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NAME AND ADDRESS DATE

CONGENITAL MEGACOLON
(HIRSCHSPRUNG'S DISEASE)

BY

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TABLE OF CONTENTS

PART ONE

Chapter I

HISTORY

Although the literature contains many descriptions of congenital megacolon prior to 1886, it was not until after Professor Hirschsprung's classic description that the medical profession became generally cognizant of this disease entity. His case report entitled "Stuhlträgheit Neugeborener in Folge von Dilatation und Hypertrophie des Colons,"[1] presented to the Society for Pediatrics in Berlin, is an accurate description of the clinical course and the pathology of the disease.

Ruysch,[2] an anatomist, is credited with the first description in the seventeenth century. The case

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[1]Professor Hirschsprung, "Stuhlträgheit Neugeborener in Folge von Dilatation und Hypertrophie des Colons [Sluggishness of Bowels of Newborn Infants as a Consequence of the Enlargement and Hypertrophy of the Colon]," in Jahrbuch für Kinderheilkunde und Physische Erziehung, 27 (1887):1-7.

[2]J. Rawson Pennington, A Treatise on the Diseases and Injuries of the Rectum, Anus and Pelvic Colon (Philadelphia, c1923), 731.

described was in a five-year-old girl. In 1820, Billard[3] described a megacolon in an infant six days of age. Five years later a case of congenital megacolon in an adult was reported by Parry.[4] Lewitt,[5] of Chicago, in 1867 became the first American to report a case in the literature.

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[3]J. M. T. Finney, "Congenital Idiopathic Dilatation of the Colon (Hirschsprung's Disease)," Surgery, Gynecology and Obstetrics, 6 (1908), 626.

[4]Ibid., 626.

[5]Ibid., 626.

Chapter II

CLASSIFICATION

According to Bockus, "Primary megacolon may be defined as a great dilatation, elongation and hypertrophy of the sigmoid colon, involving at times the entire colon and rectum, associated with retention of enormous amounts of feces . . . ."[1] Such a definition could also include secondary megacolon, as the two types differ mainly in etiology. The former is commonly considered to be of congenital origin without a demonstrable obstruction,[2] whereas the latter is acquired secondary to such obstructing lesions as neoplasms and congenital strictures. Secondary megacolon will not be discussed in this paper.

Primary or congenital megacolon has recently been further divided into two groups by Higgins[3] and

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[1]Henry L. Bockus, Gastroenterology (Philadelphia, c1944), II, 398.

[2]See page 9 for a discussion of the present concept of etiology.

[3]T. Twistington Higgins and F. Douglas Stephens, "Discussion on Hirschsprung's Disease," Proceedings of the Royal Society of Medicine, 42 (1949):223.

Stephens.[4] These workers differentiate between "true Hirschsprung's disease" (Stephens) or "true Hirschsprung--severe type" (Higgins) in which they are able to demonstrate a narrow rectosigmoid segment distal to the dilated colon, and those cases which have a "terminal reservoir" (Stephens) [="chronic constipation" (Stephens) or "true Hirschsprung--mild type" (Higgins)] in which the dilatation includes the rectum to the anus. This classification is based on the clinical course of the disease as well as roentgenologic findings. Earlier Grimson, Vandergrift, and Dratz[5] made the observation that the prognosis in patients with dilatation of the rectum was better than in those where the dilatation was present only in the sigmoid and proximally. Thereby they suggested such a differentiation.

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[4]F. Douglas Stephens, "Hirschsprung's Disease," Proceedings of the Royal Society of Medicine, 41 (1948): 831.

[5]K. S. Grimson, H. N. Vandergrift, and H. M. Dratz, "Management and Prognosis of Megacolon (Hirschsprung's Disease)," American Journal of Diseases of Children, 68 (1944):110ff.

Chapter III

ETIOLOGY

After the medical profession had gradually become more aware of this disease entity, multitudinous articles were written, many of which offered a new theory or a variation of a previously suggested one. Bockus gives a concise summary of these as follows:

> "Etiology of Primary Megacolon
> Modified from causes given by Finney (1908), Barrington-Ward (1914) and Rankin, Bargen and Buie (1932).
>
> "Mechanical Causes (usually developmental)
>
> 1. Extreme mobility of sigmoid (mesosigmoid too long) resulting in torsion (Barth).
> 2. Increased length of colon, particularly of sigmoid loop (Marfan).
> 3. Mucosa of sigmoid thrown into valve-like folds as a result of redundancy (Perthes, Roser).
> 4. Kinking, angulation, or adhesions at rectosigmoid.
> 5. Overdevelopment of sphincteric structure at rectosigmoid.
> 6. Spasm of sphincter at the rectosigmoid (Goebel).
> 7. Aplasia of musculature of rectosigmoid (Concetti).
> 8. Drag of a sigmoid overloaded with meconium acting as a valve.

9. Partial atresia of anal canal, rectum, or sigmoid.

10. Imperforate anus (partial).

11. Congenital stricture of the rectum (David).

12. Spasm of anal sphincter due to fissure, ulcer, or other causes (Fenuuck, Kästner, Bensaude, Hurst).

"Inflammatory Causes

1. Infective or inflammatory process involving the colon primarily (Walker and Griffiths).

"Deranged Nervous Mechanism

1. Abnormality in sympathetic innervation of longitudinal muscle fibers of colon (Formad, 1891).

2. Neuromuscular segmental defect in colon or paralysis of a gut segment (Hawkins, 1907).

3. Hyperactivity of sympathetic innervation of distal colon (relief of lumbar sympathetic ramisectomy--Wade and Royle).

4. Hypoactivity of sacral parasympathetic innervation to distal colon.

   (a) Anal achalasia (Hurst, Wade, Lehman, Gask, and Ross).

   (b) Achalasia of musculature at rectosigmoid (degenerative changes in Auerbach's plexus--Cameron, Robertson and Kernohan).

   (c) Disease of sacral autonomic fibers (Ishikawa).

(d) Vitamin B_1 deficiency as cause of achalasia (Etzel).

(e) Lack of propulsive motility of distal colon."[1]

Hirschsprung believed that " . . . it must be either blamed upon an anomaly of development or an abnormal foetal process"[2] because the onset of symptoms in the two cases which he reported began at birth. In his review of the literature in 1908, Finney[3] suggested "hypernutrition, a species of giantism" based on his observation of greatly dilated lymph and blood vessels supplying the area of dilatation. A retroflexed uterus was incriminated by Richardson.[4] Levi[5] believed it was due to diastasis of the recti muscles.

As early as 1900 Fenwick observed that "the general appearance of the colon is so strikingly like that of a piece of intestine above an organic stricture that one is forced to the conclusion that some form of mechanical

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[1]Bockus, op. cit., 399f.

[2]Hirschsprung, op. cit., 7.

[3]Finney, op. cit., 631.

[4]Ibid., 630.

[5]Ibid., 630.

obstruction must have been present . . . ."[6] In several of his cases it was necessary to insert a tube " . . . some way up the bowel before the sigmoid could be evacuated, the rectum having apparently lost all power of contraction."[7] Our attention is again focused on this situation by Hawkins in 1907 who said: "A neuropathic origin admits of no proof. It is probable that Hale White's expression 'congenital inertness of the colon' takes us as far as we can go into the mystery. If by 'inertness' we mean a neuro-muscular defect, through which a section of the colon, though it opposes no obstacle, is yet (continuously or from time to time) incapable of forwarding its contents, I believe we have in this the basis of the disease. I think we may realize a difference between a physiological and a physical obstruction of the bowel."[8]

Cameron (1928)[9] agreed with a suggestion made previously by Hurst, that megacolon was comparable to

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[6]W. Soultau Fenwick, "Hypertrophy and Dilatation of the Colon in Infancy," British Medical Journal, 2 (1900):566.

[7]Ibid., 566.

[8]Herbert P. Hawkins, "Remarks on Idiopathic Dilatation of the Colon," British Medical Journal, 1 (1907):477.

[9]J. A. Munro Cameron, "On the Etiology of Hirschsprung's Disease," Archives of the Diseases of Childhood, 3 (1928):211.

cardiospasm. De Takats and Biggs (1938) made a statement similar to the one quoted from Fenwick (1900) as follows: "The intestinal obstruction of these patients has all the earmarks of a mechanical obstruction, and yet at autopsy no such obstruction can be found."[10]

Although there has been much discussion about a neurogenic origin, little attention has been given to the distal segment as a source of obstruction until recent years. Alvarez has given us a concrete idea regarding the distal segment in his hypothesis " . . . that some of the unyielding muscle rings seen in so-called cardiospasm, Hirschsprung's disease, . . . are due, not to an excess of nervous action, but to an absence of it."[11] He goes on to say " . . . that the simplest explanation for the production of cardiospasm and Hirschsprung's disease would be a loss of function in certain ganglion cells in the intestinal plexuses."[12] Earlier Adamson and Aird[13] showed that progressive megacolon

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[10]Geza de Takats and Alfred D. Biggs, "Observations on Congenital Megacolon," Journal of Pediatrics, 13 (1938):822.

[11]Walter C. Alvarez, An Introduction to Gastroenterology, (New York, 1948), 219.

[12]Ibid., 321.

[13]W. A. D. Adamson and Ian Aird, "Megacolon: Evidence in Favour of a Neurogenic Origin," British Journal of Surgery, 20 (1932):226ff.

could be produced in the cat by removal of the parasympathetic nerve supply to the distal colon. During the past few years Whitehouse and Kernohan[14] in this country and Bodian[15] in England have shown microscopically an absence of the ganglion cells of the myenteric plexus in the distal nondilated segment of bowel in cases of Hirschsprung's disease.[16]

Swenson and associates,[17] of Boston, recently published the results of balloon studies made on patients with congenital megacolon and added further evidence to their postulation that " . . . congenital megacolon is due to malfunction of the rectosigmoid that results in partial colonic obstruction." On patients with colostomies, they positioned balloons in the splenic flexure, lower descending colon, and rectosigmoid.

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[14]Francis R. Whitehouse and James W. Kernohan, "Myenteric Plexus in Congenital Megacolon: Study of Eleven Cases," Archives of Internal Medicine, 82 (1948): 75-111.

[15]Martin Bodian, F. Douglas Stephens, and B. C. H. Ward, "Hirschsprung's Disease and Idiopathic Megacolon," Lancet, 256 (1949):8.

[16]See page 19 for further discussion.

[17]Orvar Swenson, Harold F. Rheinlander, and Israel Diamond, "Hirschsprung's Disease: A New Concept of the Etiology," The New England Journal of Medicine, 241 (1949):551ff.

Food was then given to stimulate peristalic action. In their normal controls they found " . . . progression of the propulsive peristalic waves from transverse colon to the anus." In five cases of Hirschsprung's disease the two balloons in the splenic flexure and descending colon showed peristalsis, but the rectosigmoid recording showed no peristalsis. Three of the tracings from the distal balloon in the rectosigmoid segment showed a " . . . larger rise in the height of the recording base-line . . . indicating increased tonus . . . ." Three also showed " . . . rhythmic series of segmental contractions of low amplitude . . . dissociated from the peristalic waves in the descending colon." In three cases, following resection of this narrowed, nonfunctioning section in the rectosigmoid portion of the bowel,[18] normal progression of peristalsis was shown by balloon studies.

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[18]See page 41 for further discussion.

Chapter IV

INCIDENCE

Much of the statistical data on congenital megacolon is from early reports. At the London Hospital Fenwick[1] found only three cases of megacolon in early life in 30,000 autopsies which had been performed over a period of sixty years. Pennington[2] gives figures by White who found one case in 16,000 autopsies at Guy's Hospital. At London Children's Hospital Barrington-Ward[3] encountered nineteen cases between 1868 and 1916. At St. Luke's Hospital, Chicago, de Takats and Biggs[4] found nine cases in 79,035 admissions, an incidence of one in 9,000 hospital admissions.

Finney[5] found that 70% of the cases occurred immediately after birth and 20% occurred within a few days

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[1]Fenwick, op. cit., 564.

[2]Pennington, op. cit., 732.

[3]Ibid., 732.

[4]de Takats and Biggs, op. cit., 819.

[5]Finney, op. cit., 634.

or weeks. Pennington, however, in summarizing 420 cases from various authors, found the age ranged from the newborn to age 78 with the following distribution:[6]

"Up to 1 year 44
1 to 5 85
6 to 10 57
11 to 20 78
21 to 30 67
31 to 40 29
41 to 50 16
51 to 60 25
61 to 70 12
over 70 . . . . . (420) 7 "

The disease is more frequent in the male than in the female. Finney[7] found two cases in the adult male to one in the adult female. Lowenstein[8] found the ratio of male to female to be three and one-half to one, but Higgins[9] gives this as high as five or six to one.

The literature contains reports of more than one case occurring in a family, but such are not common. Cases from a series at Johns Hopkins Hospital were

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[6]Pennington, op. cit., 733.

[7]Finney, op. cit., 634.

[8]Ibid., 634.

[9]Higgins and Stephens, "Discussion on Hirschsprung's Disease," op. cit., 222.

reported by Duhamel and Bing.[10] Pennington[11] states there is a familial tendency and cites four probable instances from various sources. In 1948, Zuelzer and Wilson[12] reported five children from one family who died, all of whom had presented either the picture of intestinal obstruction or congenital megacolon. The father of these children had a history of constipation since birth with severe illnesses during childhood because of his constipation.

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[10]Finney, op. cit., 634.

[11]Pennington, op. cit., 738.

[12]W. W. Zuelzer and J. L. Wilson, "Functional Intestinal Obstruction on Congenital Neurogenic Basis in Infancy," American Journal of Diseases of Children, 75 (1948):49ff.

Chapter V

PATHOLOGY

The degree of involvement may vary greatly from case to case. According to Finney,[1] the sigmoid alone is involved in more than one-third of the cases, only 15% have involvement of the whole colon, and the rectum and small intestine are "rarely" included. David[2] gives a higher per cent for cases involving only the sigmoid, placing it at about 50%. Involvement of the whole colon is next in frequency, and he states that the rectum is usually normal. In contrast to the figures of these authors regarding the frequency of involvement of the rectum, Herrmann[3] gives figures from cases at the

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[1]Finney, op. cit., 636.

[2]Vernon C. David, "Diseases of the Colon and Rectum," in Irvine McQuarrie (ed.), Brennemann's Practice of Pediatrics (4 vols., Hagerstown, Maryland, W. F. Prior Co., Inc., 1948), Vol. III, Chap. 7, p. 11.

[3]Louis G. Herrmann, "The Management of Megacolon (Hirschsprung's Disease)," Surgical Clinics of North America, 26 (1946):1178f.

Mayo Clinic and states that 42.3% had involvement of the rectum.

At times the bowel reaches an astounding size. Many[4, 5, 6] have reported specimens with a circumference of about 20 inches.

