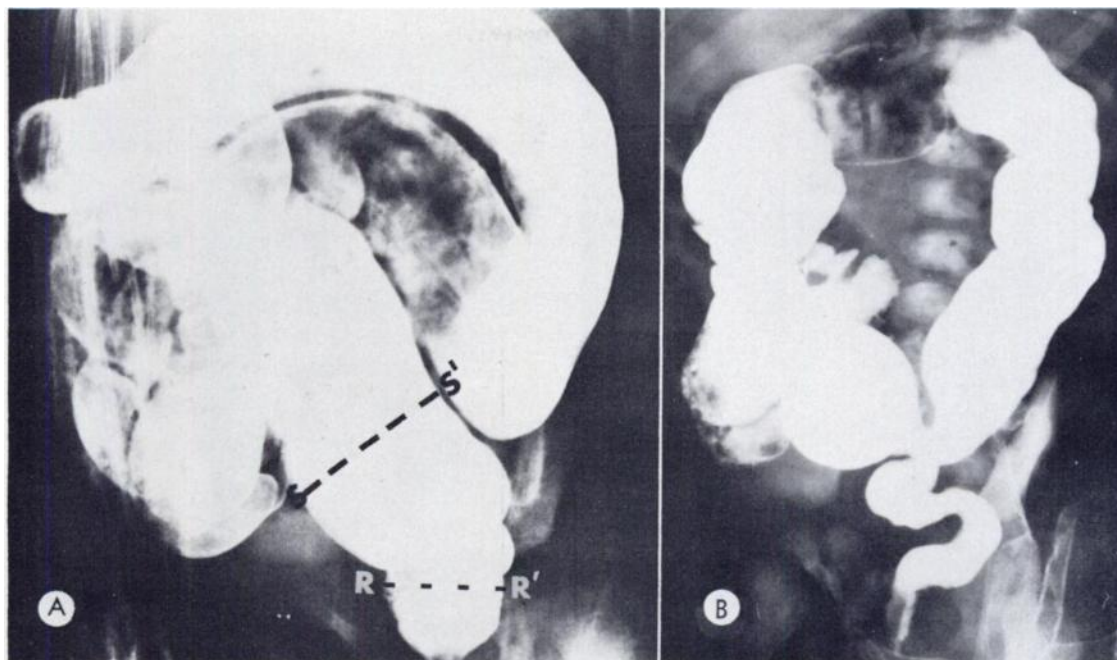


FIG. 4. Case 1. Newborn female with history of abdominal distention.

(A) Barium enema examination performed at 3 days of age. A transition segment is not apparent. The rectum, however, is narrower than the sigmoid. The "Recto-Sigmoid Index" (RR'/SS') measures 0.8 in this projection. (B) Repeat barium enema examination performed 1 week later. An obvious transition segment involving the rectum and distal segment is now present.

MATERIAL AND METHOD

The case material studied consisted of 21 biopsy proven cases of Hirschsprung's disease, 10 cases of the meconium plug syndrome and 12 cases of term newborn infants considered to be normal. Five cases of the authors' material of other diseases of the gastrointestinal tract including necrotizing enterocolitis of infancy,⁹ meconium ileus⁸ and midgut volvulus were also reviewed. All cases were studied with contrast enemas within the first 7 days of life, with the majority being examined during the first 4 days. All infants with the meconium plug syndrome, other gastrointestinal disease, and "normal" babies have been followed clinically for a period of at least 2 years and demonstrated no clinical evidence of Hirschsprung's disease.

Regarding the "normal" controls, these were infants who had a variety of mild disorders from poor feeding, vomiting and mild diarrhea, to unexplained mild rectal

bleeding for which no cause was revealed. In all instances, the contrast enemas were normal and there has been no evidence of gastrointestinal disorder. No rectal biopsies have been performed, however, in any of the control patients. We believe that it can be safely assumed that they do not have Hirschsprung's disease.

