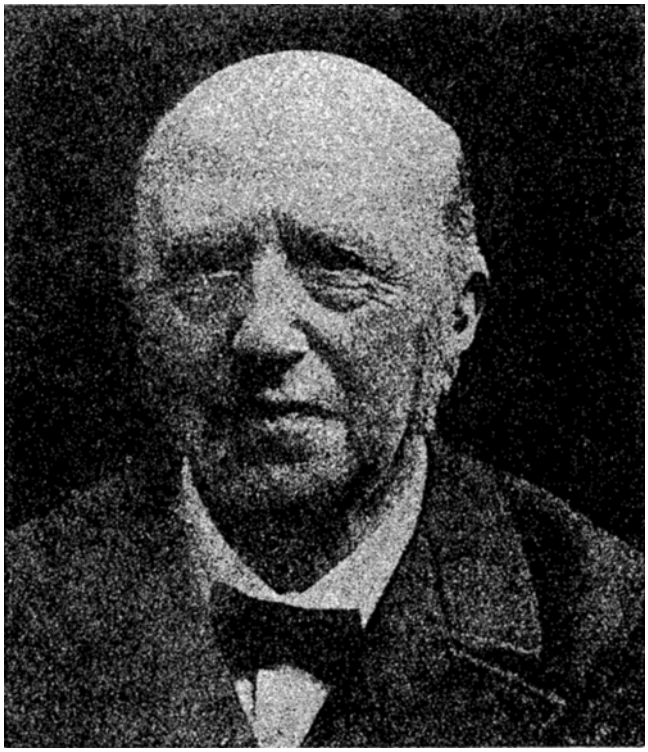
Classic Articles in Colonic and Rectal Surgery

MARVIN L. CORMAN, M.D., *Editor*

HARALD HIRSCHSPRUNG
1830–1916



Harald Hirschsprung was born in Copenhagen, Denmark, December 14, 1830. He passed his examinations in 1855 and was made a teacher in 1861. He was a Professor of Pediatrics at the University of Copenhagen and head physician to the Queen Louise Children's Hospital in Copenhagen.

He is best known for his description of the disease that was named after him. This was perhaps because of his excellent account of the condition, since his was not the original report. Parry described a case that is recorded in his collected papers in 1825. Levin of Chicago in 1867 described the first American case.

Hirschsprung contributed extensively to the pediatric literature, publishing one of the first comprehensive reports on pyloric stenosis (1888), and he was a pioneer in the use of hydrostatic pressure for the reduction of intussusception (1905).

Hirschsprung in this paper does not identify the pathogenicity, nor does he offer a suggestion for treatment. While theories abounded for 50 years (Lennander in 1900 was one of the first to suggest a neurogenic origin), there was so much confusion and difference of opinion that it was not until the 1940s that the absence of ganglion cells was felt to be the etiologic factor.

Hirschsprung died on April 11, 1916, at the age of 85.

I.

Stuhlträgheit Neugeborener in Folge von Dilatation und Hypertrophie des Colons.

Casuistische Mittheilung von

Prof. HIRSCHSPRUNG, Kopenhagen

in der Gesellschaft für Kinderheilkunde Berlin 1886.

Ich erlaube mir, dieser sachkundigen Versammlung zwei pathologische Präparate aussergewöhnlicher Art nebst ganz kurzgefassten Krankengeschichten vorzulegen; es ist meine Hoffnung, möglicherweise dadurch Bericht über ähnliche Beobachtungen oder einschlägige Bemerkungen von Seite der vielen anwesenden Collegen hervorzurufen.

From *Jahrbuch für Kinderheilkunde* 1888;27:1, with permission of the publisher.

Constipation in the Newborn as a Result of Dilation and Hypertrophy of the Colon

HARALD HIRSCHSPRUNG

LET me be permitted to present before this learned assembly two pathologic specimens of an unusual nature, together with short case histories. It is my hope that through this report I will learn of similar observations or call forth pertinent remarks on the part of many of my colleagues.

I shall at once present the first specimen to you. As you see, it is a large intestine, but of such size that it will certainly surprise you to learn that it came from a child who was only 11 months old when he died. When the abdomen was opened, there appeared a

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pair of enormously distended loops of intestine. The entire colon appears dilated. Only the rectum was not enlarged, but is actually somewhat narrowed. The described parts mentioned are not only dilated, but the wall is also noticeably thickened, particularly in the muscular part. There are very small superficial erosions and ulcerations throughout the mucous membrane, which show great differences in size and depth. These are discrete, small losses of substance and larger ones that apparently are formed by the coalescing of two or three others. In fact, in certain places the surface of the bowel presents a characteristic meshlike appearance over large areas. All the ulcers have a round or oval form with regular or undermined borders, as if they are carved out. Here and there one sees small follicle openings, but no swelling of the follicles. There is no trace of inflammation on the serosa. Ligaments and colon haustrae are not recognizable, but the appendices epiploicae are strongly developed. The mesentery of the sigmoid is high, broad, and thickened with rows of hypertrophied, bluish mesenteric glands the size of beans. In the under part of the ileum, Peyer's patches stand out strongly, but in the body there are no other important abnormalities

What symptoms did this unusual disease of the colon cause?

Shortly after the birth of the child, which took place in the Maternity Hospital in Copenhagen, it was seen that in spite of several different purges there was no stool. Only after repeated enemas did the bowels move. The same bowel problem continued during the following months, requiring various methods alternately employed; when evacuation was successful the stool was always of normal consistency and appearance. The child, moreover, got on well and thrived on breast milk and biscuits.