During a surgical procedure Herrmann[7] removed 42 pounds of fecal material and colon. Formad[8] reports a case with 40 pounds of fecal material. In other instances the capacity has been found to be 16 liters (Peacock),[9] 15 quarts (White),[10] and 17 quarts (Puls).[11]

Besides the tremendously dilated and hypertrophied colon there is thickening of the mesocolon and enlargement of the blood vessels, lymph channels, and glands

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[4]Hawkins, op. cit., 478.

[5]Frederick Treves, "Idiopathic Dilatation of the Colon, Illustrated by a Case in Which the Entire Rectum, Sigmoid Flexure, and Descending Colon Were Excised," Lancet 1 (1898):277.

[6]Pennington, op. cit., 740.

[7]Herrmann, op. cit., 1190.

[8]Finney, op. cit., 637.

[9]B. M. Bosworth, D. S. Hymen, and J. R. Lisa, "Modern Management of Megacolon," American Journal of Surgery, 75 (1948):811.

[10]Treves, op. cit., 277.

[11]David, op. cit., in Brennemann's Practice of Pediatrics, Vol. III, Chap. 7, p. 11.

in that area. The haustral markings and taenia become less pronounced or may be obliterated. Little and Calloway[12] state that the thickness of the wall[13] ranges from 1/3 to 1/2 inch. Ladd and Gross[14] give the thickness as 1/8 to 1/4 inch. Carr[15] found calcification in the wall of the upper rectum and lower pelvic colon. The mucosa is frequently ulcerated--a pressure necrosis. Petechial hemorrhages (de Takats and Biggs),[16] areas of pigmentation (Finney),[17, 18] and ecchymoses (Pennington)[19] have been found in the mucosa. Hirschsprung[20] reported submucosal abscesses.

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[12]Pennington, op. cit., 740.

[13]Normal bowel wall thickness usually ranges from 1/15 to 1/25 inch, according to Pennington, op. cit., 740.

[14]William E. Ladd and Robert E. Gross, Abdominal Surgery of Infancy and Childhood (Philadelphia, 1941), 141.

[15]Pennington, op. cit., 741.

[16]de Takats and Biggs, op. cit., 839.

[17]Finney, op. cit., 637.

[18]Pennington, op. cit., 741.

[19]Ibid., 741.

[20]Hirschsprung, op. cit., 6.

The microscopic changes involve all layers of the bowel wall, the hypertrophy of the muscle, however, usually being the predominant feature. As described by Finney,[21] the mucosa was found to be two times its normal thickness with an increase in the connective tissue stroma. The mucosal vessels were enlarged, particularly the veins. There was a diffuse round cell infiltration and masses of yellowish brown pigment were found. In some areas the surface epithelium was missing and in others the columnar epithelium had become more cuboidal in type. In the submucosa endarteritic changes were present in the vessels. He described areas of hyalinization of the tissue as well as "fascicles of connective tissue." Pennington[22] mentioned a round cell proliferation in this layer.

The circular muscle constitutes the largest portion of the bowel wall making up 42% of the wall, whereas normally it would be 20% or less (Finney).[23] The muscle cells show no abnormality.

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[21]Finney, op. cit., 637ff.

[22]Pennington, op. cit., 741.

[23]Finney, op. cit., 639.

The most interesting and significant changes are those found in the myenteric plexus. Although there have been descriptions of the ganglion cells by some authors, it was not until 1948 that a complete, well-controlled study of the myenteric plexus in megacolon was published by Whitehouse and Kernohan. In addition to studying sections from eleven cases of congenital megacolon, they studied a series of 78 control cases which included consecutive post-mortem records, cases of peritonitis, intestinal obstruction, and others. In the control cases they found:

"1. Ganglions are present in practically every section taken from the colon.

2. So-called degenerative changes in the ganglion cells are commonly encountered in sections taken under the conditions of the control cases and the cases of megacolon.

3. Nerve trunks (nonmyelinated) in the location of the myenteric plexus are extremely rare in the rectum and in other parts of the colon.

4. Extreme dilatation of the colon has relatively little effect on the myenteric plexus apart from the fact that the ganglions seem to be somewhat more widely spaced than normal.

5. Inflammatory changes in the colon have little effect on the myenteric plexus in the majority of instances.

6. The rectum contained as many ganglions per unit area as other parts of the colon or more."[24]

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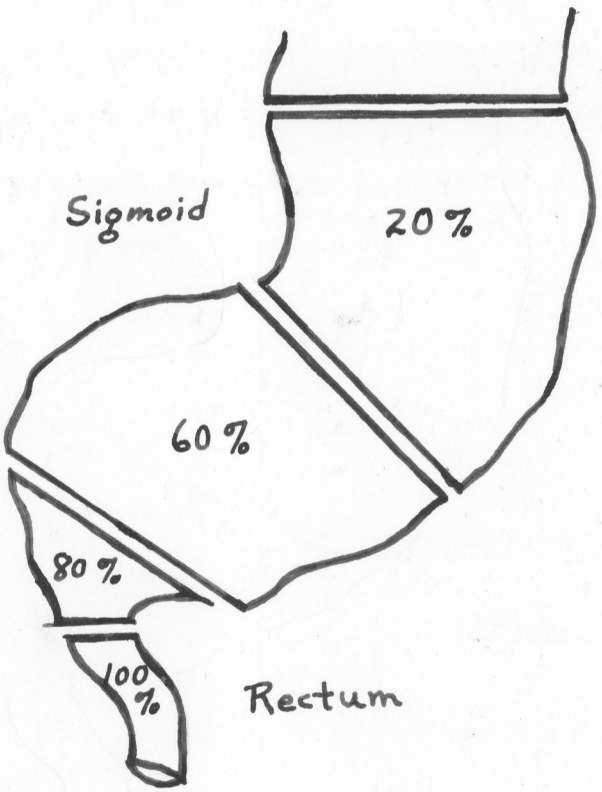
[24]Whitehouse and Kernohan, op. cit., 82f.

Then in the eleven cases of megacolon which they studied, no ganglion cells were found in the narrow distal segment and only 20% of the cases showed ganglion cells in the "transitional area" at the distal end of the megacolon. In the narrow distal segment all cases of congenital megacolon showed nerve trunks in the location of the myenteric plexus. These, as noted above, were not seen in the controls. The positive findings of Whitehouse and Kernohan are well summarized in the following figure taken from their article:

(See the following page.)



"Percentage of cases in which the myenteric plexus was absent from the terminal part of the rectum to the indicated level."[25]

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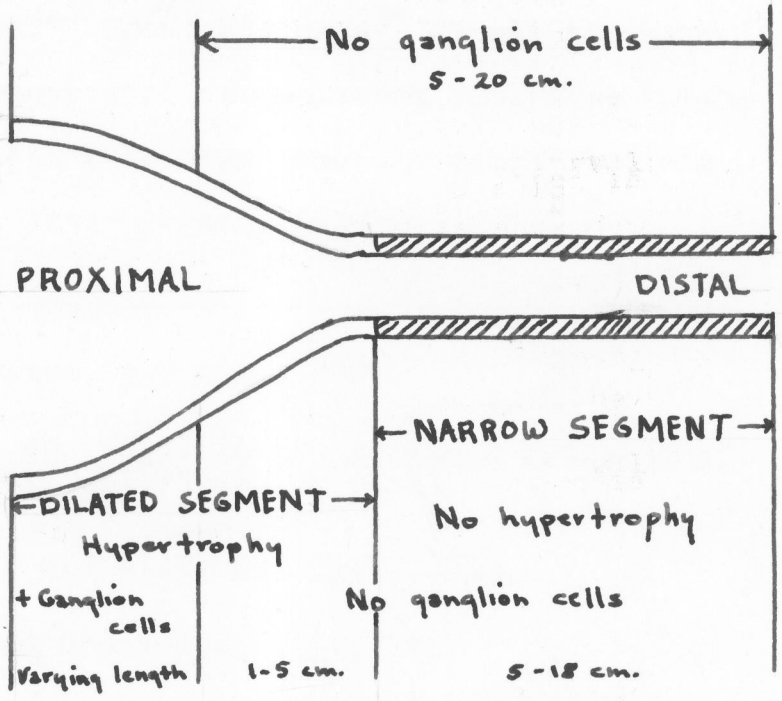
[25] Ibid., 90, figure 5.

In 1949, the work of a similar study by Bodian was published. In the eleven control cases he was able to demonstrate ganglions at close intervals to within 3.15 millimeters of the anocutaneous junction. The results of studies made of fifteen specimens, which included fourteen cases of typical Hirschsprung's disease and one case of acute intestinal obstruction, are best shown in the following figure which is found in the report in Lancet, 1949.



"Pathology of 15 cases of Hirschsprung's disease with narrow segment."[26]

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[26]Bodian, Stephens, and Ward, "Hirschsprung's Disease and Idiopathic Megacolon," op. cit., 8, figure 3.

Although most of the pathology is found in the gastrointestinal tract, other changes sometimes occur secondarily. Dilatation of the bladder has been found in many cases.[27, 28, 29] Ladd and Gross[30] state that occasional megalo-ureter occurs. Fenwick[31] reported a case in which there was bilateral dilatation of the ureters and kidney pelves, and Gee[32] found a double hydronephrosis in a case of megacolon. On microscopic examination of the liver, de Takats and Biggs[33] found "fatty changes." Fenwick's case[34] showed atelectasis of the lung bases with compensatory emphysema in the upper parts. In this same case the myocardium was shown to have fatty degeneration.

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[27]Pennington, op. cit., 740.

[28]David, op. cit., in Brennemann's Practice of Pediatrics, Vol. III, Chap. 7, p. 11.

[29]Adamson and Aird, op. cit., 221.

[30]Ladd and Gross, op. cit., 142.

[31]Fenwick, op. cit., 567.

[32]Finney, op. cit., 632.

[33]de Takats and Biggs, op. cit., 839.

[34]Fenwick, op. cit., 566.

Chapter VI

SYMPTOMS

The classic symptoms of Hirschsprung's disease are abdominal distention and a marked degree of constipation. In many cases the first sign of malfunction is a delay in the passing of meconium after birth. Ravitch[1] believes that a history of constipation from birth is essential before the diagnosis of megacolon can be made. The degree of constipation varies, but some patients do not have a bowel movement for weeks. Pennington[2] presents three unusual cases from the literature which went 7, 3 3/4, and 2 months, respectively, without a bowel movement.

Fenwick[3] stated that " . . . minor symptoms of the disease [,] loss of flesh, a foul tongue, great thirst, and enormous appetite, and a strong disinclination to mental or physical exertion were frequently present."

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[1]Mark M. Ravitch, "New Trends in Pediatric Surgery," Surgical Clinics of North America, 29 (1949), 1548.

[2]Pennington, op. cit., 744.

[3]Fenwick, op. cit., 566.

Anorexia, lassitude, and malnourishment are commonly found in untreated cases. Emesis and diarrhea are not common presenting symptoms, but do occur. Diarrhea may occur if a fecal impaction is present. In 1898, Treves[4] stated that there may be " . . . obstinate and often most persistent hiccough."

In severe, uncontrolled cases the greatly increased intra-abdominal pressure sometimes causes atelectasis of the lung bases. In these cases dyspnea would be a prominent symptom.

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[4]Treves, op. cit., 277.

Chapter VII

PHYSICAL FINDINGS

Many of the physical findings are suggested by the symptoms. On inspection one commonly sees a poorly nourished child with a protruding abdomen. If the case is long-standing, the costal margins are flared. Peristalsis or the pattern of intestinal loops is usually visible. The umbilicus may be everted and there may also be diastasis recti. The dilated veins are prominent in the thin, tense abdominal wall. In severe cases the patient may show evidence of dyspnea or orthopnea. If there is compression of the large veins in the abdominal cavity, the legs will be edematous. Palpation of the abdomen reveals an enlarged colon which is easily outlined because of the large amount of retained feces. The percussion note is usually tympanitic. In percussing the lung fields, the resonance may stop at a higher level than normally because of the elevation of the diaphragm, and the heart may be displaced laterally. The bowel sounds are more active than normal but may become high-pitched and tinkling if impacted, inspissated feces or volvulus of the redundant colon causes obstruction. The findings on digital examination of the rectum

are not constant--a large, empty rectum or a fecal impaction may be found. Peterman[1] reports finding dilated hemorrhoidal veins.

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[1]M. G. Peterman, "Congenital Megacolon. Treatment with Mecholyl Bromide," Journal of Pediatrics 27 (1945): 490.

Chapter VIII

OTHER CONGENITAL ANOMALIES

There have been several cases of congenital megacolon reported in which there have been coexistent anomalies of development. The most common of these is an imperforate anus. Imperforate anus may be a cause of secondary megacolon, but in the cases presented here the two conditions seem to be independent of each other. Peterman[1] reported a case in which a patient was constipated and passed large stools from birth after a stab wound was made to correct an imperforate anus. In four cases presented by Bodian, Stephens, and Ward[2] an "anal membrane" was perforated digitally after birth. De Takats and Biggs[3] operated on a patient seven days of age to correct an imperforate anus. This child did not present symptoms of megacolon until four to five years of age. In this same patient there was also found to be a transposition of the heart and liver.

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[1]Peterman, op. cit., 487.

[2]Bodian, Stephens, and Ward, "Hirschsprung's Disease and Idiopathic Megacolon," op. cit., 6.

[3]de Takats and Biggs, op. cit., 841.

Chapter IX

LABORATORY AND ROENTGEN FINDINGS

The pertinent laboratory findings are of a non-specific type. In many cases a secondary anemia is present. This is probably a nutritional type in most instances. However, Bodian, Stephens, and Ward[1] have reported finding occult blood in the feces of two patients. Although not commonly reported, it probably occurs frequently because of the superficial ulceration of the mucosa and may explain the anemia in some of the cases. Albuminuria has been present in some. Fenwick[2] reported hematuria in a patient who had compression of both ureters.

The common X-ray findings are the great dilatation and large capacity found with barium enema studies, as well as decreased haustral markings and the usual delay of evacuation. In 1948, Swenson and Bill[3] reported an

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[1]Bodian, Stephens, and Ward, "Hirschsprung's Disease and Idiopathic Megacolon," op. cit., 6.

[2]Fenwick, op. cit., 567.