In each instance, the widest diameter of the rectum was obtained at any level below the 3rd sacral vertebra. This point was established arbitrarily on the basis of the usual position of transition segments. The sigmoid loop was usually measured at 3 points along its course (proximal, peak of the loop and distal sigmoid colon). The largest measurement was selected. In almost all instances the widest part of the sigmoid proved to be the distal sigmoid loop. All measurements were obtained along a transverse axis, vertical to the longitudinal axis of the colon at that point (Fig. 1, A and B).

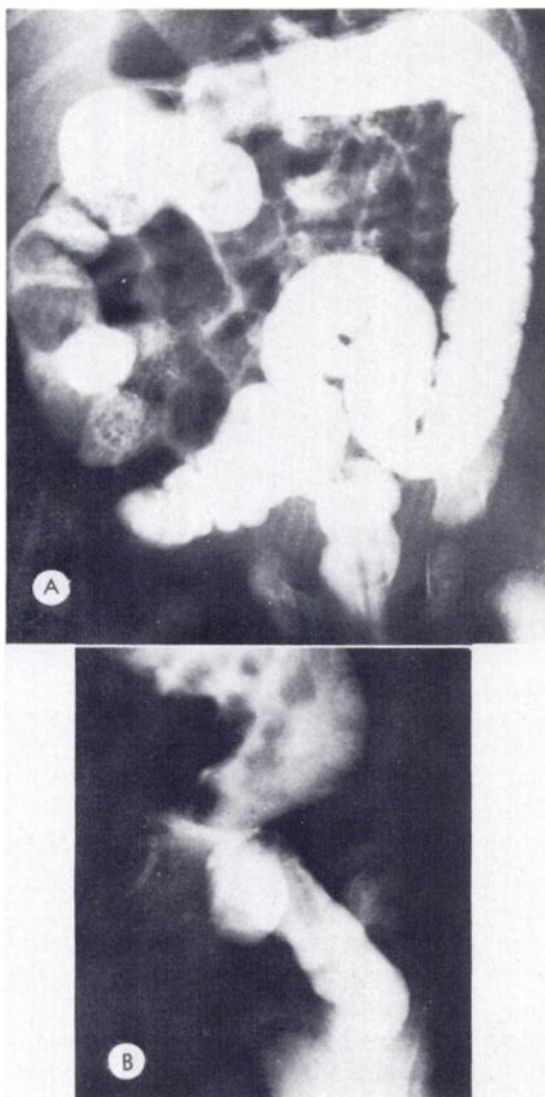


FIG. 5. Case II. Hirschsprung's disease. Barium enema examination performed at 2 days of age.

(A) Anteroposterior projection. (B) Lateral view. A definite transition segment could not be identified in this study, although the rectum appears to be of slightly smaller caliber than the sigmoid. The average "Recto-Sigmoid Index" in this study is 0.9. A repeat barium enema study performed 2 months later showed a definite transition segment.

Measurements were made in 3 projections, including anteroposterior, left posterior oblique and lateral. The "Recto-Sigmoid Index" was obtained by dividing the widest diameter of the rectum by the

widest diameter of the sigmoid loop when the colon was fully distended by contrast medium. Although absolute numbers varied according to the projection, the index proved to be fairly constant in any of the above projections, so that they consistently averaged out (Fig. 1, A and B).

Several types of rectal catheters were used, including a specially designed pediatric rectal catheter.¹² Internal rectal retention balloon catheters were used only in a few instances. These should be avoided since, in addition to other undesirable features,^{11,12} they may interfere with the desired measurements.

RESULTS

Among the 21 cases of Hirschsprung's disease, no definite or only a questionable colonic transition segment could be identified in 10 cases on the first contrast study. The average values of the "Recto-Sigmoid Index" taken in multiple projections of the latter 10 cases ranged from 0.7 to 0.9 with a mean value of 0.8 (Fig. 4-7; and Table I).

The average "Recto-Sigmoid Index" in the 12 normal newborns ranged from 1 to 1.9 with a mean value of 1.4 (Table II; and Fig. 2; and 3).