Because of the persistent difficulty with defecation, the child was referred for care to the Polyclinic. When I saw it for the first time it had had only one insignificant stool during the last 14 days. Nevertheless, the child did not seem especially distressed and its appetite remained good. The child had vomited only once. The abdomen was somewhat enlarged, but not to a great degree. An accumulation of fecal matter could be felt in the rectum and was removed partly by the finger and partly by irrigations. With the help of various laxatives, the condition remained quite good for a time, but when the appetite and cheerfulness began to wane, and the skin color became somewhat sickly, the child was brought into the hospital at the wish of the mother in November 1880. Its age was 8 months, weight 9,000 g, in a satisfactory nutritional status. During the entire stay in the hospital, which lasted two months, our interest was entirely concentrated on the bowel and particularly the purges. As a rule, the child was not feverish, rarely vomited, and had a good appetitie; in short, he gave no impression of suffering a serious illness. Bowel movements never occurred voluntarily. At times the abdomen was greatly distended, and all our efforts were directed at producing a bowel movement. When this was accomplished the abdominal distention disappeared, and the child felt noticeably better; but remission was always of short duration.

I admit that I then knew of no similar case either in the literature or in our own experience. A constriction of any kind affecting the bowel did not seem possible under the circumstances, at least not one of a permanent nature. The fact that a thick, elastic tube, more than an ell in length, as a rule extended with the greatest of ease, and that the rectum at examination was found full of fecal matter, which did not empty spontaneously, pointed as much toward a lower as a higher constriction in the intestinal tract and seemed rather to denote an atonic condition in the lower part of the gut. But the methods of treatment used in this connection, massage and electricity, proved meaningless. On the contrary, through daily enemas and laxatives, the bowels became regulated. The abdomen was seldom distended to a great degree, and the child left the hospital having gained 700 g during his stay. The time at home was only of short duration. For the first eight days the child got on very well. There was a spontaneous bowel movement that was thin in contrast to the earlier condition. Then the bowel again became markedly distended and the movements unusually large, frequent, and mustard colored. The child quickly became emaciated and he died the same day on which he was admitted again to the hospital. His weight was only 6,900 g.

This case study of such a long illness arouses a certain interest, but when one can report others of the same kind it takes on added significance, and some years later I saw a similar case.

Waldemar H., a seven-month-old infant, was taken to the Children's Hospital on April 19, 1885. Ever since birth he had suffered from retention of stools, and only by daily use of various aperients was it possible to establish defecation. At no time was he in pain. The child on the whole complained seldom, had no nausea or vomiting, and grew normally. Now and then the abdomen became distended, and the distention gradually increased. For a month it became so noticeable that the distressed mother brought the child to the Community Hospital, where capillary puncture with draining of air was carried out. Yet in a short time the abdominal distention increased again. After daily doses of castor oil in combination with enemas the boy left the hospital in satisfactory condi-

tion. Soon thereafter the abdomen began to enlarge again, in spite of the fact that bowel movements became copious, watery, and involuntary. He vomited only once, but the emaciated youngster was crying and manifestly suffering. He was then brought to the Children's Hospital.

He was a thin and frail child with an enormous abdomen, which measured 56 cm in girth. There was visible peristalsis. Rectal temperature was 38.4 C. No stool was felt in the rectum, but evacuation followed withdrawal of the finger.

During the stay in the hospital, which lasted four or five weeks, diarrhea alternated with inability to evacuate. The abdominal girth varied between 56 and 41 cm; he died with emaciation, which was the most outstanding symptom. Microscopic examination of the stool showed fine granular detritus, many small and larger fat droplets, and granular and epithelial and pus cells in small quantities.

There is certainly a great similarity in the case histories. From the first moment of life, the same unusual defecation problem was noted as was the identical abdominal distention and the same good health and growth. Vomiting was seldom seen, and other symptoms of ileus were entirely absent. Finally, in each case, diarrhea lessened the child's strength, causing weight loss and hastening the end of life. With the first case in mind, the astonishing similarity could not possibly elude us, and it seemed to us very probable that both cases must be of the same disease.

Now gentlemen, you should be able to convince yourselves of it. You perceive the obvious dilation of the transverse colon and the thickening of the intestinal wall. The dilation in the second case is a little less (16 cm) compared with the first patient (19 cm). You note on the mucous membrane the same type of damage with small and larger erosions, round and oval, through the entire thickness. Thus far, the cases are similar. Now you see in the second case separate larger and deeper ulcerations that penetrate to the serosa, and indications of peritonitis can be seen on the serosal surface. Near the larger ulcerations, we find an abscess under the mucosa that measures 2 cm in one direction and 1 cm in the other. Mottled spaces can be seen in the submucosa containing pus. Similar smaller spaces can be seen, but only a few at a time. That submucosal abscesses could penetrate further into the serosa than the mucous membrane is quite evident.

Let us now turn to solving the question of the nature of the ulcerations. It seems to me not possible to solve it with a word. There is no report of tuberculosis or dysentery.

[*At this point the author comments in some detail upon the nature of the ulcers. Hirschsprung concludes with the following comments.—Editor*]

. . . I shall abstain from any theoretical deductions. But upon consideration, one notices that the difficulty in defecation is present at the beginning of life. It appears unquestionable that the condition is caused *in utero,* either as a developmental anomaly or as a disease process.

Acknowledgment

Dr. Hirschsprung's article was translated from the German by Miss Anne C. Messer, Research Assistant, Sias Biophysics Research Division, Lahey Clinic Foundation. Miss Messer died in February 1981.

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