[3]Orvar Swenson and Alexander H. Bell, Jr., "Resection of Rectum and Rectosigmoid with Preservation of the Sphincter for Benign Spastic Lesions Producing Megacolon," Surgery, 24 (1948):212.

interesting observation. In twenty patients they were able to demonstrate "an area of spasm" just distal to the area of dilatation. They found this segment under the flouroscope by a slow instillation of the barium suspension which was stopped as soon as the lower end of the dilated bowel was seen. This narrow segment was again shown to be demonstrable by Ward[4] in England. He made the interesting observation in three cases that there were segmentation movements in the narrow segment. This finding corresponds to a similar observation made by Swenson, Rheinlander, and Diamond[5] in their balloon studies.[6]

On the basis of his X-ray studies, Ward[7] also classified his cases into two groups,[8] according to whether the dilatation ended in the sigmoid or included the rectum. In addition to these two groups, twelve cases which clinically had been diagnosed as megacolon showed no abnormality on radiological examination.

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[4]Bodian, Stephens, and Ward, "Hirschsprung's Disease and Idiopathic Megacolon," op. cit., 7.

[5]Swenson, Rheinlander, and Diamond, "Hirschsprung's Disease: A New Concept of the Etiology," op. cit., 554.

[6]See page 10 for further discussion.

[7]Bodian, Stephens, and Ward, "Hirschsprung's Disease and Idiopathic Megacolon," op. cit., 7f.

[8]See page 4 for further discussion.

Chapter X

TREATMENT

Many procedures have been advocated; until recently, however, no method of treatment seemed to be successful consistently. In general the methods of treatment fall into the two large categories of a medical and a surgical approach to the problem. Prior to comparatively recent advances in therapeusis--which have made surgery safer and medical treatment more specific--the mortality with either type of treatment was high. Considering both types, surgery offered the best prognosis. De Takats has summarized some early statistics on the mortality following surgical and medical treatment.

"Mortality Statistics Following Conservative and Surgical Treatment Exclusive of Sympathectomy."[1]

Author	Number of Cases	Mortality Following	
		Conservative Treatment Percentage	Surgical Treatment Percentage
Danziger	94	75	34
Lowenstein	2	88	22
Schmidt	119	60	40
Neugebauer	254	68	5
Schneiderlöhn	268	79	52
Jadd & Thompson	42	19	23
Ask-Upmark	102	37	52

[1] de Takats and Biggs, op. cit., 828.

Until about ten years ago medical treatment was of a nonspecific type and consisted mainly of enemas and purges to keep the colon empty in an attempt to promote the general well-being of the patient as well as to prevent such complications as perforation and volvulus. Finney advocated " . . . cathartics, enemata, the use of the rectal tube, massage, electricity, tonics, exercise, regulation of the diet, etc."[2] Fenwick believed that enemas aggravated the colon and suggested suppositories, massage, and a well-fitting belt.[3] Some have advocated rectal dilatation.[4, 5] De Takats and Biggs[6] observed weeks of relief in two patients following manual removal of rectal impactions. They believed the indications for medical management to be as follows:

> " . . . (1) in an effort to minimize distention of the bowel and prevent nutritional changes in all infants and children up to the age of three years;

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[2]Finney, op. cit., 635.

[3]Fenwick, op. cit., 567.

[4]Ladd and Gross, op. cit., 147.

[5]de Takats and Biggs, op. cit., 826.

[6]de Takats and Biggs, op. cit., 826.

(2) in all mild and latent cases, even after that age, if response to treatment is satisfactory and the child is growing normally;

(3) as a preoperative measure, once it is recognized that the case is not medically tractable."[7]

Bodian, Stephens, and Ward stress the following principles in medical management:

"1. Thorough and repeated evacuation of the bowels.

2. Purgation.

3. Education in normal bowel habits."[8]

They believed that this regime should be used in the cases of "Hirschsprung's disease--mild" or cases of "chronic constipation."[9]

Between 1935 and 1940 another tool was added to aid in conservative therapy, viz., drugs acting directly on the colon. In 1938, Klingman[10] reported that five of seven cases responded to the use of syntropan (an

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[7] Ibid., 827.

[8] Bodian, Stephens, Ward, "Hirschsprung's Disease and Idiopathic Megacolon," op. cit., 10.

[9] See page 4 for further discussion of classification.

[10] Walter O. Klingman, "The Treatment of Neurogenic Megacolon with Selective Drugs," Journal of Pediatrics, 13 (1938):805ff.

atropine-like substance that is less toxic and with fewer disagreeable side effects). Peterman[11] reported a case in 1945 which was treated successfully with syntropan. This patient had failed to respond to a parasympathicomimetic drug.

The action of the drug which paralyzes the parasympathetic nerves was not well understood by Klingman who stated that "in some respects the use of a parasympathetic paralysant is almost paradoxical, and one is forced to accept it on the grounds that it is effective, the means by which it acts being as yet unexplained."[12] Myerson[13] believed its effectiveness might be explained by its inhibition of acetylcholine at the junction of the preganglionic and postganglionic fibers in the sympathetic nervous system, thus bringing the sympathetics and the parasympathetics into a better balance.

In 1937, Myerson and his group published the results

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[11]Peterman, op. cit., 490f.

[12]Klingman, op. cit., 808.

[13]Merl J. Carson, "Parasympathicomimetic Drugs in the Treatment of Hirschsprung's Disease," Journal of Pediatrics, 35 (1949):570.

of studies on the effect mecholyl[14] had on schizophrenic patients with constipation. The conclusions were:

> " . . . 1. mecholyl increases the tonus of the colon to spasticity and at the same time increases the motility of the colon resulting in increased evacuation rate . . . ;
>
> 2. atropine banishes any effects produced by mecholyl."[15]

Three years later Law[16] reported the successful treatment of patients with Hirschsprung's disease using mecholyl. Eight years before Law's report Cowie[17] had successfully treated a patient with acetylcholine and mecholyl. However, Law was the first to report a series of cases. In his cases the results were gratifying. He stressed " . . . habit, diet, and reasonable care . . . ,"[18] together with oil and enemas as necessary with the mecholyl for a trial of at least five to

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[14]Acetylbetamethylcholine bromide or hydrochloride.

[15]A. Myerson, Purcell G. Schube, and Max Ritvo, "Human Autonomic Pharmacology. V. - The Effect of Acetyl-beta-methylcholine (Mecholyl) on the Atonic Colon," Radiology, 28 (1937):557.

[16]John L. Law, "Treatment of Megacolon with Acetyl-betamethylcholine Bromide," American Journal of Diseases of Children, 60 (1940):267ff.

[17]Ibid., 266.

[18]Ibid., 266.

ten days in order to " . . . right an autonomic imbalance and to set up acquired reflexes . . . ."[19] Later Law[20] used prostigmine bromide to augment the mecholyl, stating that prostigmine prevented the destruction of acetylcholine by esterases.

Greenblatt[21] had good results with carbaminoylcholine, another parasympathicomimetic drug, although he failed to get good results with mecholyl, mecholyl and prostigmine, prostigmine, or urecholine. Carson[22] reported relief of symptoms with the use of urecholine, also a parasympathicomimetic drug.

An interesting observation which has been made by Peterman,[23] Greenblatt,[24] Carson,[25] Cowie,[26] and Law[27]

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[19]Ibid., 267.

[20]Ibid., 268.

[21]Jacob Greenblatt, "Carbaminoylcholine (Doryl) in the Treatment of Congenital Megacolon," Journal of Pediatrics, 31 (1947):687ff.

[22]Carson, op. cit., 570ff.

[23]Peterman, op. cit., 485ff.

[24]Greenblatt, op. cit., 688f.

[25]Carson, op. cit., 571f.

[26]Law, op. cit., 279f.

[27]Ibid., 266.

is that some of these patients do well for weeks or months after specific medication has been stopped.

The surgical approach to the treatment of congenital megacolon dates back to the latter part of the nineteenth century. In a review of the literature in 1908 the following surgical methods of treatment were presented:

1. "Intestinal puncture, either through the abdominal wall or after laparotomy." This was considered to be of an "unsurgical manner" by the author who stated that the result was ultimately unsatisfactory.

2. Exploratory laparotomy. This was done to aid in the diagnosis and offer the opportunity to "milk" the contents of the colon through the anus.

3. Colotomy. This procedure was performed in order to evacuate the contents of the bowel. The results were "fairly satisfactory."

4. Colostomy.

5. Colopexy.

6. Enteroanastimosis.

7. " . . . Procedure similar to the operation of pyloroplasty for pyloric obstruction." The results of this operation were termed "unsatisfactory."

8. Resection with subsequent enteroanastimosis. This was said to be " . . . unquestionably the operation of choice, other things being equal."[28]

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[28]Finney, op. cit., 635f.

Through the years colostomy and resection have been common surgical procedures in the treatment of megacolon. A colostomy or cecostomy has frequently been done as an emergency procedure where there were such complications as complete obstruction or respiratory embarrassment from the greatly increased contents of the abdominal cavity. It has also been used as a preliminary step to a subtotal or total colectomy or to the more recent procedure, viz., resection of the narrow rectosigmoid segment.

Ask-Upmark[29] reported 102 cases in which a partial colectomy had been done, with good results observed immediately, and it was thought at that time that the results would be permanent. Ladd and Gross[30] did partial or total colectomies in thirteen patients, but only about one-half showed satisfactory results. Dixon[31] reported good results from subtotal colectomy in six of seven cases.

In addition to the risk of this major surgical procedure, the patient faces the possibility of recurrence of

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[29]Herrmann, op. cit., 1190.

[30]Ladd and Gross, op. cit., 152ff.

[31]Claude F. Dixon, "The Management of Megacolon," Transactions of the Western Surgical Association, 48 (1938):433ff.

dilatation proximal to the point of anastimosis. This is a generally accepted fact and has been reported by several authors.[32, 33, 34]

In 1927, Wade and Royle[35] of Australia reported their results in a case treated surgically by sympathectomy. Five months after the procedure the patient was having a daily bowel movement (which he had not had for ten years prior to surgery), had a flat abdomen with no visible peristalsis, and was generally in good health. They reported, also, that less barium was needed to outline the descending and pelvic colon.

The results of this procedure--which has since been used extensively--are explained thus:

> "(1) . . . diminution of plastic tone of the colon,
>
> (2) allowing the weakened parasympathetic nerves to exert their action unopposed by the antagonistic sympathetic innervation, and
>
> (3) the release of a reflex sympathetic inhibition."[36]

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[32]Ladd and Gross, op. cit., 153.

[33]Swenson, Rheinlander, and Diamond, "Hirschsprung's Disease: A New Concept of the Etiology," op. cit., 552.

[34]R. B. Wade and Norman D. Royle, "The Operative Treatment of Hirschsprung's Disease: A New Method," Medical Journal of Australia, 1 (1927):138.

[35]Ibid., 139ff.

[36]de Takats and Biggs, op. cit., 824.

Passler[37] reviewed 117 cases in which sympathectomy had been performed. Of this number, thirty-eight cases had "complete relief," sixty-four had evidence of "improvement," twelve had received no help, and three had died. Herrmann had a series of fourteen cases, thirteen of which had "satisfactory function of the bowel."[38]

In 1931, Scott and Morton[39] suggested spinal anesthesia as a preliminary test to determine the results to be obtained by sympathectomy. Law believes, however, that it is " . . . difficult to demonstrate clinically overactivity of the sympathetic nerves. Spinal anaesthesia . . . may block the parasympathetic nerves and the interpretation is, therefore, equivocal."[40] Another limiting factor presented by Law, other than the inability to determine the prognosis following sympathectomy, is that children do poorly following the procedure if the colonic musculature has been greatly dilated and has become weak.

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[37]Herrmann, op. cit., 1183.

[38]Ibid., 1183.

[39]W. J. Merle Scott and J. J. Morton, "Sympathetic Inhibition of the Large Intestine in Hirschsprung's Disease," Journal of Clinical Investigation, 9 (1931): 247ff.

[40]Law, op. cit., 264.

In view of the work done by Kernohan and Whitehouse, by Swenson, Rheinlander, and Diamond, and by others,[41] the recent theory of etiology which states that the rectosigmoid is nonfunctioning and causes an obstruction gives a key to a more logical and definitive type of treatment, viz., resection of the narrowed, nonfunctioning rectosigmoid. In 1948, Swenson and Bill[42] reported results of fifteen experimental rectosigmoidectomies in dogs. This procedure included resection to about one and one-half centimeters above the anus and an end-to-end anastimosis done by a pull-through procedure in which the lower remaining segment was inverted and the proximal segment was pulled through until the ends were approximated and the anastimosis done. In the fourteen dogs which lived, there were no postoperative strictures and all had good sphincter control.

In August, 1949, the results of this procedure in twenty-three patients with congenital megacolon were published by Swenson, Neuhauser, and Pickett.[43] Although

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[41]See page 9 for further discussion.

[42]Swenson and Bill, "Resection of Rectum and Rectosigmoid with Preservation of the Sphincter for Benign Spastic Lesions Producing Megacolon," op. cit., 216f.

[43]Orvar Swenson, Edward B. D. Neuhauser, and Lawrence K. Pickett, "New Concepts of the Etiology, Diagnosis and Treatment of Congenital Megacolon (Hirschsprung's Disease)," Pediatrics, 4 (1949):201ff.

the youngest patient was only two months of age, there had been no deaths. Postoperatively all were having normal bowel movements without the aid of laxatives or enemas and all had normal sphincter control. Follow-up barium enema studies done on five of these patients showed that the colon was still "slightly larger than normal" but the emptying was "excellent." In fifteen of these cases a colostomy was done as a preliminary procedure because of an "advanced" stage of the disease. At the time their report was written all except two had been closed.

Bodian, Stephens, and Ward[44] have reported twelve cases in which a rectosigmoidectomy was done. In one case the colostomy had not yet been closed. In the eleven completed cases, which had been followed for lengths of time varying from one week to three months, the patients were having regular bowel movements without the symptoms of constipation or distention.

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[44]Bodian, Stephens, and Ward, "Hirschsprung's Disease and Idiopathic Megacolon," op. cit., 10.

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PART TWO

Chapter XI

REVIEW OF CASES

Review of Thirteen Cases Seen at Wisconsin General Hospital Since January, 1944

AGE AT ONSET OF SYMPTOMS:

Number of Cases	Age at Onset of Symptoms	Age at Admission
3	Birth	19 mos.; 5 yrs.; 8 yrs.
1	24 hrs.	16 days
1	Symptoms present all his life (onset, ?)	51 yrs.
1	10 days	15 yrs.
1	0-2 mos. (?)	7 yrs.
2	2 mos.	19 mos.; 9 yrs.
1	1½ yrs.	3½ yrs.
1	2 yrs.	2½ yrs.
1	40 yrs.	45 yrs.

Number of Cases

BIRTH:

8 . . . . Normal delivery (7 had normal weight; R. A. weighed 4 pounds)

1 . . . . Difficult breech, nonoperative delivery, normal weight

1 . . . . Cesarean section, normal weight

3 . . . . History not available

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DEVELOPMENT:

7 . . . . Within normal limits, i. e., according to available information or to what informant considered normal (However, R. A., age 9 yrs., at admission had a bone age of 6 yrs., 9 mos.; R. W., age 15 yrs., appeared small for his age at time of admission.)