The "Recto-Sigmoid Index" was above

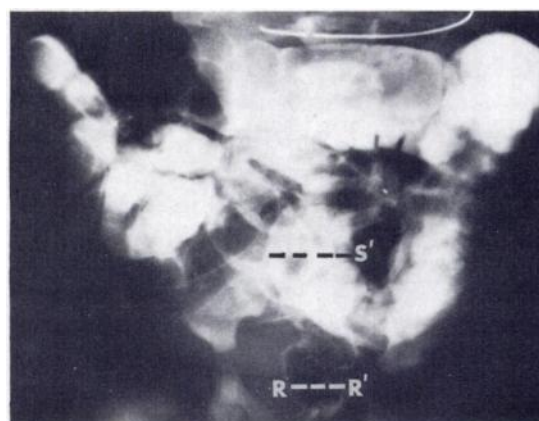


FIG. 6. Case III. Hirschsprung's disease. Contrast enema examination performed at 1 day of age. Anteroposterior projection. The rectum is not as wide as the sigmoid. The "Recto-Sigmoid Index" (RR'/SS') in this projection measures 0.6.

1 in all 5 cases of newborns with other pathologic processes of the gastrointestinal tract, with values ranging from 1.3 to 1.5 and the mean of 1.4 (Table III; and Fig. 9; and 10). A limited selective review of the literature^{2-10,13-15} was also undertaken and the "Recto-Sigmoid Index" was calculated when feasible. The results appeared to confirm the measurements of our own cases.

DISCUSSION

The above results suggest that the "Recto-Sigmoid Index" may be of value in the diagnosis of Hirschsprung's disease in the first few days of life, in the absence of a definite roentgenographic transition segment on contrast enemas (Fig. 4A; and 5-7). A "Recto-Sigmoid Index" of less than

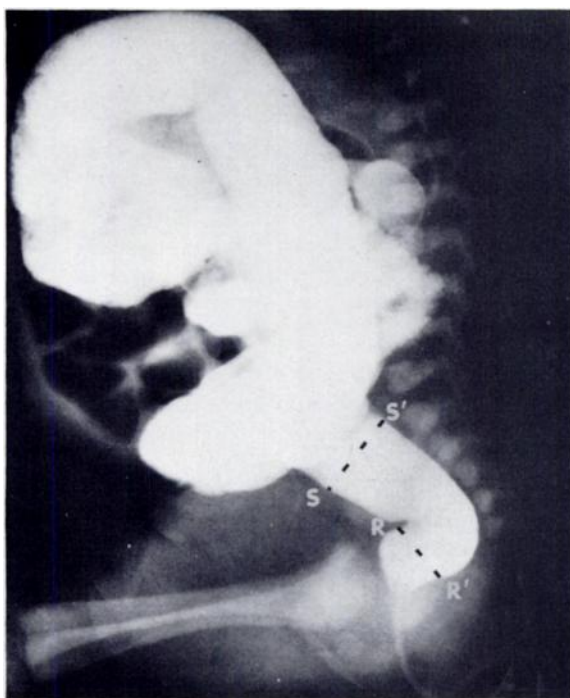


FIG. 7. Case x. Hirschsprung's disease. Four day old male with history of failure to pass meconium and abdominal distention. Contrast enema examination revealed only a questionable transitional segment. The "Recto-Sigmoid Index" (RR'/SS') measures 0.7 in this lateral view. (Courtesy of Dr. Henry Pritzker, Montefiore Hospital and Medical Center, New York City.)

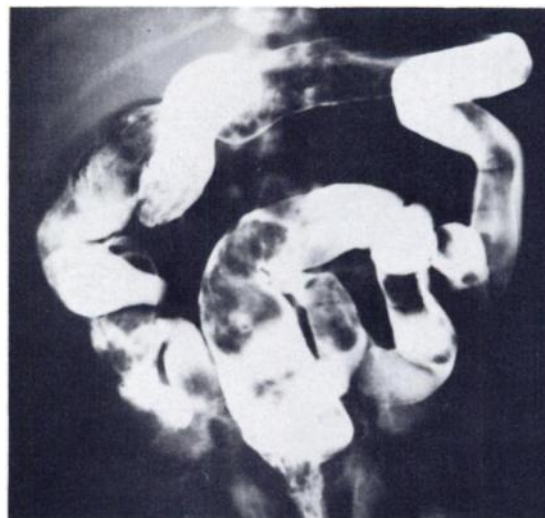


FIG. 8. Meconium plug syndrome. Premature, 18 hour old male with a history of abdominal distention and failure to pass meconium. The barium enema examination shows large intracolonic meconium masses. The rectum is wider than the sigmoid. The "Recto-Sigmoid Index" in this projection is 1.3. A large meconium plug was passed following completion of the study.