1 . . . . Walked at age of 25 months

1 . . . . Walked at age of 29 months

4 . . . . History not available

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PAST ILLNESSES:

Past medical history was essentially noncontributory.

One child had had recurrent episodes of pneumonia and otitis media. At the time of admission he had pneumonia; and at a recent outpatient visit he gave a history of another recent pneumonic infection. This child is a diabetic.

FAMILY HISTORY:

There was no history recorded on the charts of other cases of megacolon or of other congenital anomalies in these families.

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SYMPTOMS:

All except two gave constipation or abdominal distention (or a complaint implying this) as the chief complaint or part of the chief complaint. One gave "stomach trouble" as the chief complaint; and another was admitted because an abdominal mass had been found on routine physical examination. In two cases no chief complaint per se was indicated, but constipation and distention were given as symptoms.

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PHYSICAL FINDINGS:

The most common physical finding was abdominal distention which was present in twelve of the thirteen cases reviewed. Other common findings included visible peristalsis, palpable fecal masses, and a tympanitic percussion note.

In six cases flared costal margins were recorded. It is interesting to note that the diagnosis of emphysema was made in one case. In another case a note of the increased anterior-posterior diameter of the chest was made.

In three cases hemorrhoids were present at the time of admission or there was a history of hemorrhoids.

One patient had varicosities of the lower extremities with swelling of the left leg on one admission and an ulcer of the left leg on another admission. He had had a saphenous ligation.

During a proctoscopic examination on one patient, an area of contraction was encountered (just distal to the dilated portion) which relaxed after about three minutes to admit the proctoscope.

OTHER CONGENITAL ANOMALIES:

In only one patient were there other anomalies of development. In this patient there was a history of an imperforate anus, findings of a megacolon with nonrotation of the colon, and an absent right testicle.

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Number of Cases

LABORATORY FINDINGS:

3 . . . . Hypochromic anemia

1 . . . . Occult blood in stools (i. e., in stools of one of the above patients with hypochromic anemia)

1 . . . . Diabetic

- - - - - - - - - - -

X-RAY FINDINGS:

3 . . . . Dilatation included the rectum

8 . . . . Dilatation did not include the rectum but started in the sigmoid and extended proximally

2 . . . . Exact extent not indicated

This differentiation is interesting because of the recent reports of rectosigmoidectomies done in cases where the rectum and lower sigmoid were apparently of normal caliber.

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TREATMENT:

Specific Methods or Agents

1. DHO 180 (dihydroergocornine) - sympathicolytic
2. Priscoline - sympathicolytic
3. Syntropan - atropine-like
4. Mecholyl - parasympathicomimetic
5. Demerol - analgesic, relaxer of spastic smooth muscle
6. Prostigmine - cholinesterase inhibitor
7. Sympathetic block
8. Spinal anesthesia

Results of Specific Medical Therapy

DHO 180 - K. P. -- Two trials without benefit
M. B. -- One trial without benefit
J. L. -- One trial without benefit
R. A. -- Spontaneous bowel movements-- medication discontinued after about a month (August, 1949) and patient has continued to have a bowel movement about every four or five days the same as when taking the drug

DHO 180 and Mecholyl - M. B. -- One trial of these two drugs combined without benefit

Priscoline - K. P. -- One trial without benefit

Syntropan - K. P. -- Two trials without benefit
M. B. -- Two trials without benefit

TREATMENT:

Results of Specific Medical Therapy (continued)

Mecholyl - G. S. -- One trial without benefit
K. P. -- One trial with slight improvement but of insufficent magnitude to warrant continuance
M. B. -- Expired before adequate dosage was reached
L. D. -- Discharged in February, 1949, on dose of mecholyl which had started spontaneous bowel movements--duration of medication unknown but with no medication is now having a bowel movement every two or three days with "very little" distention
P. U. -- Mecholyl taken from July, 1944 to July, 1949 with apparent good health and spontaneous bowel movements--expired suddenly in July, 1949 with post-mortem diagnosis of volvulus

Demerol - K. P. -- Relief of symptoms for two and one-half months when distention returned

Prostigmine - G. S. -- Spontaneous bowel movements since started on prostigmine in February, 1949
K. P. -- Benefit during hospitalizzation but none when discharged and when prostigmine was taken only about two times weekly instead of daily
R. W. -- Spontaneous stools since started on this medication in December, 1949

TREATMENT:

Results of Specific Medical Therapy

Prostigmine - (continued)

M. R. -- Discharged in October, 1947, having spontaneous stools--parents stopped medication after seven months and patient now has daily bowel movement with only occasional enema

L. D. -- One trial without benefit

M. B. -- One trial without benefit

Mecholyl and Prostigmine - J. L. -- One trial without benefit

Sympathetic block - K. P. -- Two blocks without benefit

M. B. -- One block without benefit

J. L. -- One block without benefit

Spinal anesthesia - K. P. -- Three trials without benefit

J. L. -- Spontaneous bowel movements over period of three months with only occasional enema following first anesthetic--no benefit following the next two spinal anesthetics

DEATHS:

P. U. -- Expired suddenly after having good results from the use of mecholyl over period of five years. The diagnosis of volvulus was established with a post-mortem examination.

M. B. -- Expired suddenly in a shock-like picture following an enema. There was no anatomical explanation at the post-mortem examination.

Chapter XII

CASE HISTORIES

Case One

K. P., 235976, white male, age 8 years, was admitted for the first time on January 28, 1944 and discharged on April 2, 1944. The chief complaint was enlargement of the abdomen. He had been constipated since birth and abdominal distention had begun shortly after birth and had been progressive up to the time of admission. Peristalic movements had become visible when the patient was about fifteen months of age. About this same time he had begun to vomit and the diagnosis of intestinal obstruction was made. At surgery a congenital megacolon was diagnosed. Enemas were necessary for all bowel movements until the "last few years" when they had been needed less frequently. At the time of admission he had a bowel movement about once daily, and they often occurred after the insertion of the tip of a rectal tube. The stools were hard and small and without blood. He passed much flatus per rectum. He occasionally had pain when distention was increased. Past medical history revealed a normal delivery with a birth weight of 6 pounds. His

development had been considered normal. At the time of admission he was doing satisfactorily in the third grade. He had had measles and chicken pox. Two other children in the family were living and well. Physical examination revealed a malnourished child with the important findings limited to the abdomen. The abdomen was greatly distended with a right longitudinal scar, tightly stretched skin, and poor musculature. The costal margins were flared and the breathing was mainly thoracic. Peristalsis became visible after enemas. The percussion note was tympanitic and tympany extended into the lower chest. The circumference of the abdomen was 75.5 centimeters above the umbilicus and at the costal margins, 78.5 centimeters. One examiner measured the pattern of the colon on the abdominal wall and gave the diameter as 4.5 centimeters. Repeated urinalyses were essentially negative. On admission he had 14.1 grams of hemoglobin and 4.35 million red blood cells. Later he developed a hypochromic anemia with 12.9 grams of hemoglobin. An X-ray of the abdomen revealed a greatly dilated colon filled with impacted fecal material and gas. The barium enema studies revealed the colon to be widely dilated proximal to the lower half of the sigmoid. The rectum and lower sigmoid were approximately of normal caliber. The abdomen seemed

to be filled completely with the colon causing distention of the abdomen and elevation of the diaphragm. His treatment included enemas, laxatives, fruit with each meal, a transfusion of 200 cubic centimeters of whole blood, and specific medication. On January 21, he was started on mecholyl--50 milligrams at 9 a.m. This was gradually increased until by February 15 he was receiving 200 milligrams twice daily. There was slight improvement on this, but he failed to show satisfactory progress. From March 5 to March 14, he received 25 milligrams of syntropan three times daily, but his condition apparently became worse. He needed daily enemas, began to have a slight elevation of temperature, and complained of abdominal pain. From March 16 to March 23, he received sulfasuxidine to reduce the intestinal flora. On March 23, demerol, 25 milligrams every four hours, was started. Almost immediately the pain stopped and the distention decreased. An upper respiratory infection and otitis media which occurred during this admission were treated with sulfadiazine and symptomatically. He was discharged on 25 milligrams of demerol four times daily, sulfadiazine in decreasing doses, and vitamins.

The second admission was from June 17, 1945 to June 27, 1945. The patient had continued on demerol

as prescribed until July, 1944, when abdominal distention recurred. A series of four injections of vitamin B had caused marked decrease in distention and he remained well until one month before this admission when injections of vitamin B were again started. One to three enemas were required daily at the time of admission. His appetite was good. In May, 1944, he had had jaundice which had been considered to be on an infectious basis. The physical examination was essentially as before with malnourishment, abdominal distention, poor abdominal musculature, visible peristalsis, tympany, and elevation of the diaphragm. Breathing was mainly thoracic. Cardiac premature contractions were heard as on the previous admission. The circumference at the umbilicus was 65 centimeters and at the costal margin, 71 centimeters. The urinalysis was negative. Hemoglobin was 13.93 grams and there were 5.21 million red blood cells. The barium enema studies showed some improvement in the proximal half of the colon which was less dilated than before. The specific medication during this admission was prostigmine. The first dosage was 15 milligrams three times daily. With this amount he had spontaneous bowel movements but complained of abdominal pain; consequently, the dose was cut to 3/4 of a pill (11.25 milligrams). On this he continued

to have spontaneous stools, and so he was discharged on this medication and vitamins.

He was admitted for the third time on July 13, 1948 and discharged on August 1, 1948. His condition at this time was essentially unchanged as compared with that seen on previous admissions. He still required one or more enemas daily and had abdominal distention. He had been taking prostigmine only about two times weekly without apparent benefit. In general his health had been good except for a poor appetite and a slow gain in weight. The physical examination revealed essentially the same as previously noted. The circumference at the umbilicus was 77 centimeters and at the costal margins, 82 centimeters. Laboratory examinations were within normal limits. The barium enema studies were reported as showing progressive dilatation. He was started on prostigmine at the time of admission (10 milligrams three times daily) but this was stopped and syntropan was started on July 15. He was started on 12.5 milligrams three times daily, and this was increased to 25 milligrams three times daily on the next day. Another approach was tried when a left lumbar sympathetic block was done July 19. This caused the passage of some flatus and increase in peristalsis. The block was repeated the next day. On July 21, a spinal anesthetic was administered and

anesthesia reached the level of the sixth thoracic vertebra. On July 20, DHO 180 was ordered--1 milligram three times daily after meals--and two days later this was increased to 2 milligrams a dose. DHO 180 was stopped on July 24. Syntropan, 37.5 milligrams three times daily after meals, was ordered July 22. By July 28, the dose had reached 100 milligrams. There was no apparent benefit from these, so the patient was discharged to wait for the late effects of spinal anesthesia. The patient was to return in six weeks if no improvement was evident.

The patient returned September 20, 1948 and was discharged October 12, 1948. For two days after discharge in August he had had spontaneous stools, but since that daily enemas had been required. The physical findings were essentially as before. One examiner noted that he had emphysema. Laboratory work was negative. No barium enema studies were done. On September 22, he was given two intramuscular injections of DHO 180, a total of 0.15 milligram. One intramuscular dose was given on the next day--0.1 milligram. On September 24, another spinal anesthetic was given to the level of the ninth thoracic vertebra with no increase in peristalsis and no evacuation. On September 27, he was given

DHO 180 by the intravenous route. A total of 0.6 milligram was given. Subjectively he experienced nausea and cramps. Objectively there was increased peristalsis. On October 10, another spinal anesthetic was given to the level of the fourth thoracic vertebra, and there was marked increase in peristalsis but no evacuation. As there still was no apparent improvement, he was discharged to return later for contemplated surgery. He was to have daily enemas, cooked fruit, and vitamins.

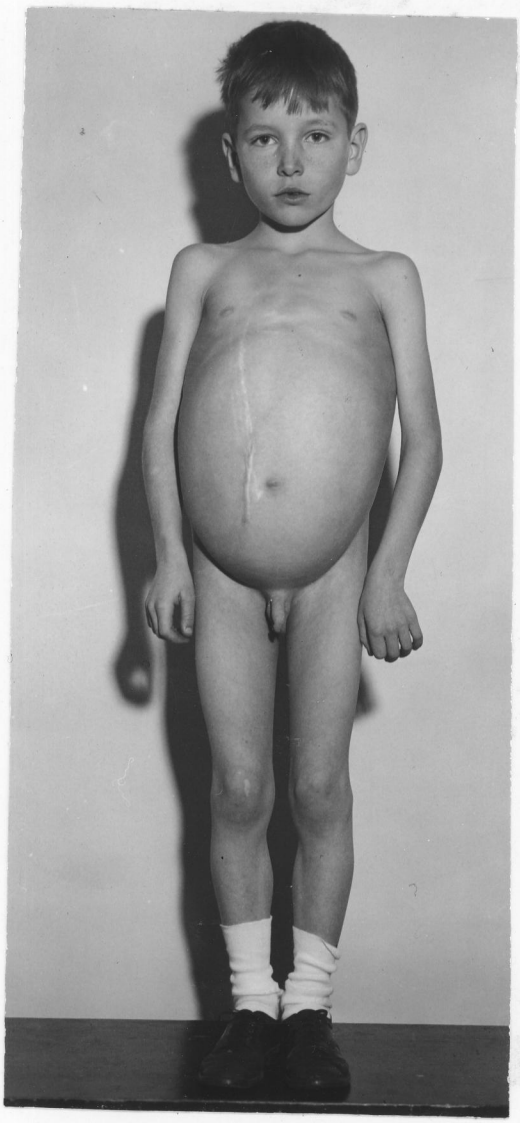
He returned June 8, 1949 and was discharged July 1, 1949. The patient presented complaints similar to those given before. He had needed fewer enemas since the last admission and had felt better. He was now in the eighth grade and was a good student. Physical findings were essentially unchanged. Abdominal circumference at the umbilicus was 74 centimeters. Laboratory work was within normal limits. A flat plate of the abdomen showed a large amount of gas and fecal material and probably some increase in the size of the colon. It was thought that surgery should not yet be done; consequently, he was started on 12.5 milligrams of priscoline three times daily before meals, on June 24. On June 28, the dose was increased to 25 milligrams. No spontaneous bowel

movements occurred. His care had included the usual cooked fruit, enemas, mineral oil, and vitamins. On discharge he was to receive daily enemas but no other medication.

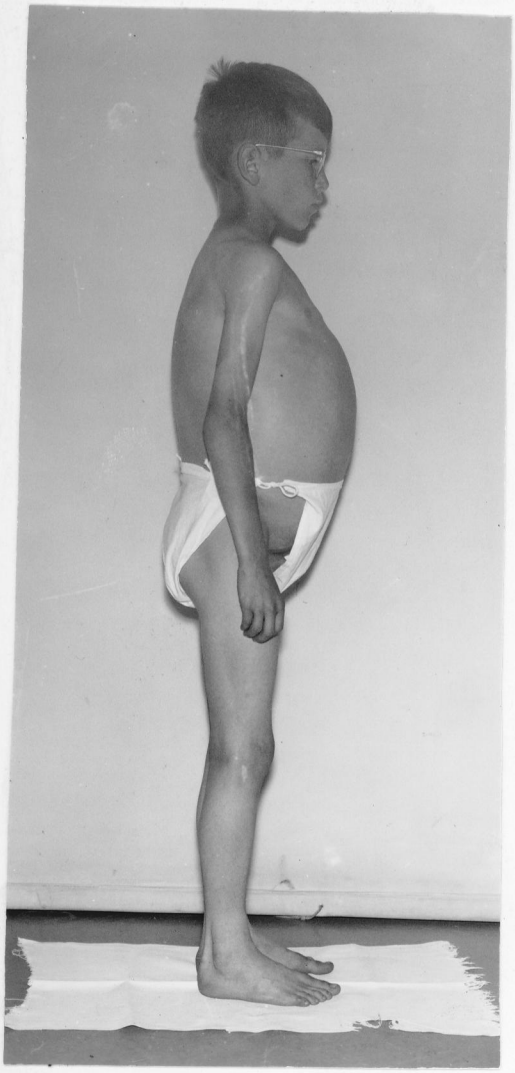
The sixth admission was on September 29, 1949 and he was discharged on October 30, 1949. He had felt well and had had a daily enema but no other medication. The examination was essentially as before. Laboratory work was negative. The introduction of a small amount of barium showed that the distal sigmoid and rectum were not involved in the megacolon. A chest X-ray showed elevation of the diaphragm. At this time he was prepared for surgery by enemas, castor oil, and a low residue diet. He was to have a colostomy preliminary to a rectosigmoidectomy, but was discharged to return after the first of the year because of the illness of the surgeon who was to do the operation.

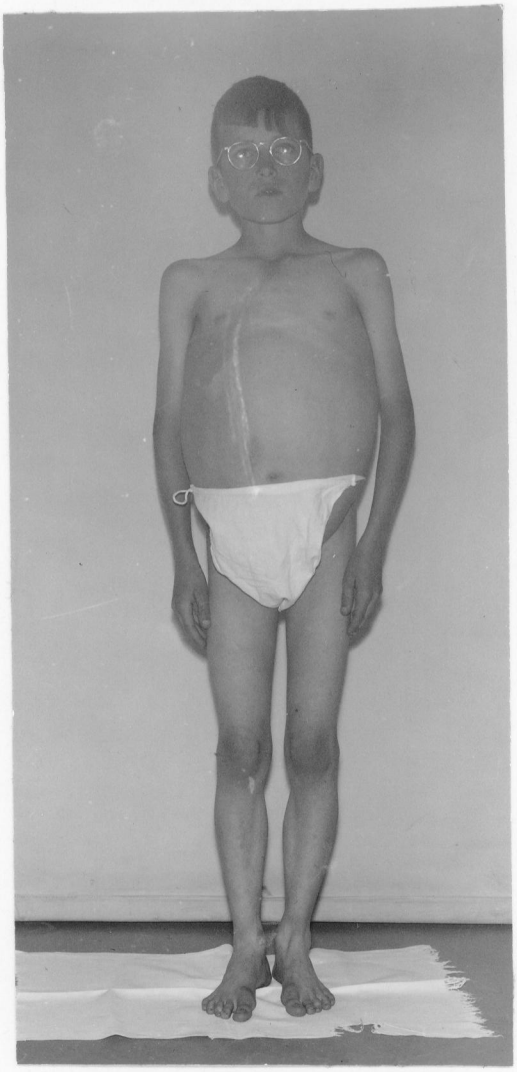
The last admission was from January 2, 1950 to January 29, 1950. He had felt well, used daily enemas as before, but had had no spontaneous bowel movements. The examination was as before. The laboratory work was negative. A flat plate of the abdomen showed tremendous gaseous distention of the colon and a large fecal impaction occupying the entire pelvis and extending along

the left side into the upper third of the abdomen. Extensive cleansing enemas and retention enemas were given. A transverse colostomy was done and the cleansing process continued. He was discharged to return for the rectosigmoidectomy and was to have rectal enemas three times weekly and daily irrigations of the proximal loop.

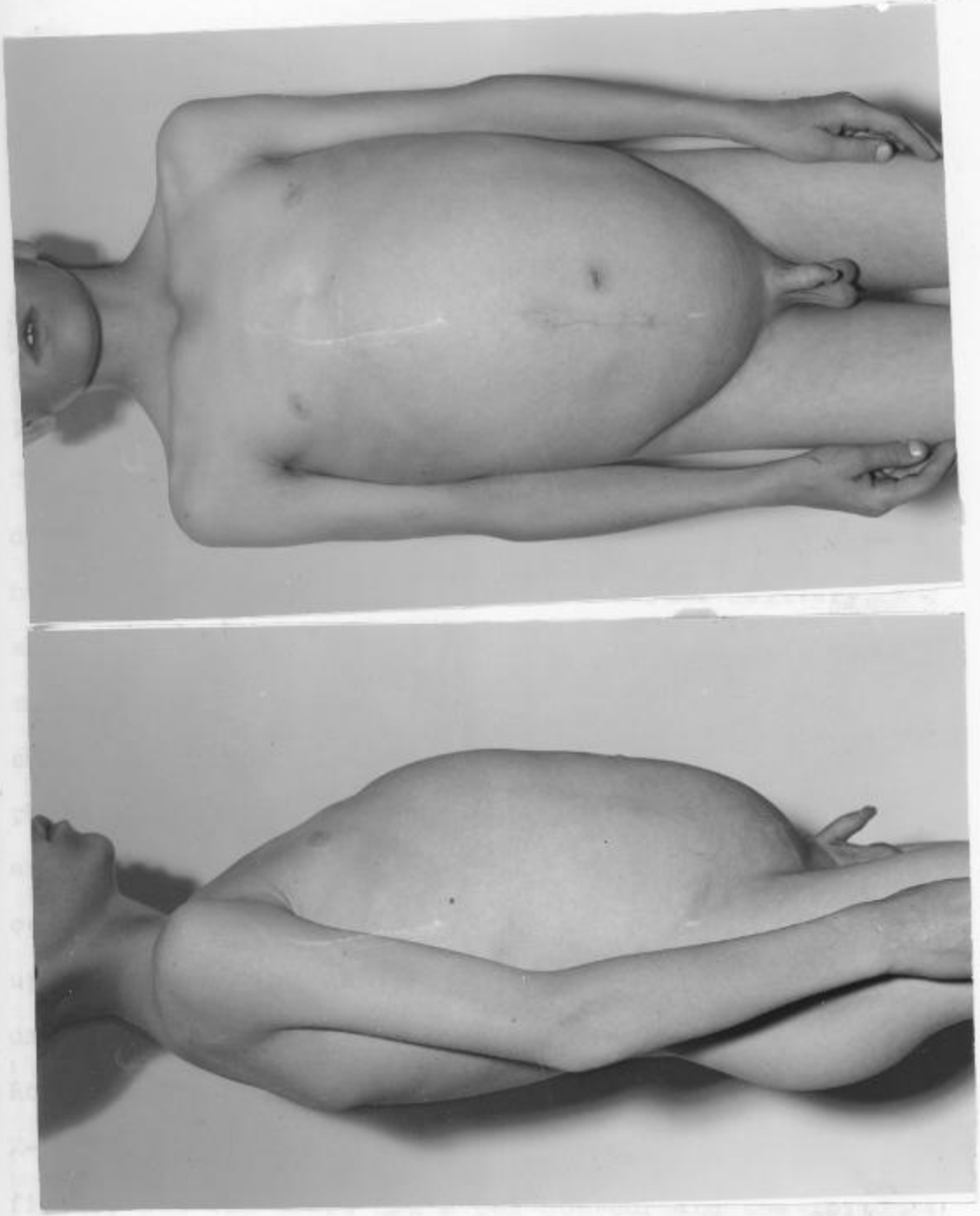


K. P., 235976. Taken on January 30, 1944.





K. P., 235976. Taken on July 13, 1948.



K. P., 235976. Taken on June 8, 1949.

Case Two

P. U., 238411, white female, age 7 years, was admitted July 7, 1944 and was discharged July 29, 1944. Her chief complaint was abdominal distention. She presented symptoms of constipation since infancy, recurrent episodes of distention since age three, periods of diarrhea, flatus per rectum, and belching. Three weeks prior to admission she had had emesis and dyspnea associated with distention. Her past medical history was not contributory with a normal delivery, a birth weight of 7 pounds, 8 ounces, normal development, and the usual childhood diseases. Two other children in the family were in good health. Positive physical findings were limited to the abdomen and chest except for the general picture of malnutrition. The costal margins were flared, the heart was displaced upward, the abdomen was protruding with an everted umbilicus, and peristalic movements were visible. Routine laboratory studies were within normal limits. X-ray of the chest revealed good areation of the lung fields, but the heart had a box contour and the left leaf of the diaphragm was elevated. Barium enema studies showed the entire colon to be dilated from the

cecum through the proximal half of the sigmoid. The terminal ileum also was dilated. The patient was started on 50 milligrams of mecholyl three times daily, but following the third dose the patient had cramps, emesis, sweating, and prostration which was relieved by atropine sulfate subcutaneously, grain 1/150. The dose was reduced to 50 milligrams two times daily for eight days and then increased again to three times daily with no untoward effects. Spontaneous bowel movements started the second day and continued until discharge. She was discharged on 50 milligrams of mecholyl three times daily, with the note that she was "markedly improved, less distention, generally improved."

An outpatient note made October 20, 1944 revealed that she was feeling well with a good appetite and three soft bowel movements daily. The heart was still displaced and the upper abdomen was prominent. There was tympany over the area of the descending and transverse colon. No peristalic waves were seen. She was to continue on mecholyl, 50 milligrams three times daily.

Correspondence in February, 1950 revealed that the patient had expired suddenly on July 11, 1949. She "had seemed so much better" during the previous one and

one-half years. She had continued on mecholyl. She had still been bothered by flatus--"depending on what she ate"--which would be relieved by emesis. Her appetite had been good and she had eaten "lots of fruit and juices." There had been no other illnesses and no additional therapy for her primary disease. Her school work had been good. Her local doctor reported in March, 1950 that "she was apparently well and active until the night of July 10, 1944 when she was awakened with severe abdominal pain which was markedly progressive . . . went into shock . . . became unconscious within a couple of hours and died within a few hours . . . ."

The post-mortem findings included the following: "Absence of normal anchorage of colon in region of cecum and hepatic and splenic flexures; marked volvulus of large and small intestines; acute mesenteric lymphadenitis; emaciation." Microscopic examination showed "marked infiltration of lipoid material in practically every cell" of the liver.

Case Three

M. B., 249664, white male, age 16 days, was admitted August 10, 1946 and was discharged October 27, 1946. The chief complaint was abdominal distention and the other symptoms presented were diarrhea and emesis. The onset had been twenty-four hours after birth. Some relief of symptoms had been obtained with the use of a high rectal tube. The delivery had been normal with a birth weight of 8 pounds, 11 1/4 ounces. There were five other children, but the state of their health was not recorded. Physical examination revealed a state of malnutrition with depression of the fontanelles. The tympanic membranes and the pharynx were congested. The abdomen was protuberant. On rectal examination there was impaction of feces. The laboratory work was essentially negative. A chest X-ray report stated that the heart was globular with questionable enlargement. The report of the barium enema stated that the colon was somewhat enlarged in caliber and distended with gas. His treatment included parenteral fluids, 560 cubic centimeters of whole blood given in six transfusions, 100 cubic centimeters of plasma, vitamins, atropine before feedings, use of the rectal

tube, etc. He gained weight on the hospital schedule and was discharged, being put on a formula suitable for his age, cream of wheat, apple sauce, and canned pears.

The second admission was from June 17, 1948 to August 17, 1948 when the child was about two years old. His symptoms included abdominal distention as before, constipation, lassitude, and poor appetite. The only significant physical finding was the abdominal distention. Barium enema showed increased involvement of the colon with dilatation extending from the upper sigmoid region through the cecum. Various drugs were tried. On June 18, prostigmine, 5 milligrams three times daily, was started. This was continued until June 23 when prostigmine was stopped and syntropan was started. The dose of syntropan was gradually increased from 5 milligrams three times daily to 50 milligrams three times daily by July 27. As there were no favorable results, syntropan was stopped August 2. On August 3, DHO 180 was started. The first day he was given 0.5 milligram three times daily after meals. The dose was increased gradually to 4 milligrams three times daily after meals by August 9. On August 11, mecholyl was started in addition to the DHO 180 and 20 milligrams

of mecholyl was given with each dose of DHO 180. A sympathetic block of L2 and L3 was done on August 10. His regime included a low residue soft diet, cooked fruit three times daily, vitamins, laxatives, and enemas as needed. There was no response to any of the specific agents used. His discharge orders were: enemas as necessary, vitamins, and neocultol.

The third admission was from June 28, 1949 to August 23, 1949 at the age of three years. In addition to the distention and constipation, he had dyspnea, anorexia, and emesis. Since the previous admission, he had had daily enemas except for one week in October, 1948 when he had daily spontaneous bowel movements. Physical examination this time revealed a malnourished child with flared costal margins and a distended abdomen with visible peristalic movements. The percussion note was tympanitic. There were impacted feces in the rectum. A chest X-ray revealed an elevated diaphragm. The barium enema studies showed "megacolon--severe." The patient was started on DHO 180 on July 12--3 milligrams three times daily after meals. The dose was increased gradually to 5 milligrams three times daily after meals. On July 15, DHO 180 was stopped and syntropan was started. Syntropan was given until August 2 during

which time the dose was increased from 5 to 50 milligrams three times daily. Prostigmine was tried from August 2 to August 11; the dosage was started at 5 milligrams three times daily and increased to 10 milligrams three times daily. The next drug to be tried was mecholyl. This was started at 20 milligrams three times daily and was being increased. Before adequate doses of mecholyl were reached, however, the child expired in a shock-like picture following an enema. He had had similar reactions after two other enemas but had responded to treatment for shock. A note on the chart ". . . postulated that the stimulation from the enemas may have upset the balance of the autonomic nervous system with resultant shock." The regime during this admission had included a general diet with cooked fruit, vitamins, and enemas as necessary.

The post-mortem findings of interest were the greatly dilated ascending, transverse, and descending colon, and pulmonary congestion. The lower half of the sigmoid and rectum were of approximately normal caliber. The death of the patient could not be explained by the pathological findings.