FIG. 9. Necrotizing enterocolitis of infancy. Two week old male. The barium enema examination demonstrates extensive pneumatosis intestinalis (arrows). The rectum is, however, wider than the sigmoid loop. The "Recto-Sigmoid Index" measures 1.3. Measurements were also based on oblique and lateral views.

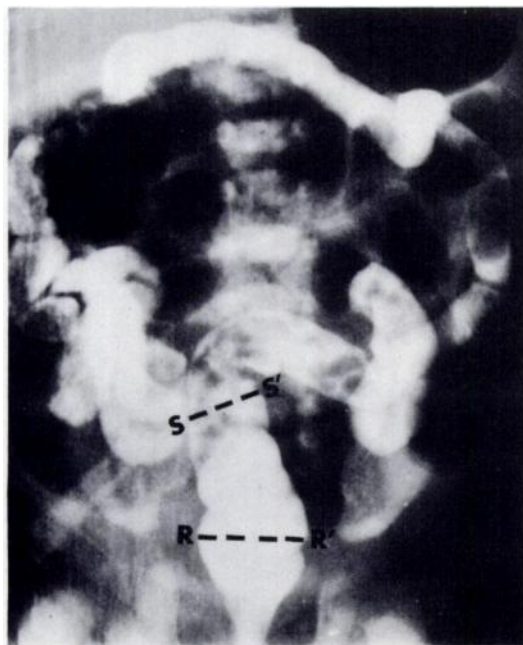


FIG. 10. Lower jejunal atresia. Newborn infant. The contrast enema examination demonstrates a microcolon. Nevertheless, the caliber of the rectum is larger than the sigmoid loop. The "Recto-Sigmoid Index" (RR'/SS') measures 1.3.

1 would be suggestive of Hirschsprung's disease.

The "Recto-Sigmoid Index" is obviously superfluous when a transition segment is clearly evident in the form of marked disproportion between the rectum and the proximal sigmoid colon (Fig. 4B). A less definite disproportion in the caliber of the distal aganglionic and normal proximal colon may be appreciated by the most experienced observers. However, we have seen considerable confusion and missed diagnoses in subtle cases. It is hoped that determination of the "Recto-Sigmoid Index" will provide a further diagnostic aid when a zone of transition is in doubt. The "Recto-Sigmoid Index" was not intended to be used in the rarer instances of aganglionosis involving other loci of the colon or with more extensive aganglionosis, including total aganglionosis of the colon and terminal ileum.^{1,2} The recto-sigmoid area remains, however, the most common site for the aganglionic segments of Hirsch-

sprung's disease.⁵ All of our cases of Hirschsprung's disease were confined to this location and the measurements may, therefore, serve as an aid to diagnosis in this region.

In the meconium plug syndrome (Fig. 8; and Table 1) and in other pathologic conditions which may simulate Hirschsprung's disease¹⁰ (Fig. 9; and 10) as well as in normal newborn infants (Fig. 2; and 3), the "Recto-Sigmoid Index" was almost invariably higher or at least equal to 1. A "Recto-Sigmoid Index" higher than 1 tended to exclude the diagnosis of Hirschsprung's disease in the current material. It should be noted, however, that in view of the limited number of cases collected and the known difficulty of establishing the diagnosis of colonic aganglionosis in early life, a rectal biopsy may be necessary in clinically borderline cases despite a "Recto-Sigmoid Index" higher than 1.

SUMMARY

The "Recto-Sigmoid Index" is an objective measurement which may prove to be useful in the early diagnosis of Hirschsprung's disease in the newborn, particularly in those cases where a colonic transition segment is not readily apparent or where its presence is questionable.

On the basis of our current case material, a "Recto-Sigmoid Index" higher than 1 may indicate a normal colon or a condition mimicking Hirschsprung's disease, such as meconium plug syndrome.

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