Case Four

J. L., 251869, white male, age 5 years, was admitted on January 15, 1947 and was discharged February 20, 1947. The chief complaint was "can't move bowels." An enema given every other day since the baby had been taken home from the hospital after birth had controlled the symptoms until December, 1946. He now had distention, loss of appetite, belching, emesis if no bowel movement, weakness, and cramps in the abdomen. The past history revealed that his delivery had been normal with a birth weight of 8 pounds, 4 ounces. The only previous illness was a questionable case of chicken pox. He had not walked until 29 months of age. Two other children in the family were living and well. On physical examination the child was malnourished. The abdomen was prominent with dilated veins over the surface. Peristalsis was visible and the enlarged colon was palpable. The percussion note was tympanitic and bowel sounds were increased. X-ray studies revealed elevation of the diaphragm on the right and gaseous distention of the entire colon except the rectum. Before admission here, he had received a transfusion of 250 cubic centimeters of blood. His regime

here included cooked fruit in his diet, one-half glass of salt water before breakfast, vitamins, enemas, milk of magnesia, and mineral oil. From January 15 to January 22 he received 5 milligrams of prostigmine three times daily. In addition to this he received 20 milligrams of mecholyl with each dose of prostigmine on January 15, 40 milligrams with each dose on January 16, and 50 milligrams of mecholyl with each dose of prostimine from January 17 to January 22. On January 22, he was started on syntropan which was continued to February 10. The initial dose was 12.5 milligrams three times daily, which had been increased to 37.5 milligrams three times daily by February 4. Spinal anesthesia was carried out to above the nipple line. There was no beneficial effect apparent from any of the above specific measures, and his discharge instructions included daily irrigation, vitamins, milk of magnesia, and mineral oil.

The second admission was from June 2, 1947 to June 4, 1947. The prescribed instructions had been followed, and he was now described as being "completely well." He had had one to two bowel movements daily with a weekly enema. For the week prior to admission two to three enemas had been required. The abdomen was still moderately enlarged and the costal margins were flared. The

enlarged colon was palpable with visible peristalsis. The rectum contained impacted feces. Barium enema studies were essentially as before. The child had had a recent fracture of the medial condyle of the left humerus which occurred in a fall from a porch. This was visualized on X-ray and appropriate treatment carried out. Spinal anesthesia to the nipple line was repeated during this admission and he was discharged with a return date.

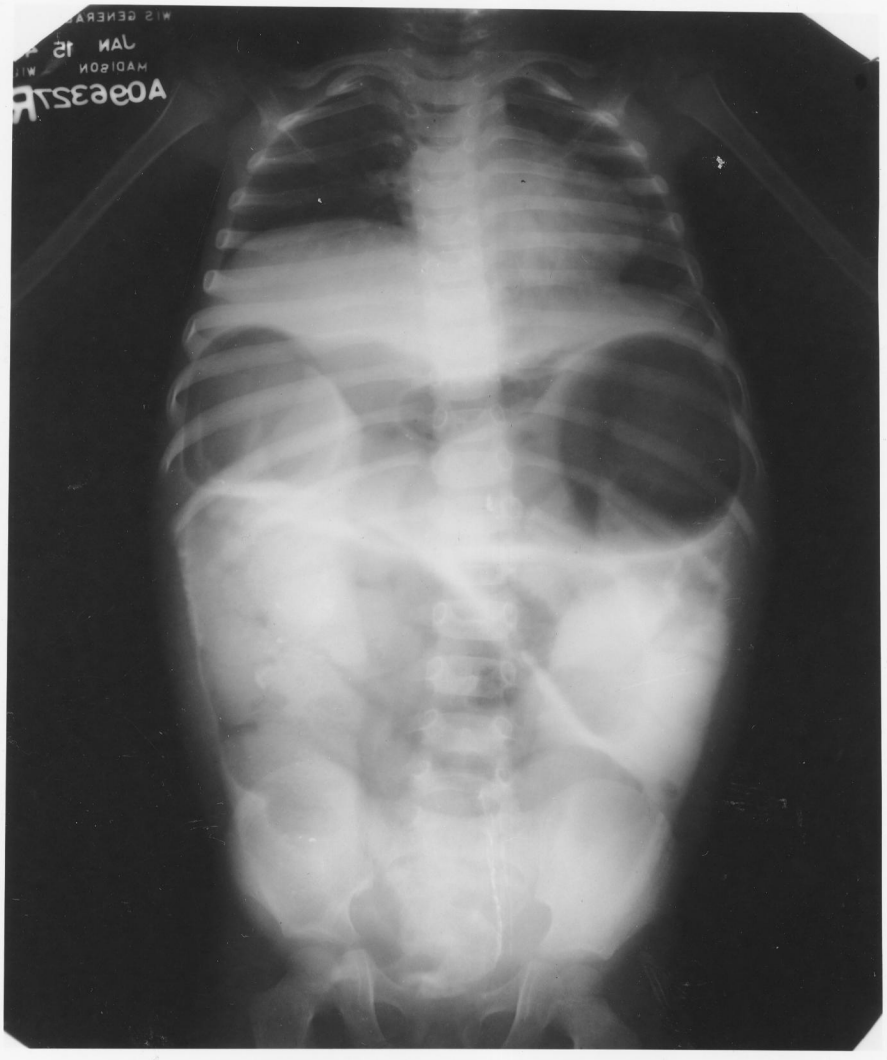
The third admission was from November 3, 1947 to November 7, 1947. There had been no spontaneous bowel movements, but with enemas every two to three days he had remained energetic and had a good appetite. The abdominal distention now was only "slight" but peristalsis was still visible and the enlarged colon palpable. After X-ray the colon was described as being "tremendously dilated throughout" proximal to the rectosigmoid junction. He was discharged after cleansing enemas and a left lumbar sympathetic block of L2 and L3. He was to continue at home with the same regime as before.

The last admission was about one year later--November 8, 1948 to November 16, 1948. His health was "excellent" and he was gaining, but still he had had no

spontaneous bowel movements. An irrigation with about a gallon of water was necessary two to three times per week. Physical examination revealed the following: no abdominal distention, slight diastasis recti and eversion of the umbilicus, and visible peristalsis. The colon was not palpable but the examination had been done following an enema. The child had a deformity of the left arm from the fracture mentioned above with only 90 degrees of flexion possible at the elbow and an ulnar deviation. The report of the barium enema was essentially as before. His regime included the usual cooked fruit, vitamins, and enemas. From November 9 through November 14, he received DHO 180. Over the first three days the dose was increased from 1 to 3 milligrams three times daily before meals. On two days he was given DHO 180 intravenously under careful observation. With 0.4 milligram intravenously, marked peristalsis was visible. About an hour after this procedure, he had an emesis. A second time he was given 0.375 milligram of DHO 180 intravenously followed after a short time with 0.5 milligram of dihydroergotamine intravenously. The DHO 180 elicited some peristalsis. Results of an enema following this procedure were "fair." During this admission spinal anesthesia was

again carried out to about the level of the third thoracic vertebra. He was discharged to return for contemplated surgery.

Correspondence in February, 1950 revealed that the boy now was 48 inches tall and weighed 56 pounds. There had been no serious illnesses, his appetite was good, and his school work was average. He still had no spontaneous bowel movements and a "complete syringing" was done two times a week with results "mostly very good." No laxatives had been used, and there had been no further treatment.



J. L., 251869. Flat plate showing gaseous distention of a megacolon together with elevation of the diaphragm and displacement of the heart.

Case Five

M. R., 255614, white female, age 19 months, was admitted on September 27, 1947 and was discharged October 8, 1947. Her chief complaints were vomiting and constipation. She had been constipated since the age of two months and had required frequent enemas. During the month prior to admission lassitude and abdominal distention had been noted. She had had a three-day episode of diarrhea a month previously and for the past two weeks had vomited about every other day. The past medical history was not contributory. Her birth weight was 7 pounds, 15 ounces, and she had been a normal breech delivery. At one year of age she walked, and at the time of admission she was able to say a few words. One other child in the family was in good health. The physical examination was negative except for the general picture of malnutrition and the abdominal findings. The abdomen was moderately distended, had a small umbilical hernia, and was tympanitic to percussion. Laboratory studies revealed a hypochromic anemia with 9.1 grams of hemoglobin and 4.8 million red blood cells. X-ray studies showed a "moderate" megacolon. Retrograde flow was obstructed at the distal

end of the transverse colon, but the part visualized was "somewhat distended and redundant." The patient was given cooked fruit in her diet, vitamins, mineral oil, enemas as needed, and started on prostigmine, 5.0 milligrams three times daily, which was increased to 7.5 milligrams three times daily after two days. She was discharged having daily spontaneous bowel movements and was to continue on the same dose of prostigmine, daily laxative, enemas as needed, cooked fruit three times daily, warm water before breakfast, and vitamins.

She was seen as an outpatient on January 2, 1947. Her appetite was fair and she was having two to three stools daily. She was to continue on 7.5 milligrams of prostigmine.

She was again seen in the outpatient department on February 20, 1948. She was having one to two bowel movements daily and was to continue on the same dose of prostigmine.

Correspondence in February, 1950 reveals that the patient is active and peppy. She is having daily, yellowish stools. She has had no laxative for two months, but an enema is necessary every ten to thirty days. There is no history of abdominal distention

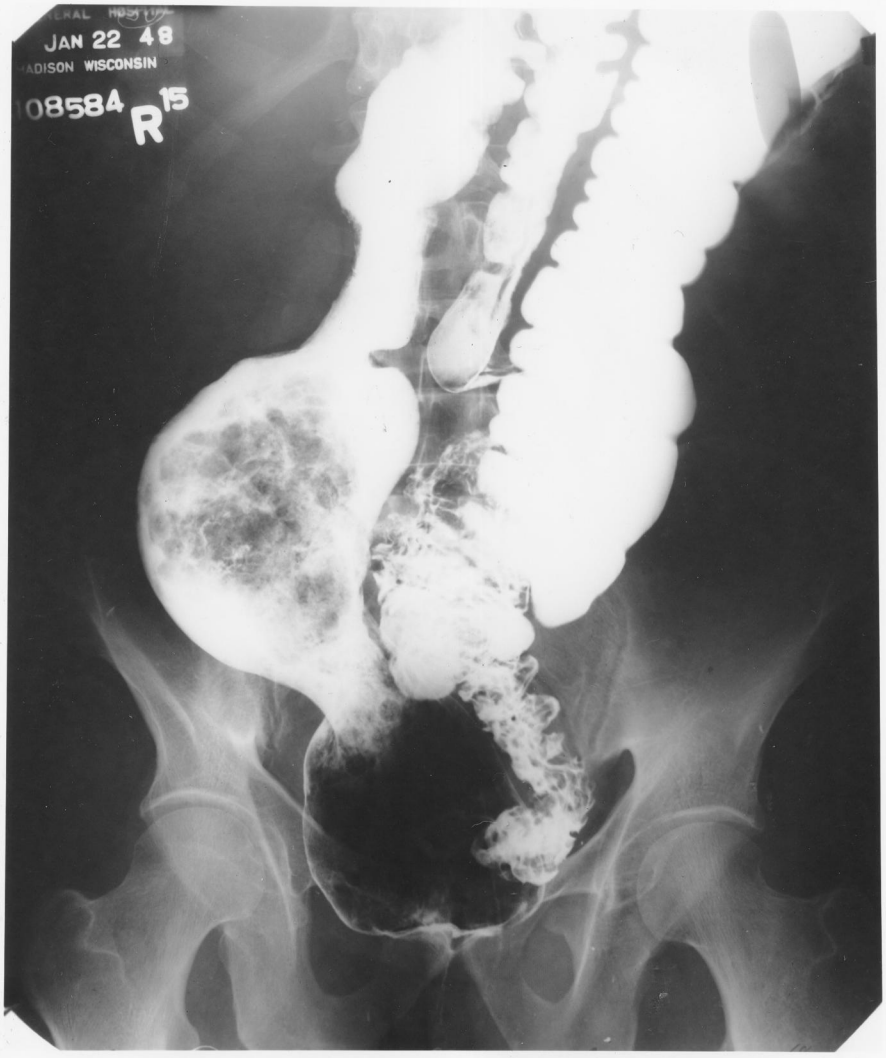
or emesis. She "seldom" has diarrhea. The prostigmine was stopped after seven months of treatment and she received about ten treatments from a chiropractor "which were very successful." In February, 1949, she had an episode of increased constipation which was cleared in about a month by using malt extract, fruits, "light" food, and daily enemas for a period of two weeks. She is 36 inches in height and weighs 33 1/2 pounds.

Case Six

D. F., 257149, white male, age 45 years, was admitted January 18, 1948 and was discharged January 29, 1948. His chief complaint was that he had gas and indigestion. His present trouble started five years before admission. He had had increasing flatulence and his history included intolerance to fatty foods and occasional right upper quadrant pain. Three years before admission he had had episodes of hematemesis. "Chronic" constipation during the past five years had required him to take frequent laxatives. His appetite was good, but he had lost 20 pounds during the previous eighteen months. He complained of weakness and easy fatigability. His past medical history revealed that he had had measles, chicken pox, whooping cough, mumps, smallpox, and allergies. He stated that he had had a plastic repair of an imperforate anus as an infant and again in 1926. Of his seven siblings, one had had jaundice, and one had died of diphtheria. On physical examination the findings included the following: a funnel chest, absence of the right testicle, poor sphincter tone on rectal examination, hemorrhoids, and impacted fecal material in the rectum. Routine

laboratory work was within normal limits. A chest X-ray was negative. The cholecystogram was negative. Barium enema studies revealed the sigmoid, descending, and transverse colon to have a large diameter. The cecum was in the left pelvis. The impression was that this patient had a megacolon and nonrotation of the colon. During his hospitalization he received laxatives and enemas as needed. He was given one dose of prostigmine (30 milligrams). He developed a fecal impaction after the barium enema which was relieved with the aid of an oil enema. On discharge he was advised to try to maintain a normal bulk in the stool with the use of metamucil.

Correspondence in February, 1950 revealed that this patient was having a daily bowel movement with the aid of one tablespoon of mineral oil taken every other day. He had gained weight and had a good appetite. There was no history of emesis or right upper quadrant pain. He had gas if he ate "acid food." No enemas had been needed. A "checking over" at the Mayo Clinic had again revealed the nonrotation of the colon.



D. F., 257149. Barium enema showing megacolon and and nonrotation of the colon. The cecum is in the left pelvis. Example of megacolon in an adult.

Case Seven

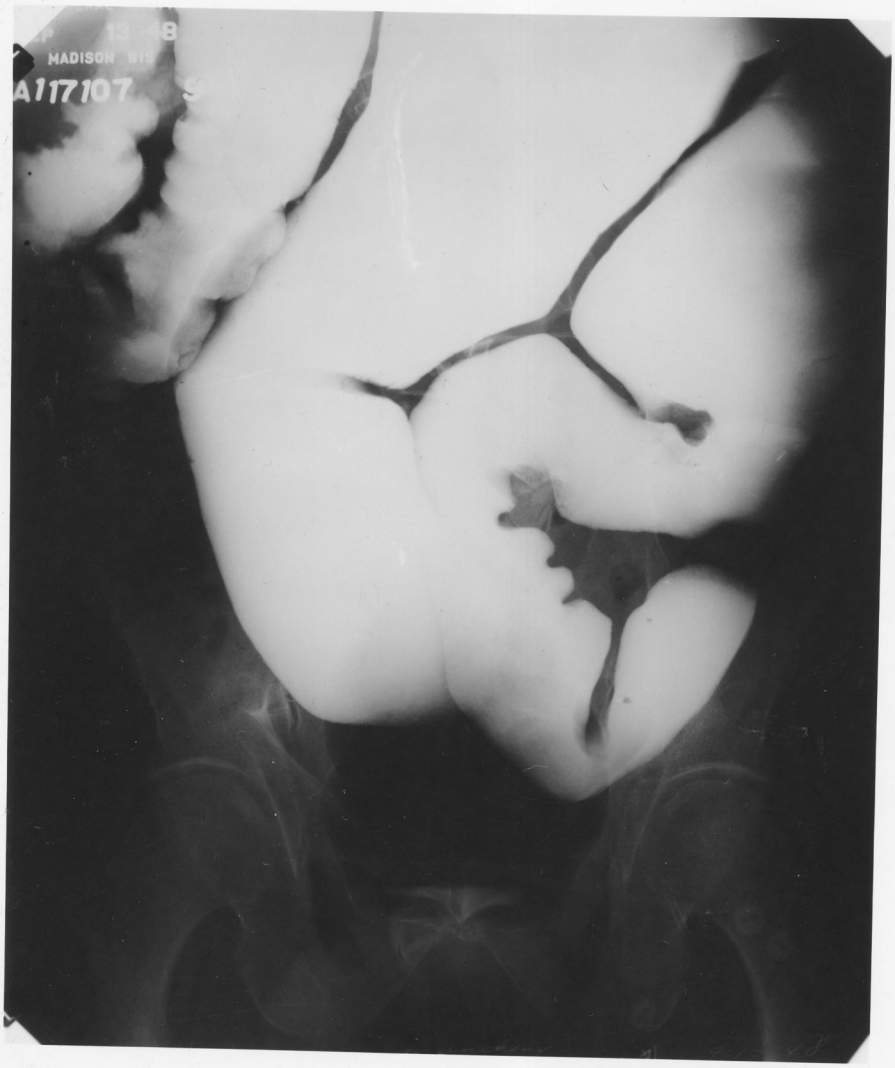
A. A., 157426, white male, age 51, was admitted September 8, 1948 and was discharged September 15, 1948. His complaints were abdominal distention and gas. He had had trouble with constipation and a certain amount of distention all his life. For the past two to three years he had used almost daily enemas and for several hours following an enema he was incontinent. His appetite was good but he had lost 25 pounds. The past medical history revealed that he had had the "usual childhood diseases," a bilateral saphenous vein ligation, and incision of a thrombosed hemorrhoid. As a boy he had been in bed for eight weeks with a back injury. His parents had died of "old age." One brother was living and well. On physical examination the positive findings included a blood pressure of 150/90 millimeters of mercury, an increased anterior-posterior diameter of the chest, dorsal scoliosis, marked distention of the abdomen with a tympanitic percussion note, increased bowel sounds, a loose rectal sphincter with a few skin tags, grade one benign prostatic hypertrophy, and varicosities of the lower extremities with edema of the left leg. The routine laboratory examinations were within normal limits. The barium enema studies revealed

that the sigmoid was four to five inches in diameter. The rectum, descending, transverse, and ascending colon were all dilated, the dilatation being most marked on the left. The treatment included a high protein, high carbohydrate, low residue diet, an oil retention enema nightly, and two ounces of castor oil nightly. He was discharged with instructions to continue on a low residue diet and take two ounces of castor oil nightly.

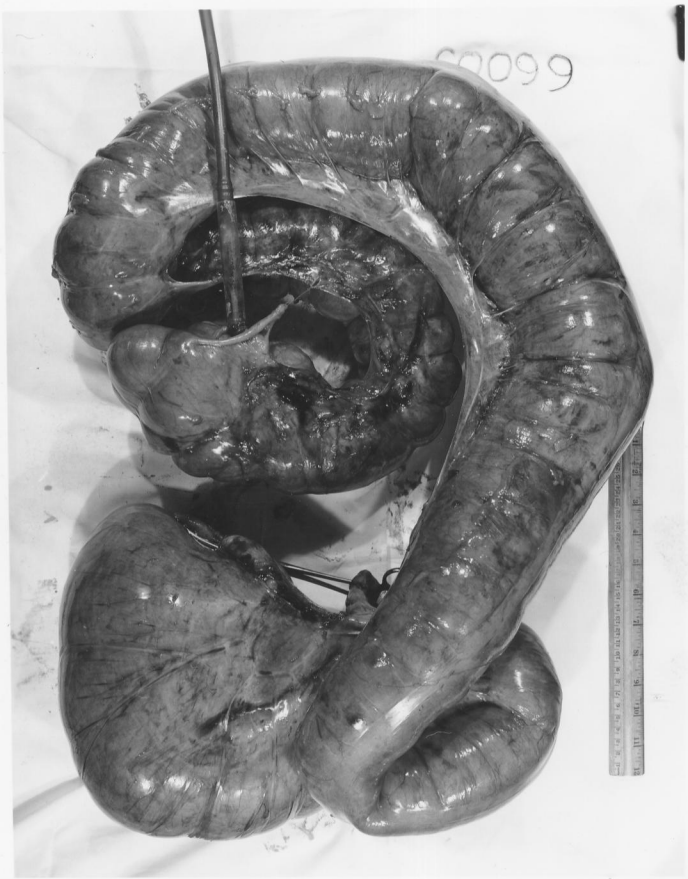
The second admission was from December 1, 1948 to January 7, 1949. He had felt somewhat better since the previous admission until the preceding month when he had felt filled up and had increased amounts of gas. Enemas were necessary three to four times weekly during this time. The physical examination revealed flared costal margins with a "very large" abdomen. Hemorrhoids were seen on rectal examination. There was an ulcer on the left leg. A urinalysis was negative. He had 12.05 grams of hemoglobin and 3.7 million red blood cells. The barium enema was essentially as before but the colon was more dilated. At this admission a colectomy was done with a side-to-side anastimosis of the ileum to the lower one-half of the sigmoid. About one week to ten days postoperatively he began to have from six to ten bowel movements each day. The number

of bowel movements was controlled by the use of paregoric and by belladonna and opium suppositories. On discharge he was having two to four stools daily, was gaining weight, was feeling well, and had a good appetite. He was advised to continue on a bland diet and take paregoric to control his bowel movements.

The third admission was from January 16, 1950 to January 20, 1950. For seven weeks prior to admission he had been having six to ten loose, watery stools each day. Two days of constipation had preceded the diarrhea. He complained of a large amount of gas. The examination revealed slight abdominal distention and high-pitched bowel sounds. On proctoscopic examination the rectum and sigmoid appeared to be ballooned out. Laboratory work was essentially within normal limits. The barium enema studies revealed that the anastimosis was working well, but the rectum, sigmoid, and ileum were moderately dilated. During his hospitalization the diarrhea was controlled with paregoric and bismuth. When discharged he was advised to eat a bland diet, take metamuscil to control the bulk of his stools, and use paregoric for diarrhea.



A. A., 157426. Barium enema demonstrating megacolon in an adult.



A. A., 157426. Megacolon removed at surgery. Colectomy done with side-to-side anastimosis of ileum and lower portion of the sigmoid.

Case Eight

L. D., 262467, Indian male, age 19 months, was admitted December 29, 1948 and was discharged February 5, 1949. The presenting complaints were alternating constipation and diarrhea. During episodes of constipation the patient would sometimes go for as long as a week without a bowel movement. There had been blood and pus in the stool. Symptoms had been present since birth. For six days prior to admission, abdominal distention had been present. The past medical history revealed that he was born in a difficult, nonoperative breech delivery and weighed 7 pounds, 11 ounces at birth. He had had measles, whooping cough, and pneumonia. (During this hospital stay he also had chicken pox.) One other child in the family was said to be anemic. The positive physical findings were as follows: two café au lait spots on the thigh, enlarged tonsils, a "hard" distended abdomen, and a palpable colon. Laboratory studies revealed a hypochromic anemia and the stools were positive for occult blood. The barium enema studies showed the colon to be enlarged from the cecum through the sigmoid to about two and one-half times its usual size. The sigmoid was redundant.

The X-ray report also stated that the small intestine showed a deficiency pattern. Prostigmine was started and the dosage was increased over a period of five days from 5 milligrams three times daily to 15 milligrams three times daily. This medication was continued for a total of 15 days without apparent effect. Two days after mecholyl, 20 milligrams three times daily, was started, he began to have spontaneous bowel movements, and these continued almost daily during his hospital stay. He was given a transfusion of 200 cubic centimeters of whole blood. His management included crude liver, elixir of feragon, vitamins, and cooked fruit with each meal. When he was discharged he was to continue on mecholyl, vitamins, Kondremul oil, and a diet suited to his age.

Correspondence in February, 1950 revealed that the patient was on no medication and was having a soft, brown stool, without blood, every two to three days. He took no laxatives nor enemas. He had "very little" abdominal distention and his appetite was good. He apparently had no sphincter control, since it was stated that "he does not know when he is going to have a stool, seems to have no control over his bowel, a little pain on passing stool . . . ." There had been no additional

illnesses. He did not walk until about two years of age. At the time of correspondence when he was about three years of age, he was 35 inches tall and weighed "about 37 pounds."

Case Nine

G. S., 262925, white male, age 7 years, was admitted on January 30, 1949 and was discharged February 15, 1949. The chief complaints were constipation and enlargement of the abdomen. The diagnosis of megacolon had been made at the age of two months because of constipation, abdominal distention, and emesis. Since that age he had had no spontaneous bowel movements, but his symptoms had been fairly well controlled by the use of enemas about every other day, mineral oil, and milk of magnesia until the past few months when the constipation had increased. The past medical history revealed that his birth had been normal with a weight of 7 pounds, 11 ounces. He had walked at fourteen months. He had had measles and mumps. Two other children in the family were living and well. Physical examination revealed a moderately distended abdomen, visible peristalsis, a palpable colon, and a tympanitic percussion note. The diaphragm was elevated. Routine laboratory work was within normal limits. Barium enema studies revealed that the entire colon was dilated and that the sigmoid was long. His general care consisted of a low residue diet with cooked fruit at each meal, vitamins, Kondremul

oil, mineral oil, and enemas. On February 3 he was started on 25 milligrams of mecholyl. On February 4 this was increased to 40 milligrams three times daily, and on February 5 he was given 75 milligrams three times daily without apparent benefit. The mecholyl was stopped and on February 6 prostigmine was started. From February 6 to February 10 the dose was increased from 7.5 to 22.5 milligrams three times daily. On prostigmine he began to have spontaneous stools and at the time of discharge was having two to three spontaneous stools each day. The instructions at the time of discharge were to continue on that dose of prostigmine, use cooked fruit with his meals, and take vitamins and Kondremul oil.

The second admission was from January 5, 1950 to January 11, 1950. He was admitted because of an episode of epigastric pain followed by fainting just prior to admission. Since his previous admission he had been having one to two bowel movements daily, some flatulence, and increased straining with bowel movements. The physical examination revealed a semicomatose child that was easily roused. The pharynx was slightly congested. The abdomen was moderately distended with increased tension. Urinalysis showed a two plus reaction

for sugar and 0.005 per cent albumin. The first white blood cell count was 27,600 but on January 10 it was 5,350. The blood sugar was 90 milligrams per cent. Barium enema studies were as before. After admission the child had an emesis and passed two loose stools. He responded well to parenteral fluids, sulfadiazine, mineral oil, and enemas. He was started again on prostigmine. On discharge the diagnosis of upper respiratory infection was added to that of megacolon. The instructions were to increase the prostigmine from 7.5 milligrams three times daily to 22.5 or 30 milligrams three times daily if necessary. He was to continue on vitamins and Kondremul oil.

Correspondence in February, 1950 revealed that he was taking a total of 72.5 milligrams of prostigmine daily, Kondremul oil, and vitamin A, and was having a firm bowel movement daily. No laxatives were being used but he had an enema about once weekly. His appetite was good. He had no abdominal distention nor emesis. His height was 53 3/4 inches and his weight was 70 pounds. There had been no additional illnesses since his last admission. He was doing well in school.

Case Ten

C. T., 264250, white male, age 2 1/2 years, was admitted April 15, 1949 and was discharged June 13, 1949. The patient was admitted because of diabetes and pneumonia but also presented complaints of abdominal distention and constipation which had been present for three to four months and were relieved by the use of laxatives. The past medical history revealed that his delivery was normal with a birth weight of 7 pounds. He had walked at the age of fifteen months. He had had pneumonia at the age of three months, recurrent episodes of otitis media, and what was thought to have been the measles. Two other children in the family were living and well. The positive physical findings were mainly those of the respiratory system and the ear. There was a mucopurulent discharge from the left ear. The pharynx was red. In the left anterior superior part of the chest there was dullness to percussion. There were moderate crackling rales throughout the chest. The abdomen showed only slight distention. The routine laboratory examinations were negative except for sugar in the urine and the fluctuating blood sugar. The chest X-ray showed a resolving pneumonic process in the right

lower lung. Barium enema studies revealed that the sigmoid and ascending colon were somewhat enlarged and this was believed to represent a mild degree of megacolon. The rectum was normal. Therapy was directed mainly toward the resolution of the pneumonic process and the control of the diabetes. When he was discharged, he was to use 10 units of protamine zinc insulin each morning before breakfast with a diet of 1185 calories. Kondremul oil was to be taken for his constipation.

The second admission, from December 23, 1949 to January 13, 1950, was for adjustment of his insulin dosage. There were no gastrointestinal complaints noted. On physical examination, the abdomen was slightly distended and was described as feeling doughy. A few scybala were palpable. The patient also had a follicular tonsillitis. Laboratory work again was negative except for sugar and acetone in the urine and the abnormal blood sugar. The tonsillitis was treated with penicillin and on discharge he was instructed to take 6 units of protamine zinc insulin and 12 units of regular insulin daily.

The patient was seen in the outpatient department on October 2, 1949. The note stated that his appetite was good and he was having normal bowel movements

generally, although he required occasional enemas and sometimes became quite distended. On examination the abdomen was somewhat distended and felt doughy.

An outpatient note on March 10, 1950 stated that he had had a severe upper respiratory infection and pneumonia three weeks previously. Ten days prior to this visit he had had a severe insulin reaction and convulsions. His appetite was poor and he required almost daily enemas. His abdomen was found to be very large and protuberant. It was recommended that he be started on 7.5 milligrams of prostigmine daily which should be gradually increased in an attempt to stimulate more frequent bowel movements. His insulin dosage was also adjusted at this visit.

Case Eleven

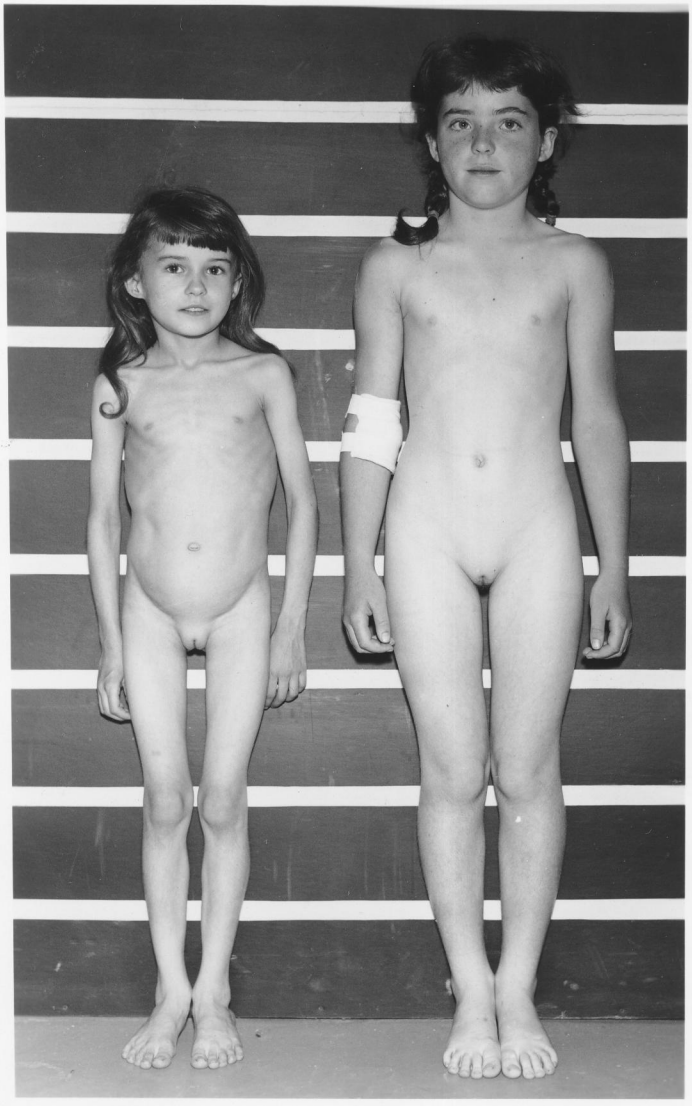
R. A., 265239, white female, age 9 years, was admitted June 21, 1949 and was discharged July 10, 1949. The onset of her illness had been at the age of two months, since when she had had constipation and abdominal distention when she did not have a bowel movement. She used laxatives daily and an occasional oil enema. For the three days preceding admission she had had diarrhea. The past medical history revealed a normal delivery and a birth weight of 4 pounds. She had talked at twelve months and walked at thirteen months. She had had measles and chicken pox. Four other children in the family were living and well. Physical examination revealed a small girl who appeared about the size of a six-year-old child. There was slight abdominal distention and peristalic waves were visible. The percussion note was generally tympanitic over the abdomen except for the left lower quadrant where the note was dull. Routine laboratory work was within normal limits. A photofluorogram of the chest was normal. The barium enema studies showed a very large rectum and sigmoid, and the colon elsewhere was somewhat enlarged. A KUB film and an intravenous pyelogram were negative. The bone age was reported as six years,

nine months. She was placed on a low residue diet, which was later changed to a general diet, with cooked fruit at each meal. She was given vitamins to supplement this. On June 28, prostigmine was started (7.5 milligrams three times daily) which was continued until July 1. On July 1, DHO 180 was started at 1 milligram three times daily before meals. By July 7 the dose had been increased to 5 milligrams three times daily before meals. She had had several spontaneous stools and consequently was discharged on DHO 180, 5 milligrams three times daily, to return in one month.

The second admission was from August 11, 1949 to August 19, 1949. She had had spontaneous bowel movements every four to five days--large in amount but otherwise normal. The DHO 180 had been discontinued about one week prior to admission and the bowel movements had continued. The physical examination was essentially negative except for slight protuberance of the abdomen with standing and tympany with percussion. The laboratory work was negative as before. The barium enema report was essentially as before except that it was thought that the rectum and sigmoid might be larger. The rest of the colon was reported as being within normal limits. During her hospitalization she developed

a pharyngitis and tonsillitis which were treated with penicillin. Cooked fruit and vitamins were given to supplement her diet, but no specific medication was given. On discharge the mother was advised to use no specific medication for a period of two weeks, but to write for a supply of DHO 180 if more enemas were needed or distention developed.

Correspondence in February, 1950 revealed that she was continuing to have a bowel movement about every four days which appeared normal. She was receiving no medication, but was given an enema if her bowels did not move regularly. There was no abdominal swelling. Her appetite was good. Her height was 49 1/2 inches and her weight was 41 pounds. Her school work was good. The only other illnesses which she had had were upper respiratory infections.



R. A., 265239. R. A. is child on left. Taken June 22, 1949. Shown with a child of approximately the same age and of normal development.

Case Twelve

R. P., 267436, white male, age 3 1/2 years, was admitted October 25, 1949 and was discharged October 26, 1949. The child was admitted for study because of an abdominal mass which was discovered on a routine physical examination. The history was that of constipation since one and one-half years of age with a bowel movement about every three days after a glycerine suppository. His appetite was good. He had emesis rarely. "Burping" had been common until one year of age. Past medical history revealed that the child had been delivered by Cesarean section and had weighed over 8 pounds. The development had apparently been normal. He sat up at six months and walked at thirteen months. The only illnesses had been an ear infection and a streptococcal throat infection. The child had been adopted. On inspection of the abdomen there was moderate distention with prominent veins over the upper abdomen and lower chest. The pattern of the bowel was visible but no peristalsis was seen. Palpable scybalous masses disappeared after an enema. The percussion note was tympanitic. The urinalysis was negative. The white blood cell count was 15.4 thousand, but the differential

count was normal as was the red blood cell count. Barium enema studies revealed marked lengthening of the left side of the colon with enlargement of the whole colon to the anal canal. The patient was discharged with instructions to take (1) liberal amounts of water, (2) cooked fruit with all meals, (3) vitamins, and (4) two to three teaspoons of milk of magnesia if needed.

Correspondence in December, 1949 stated that the child was having a bowel movement every three days with a teaspoon of milk of magnesia nightly. In a letter returned to the mother she was advised to increase the dose of the milk of magnesia to two or three teaspoons or more nightly in order to have a bowel movement every one to two days. She was also advised to see a pediatrician in California where they were moving, where perhaps the use of prostigmine could be considered.

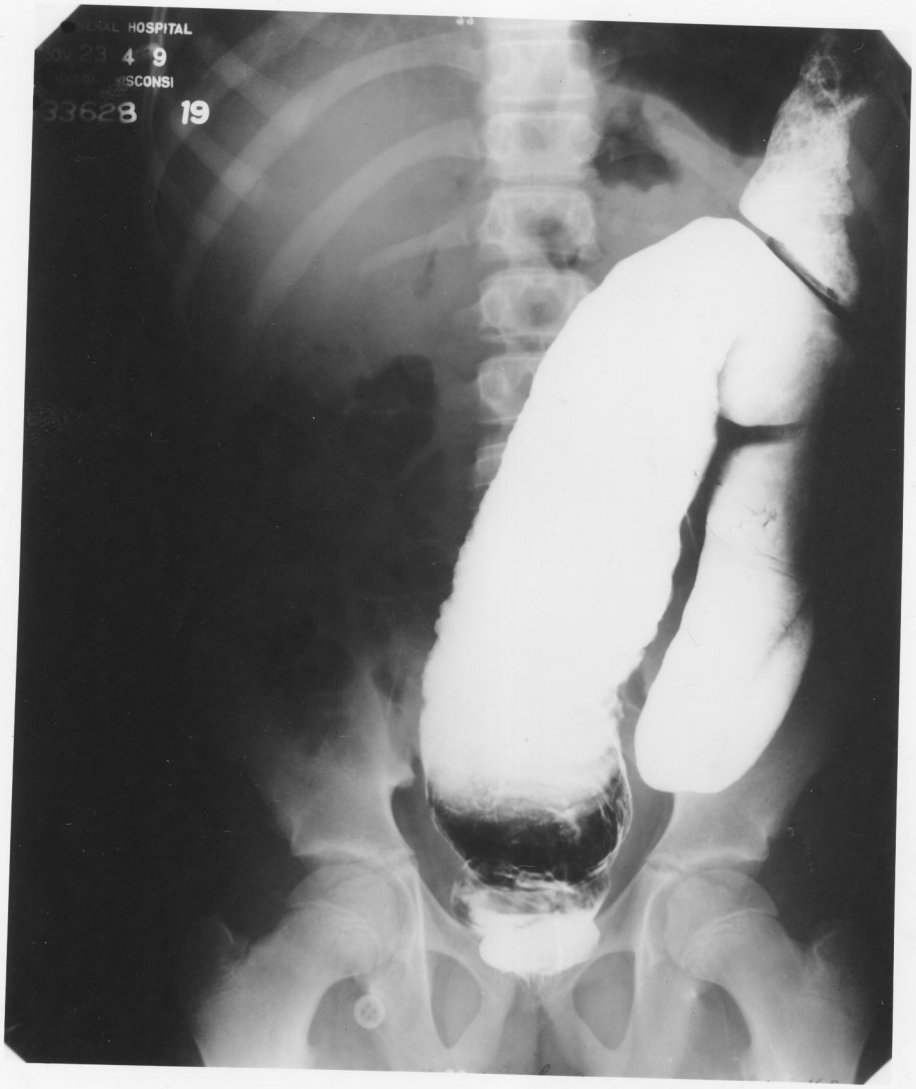
Case Thirteen

R. W., 267854, white male, age 15 years, was admitted November 21, 1949 and was discharged December 10, 1949. His chief complaint was "bowels don't move regularly." He had had chronic constipation with bowel movements from one day to one week apart. His mother stated that his symptoms had started when he was ten days of age. He had abdominal distention which was somewhat relieved after a bowel movement. The stool usually was hard at first, followed by a large, soft, foul stool. Cathartics and enemas had been used to control the symptoms. He complained of flatus. Nausea and vomiting were relieved by a bowel movement. Frequently he could see peristalsis through the abdominal wall. He had had hemorrhoids. The past medical history revealed that he had measles, mumps, and chicken pox. Three brothers were living and well. One brother had died at Wisconsin General Hospital of generalized moniliasis. The physical examination revealed a 15-year-old boy who appeared small for his age. The skin was tight over the abdomen and the veins were prominent. There was no visible peristalsis. A hard mass was palpated in what was thought to be the ascending

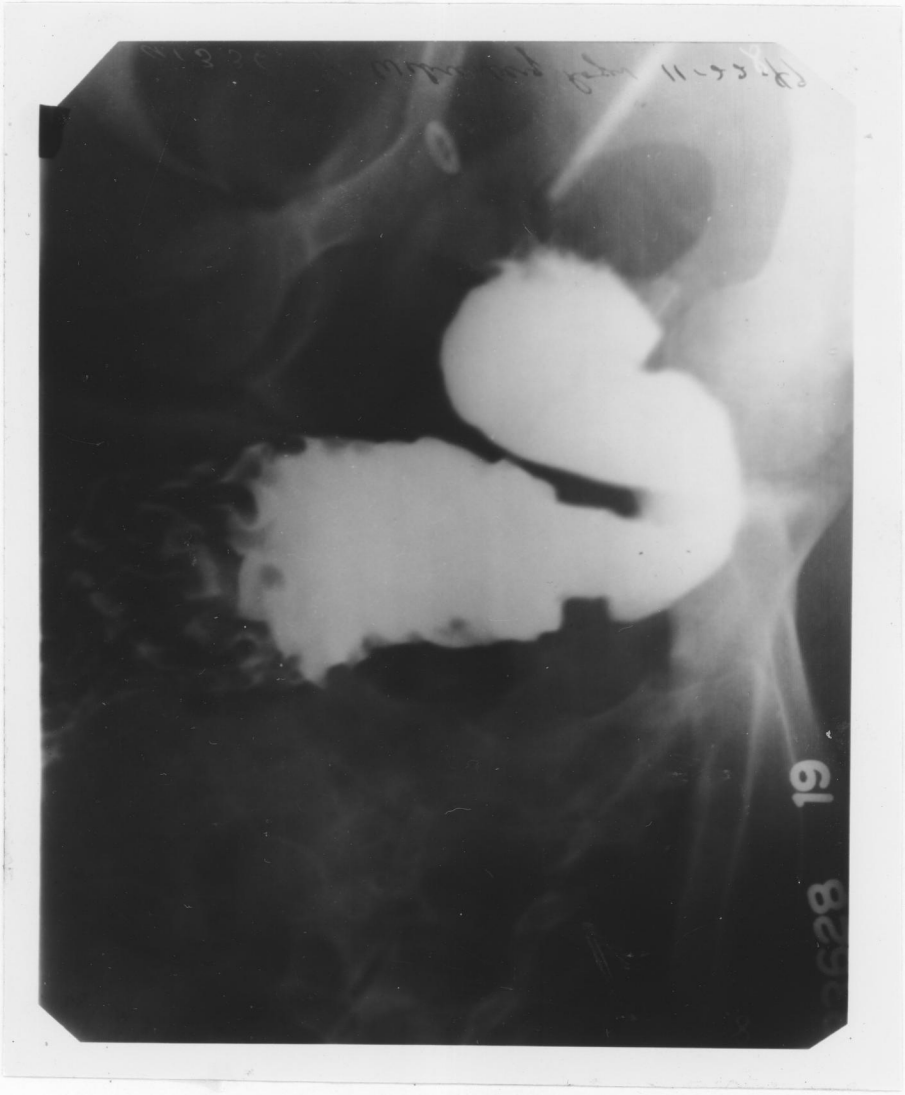
colon. The percussion note was tympanitic. Bowel sounds were decreased. Other findings included flared costal margins, a diffusely enlarged thyroid, and large tonsils. The boy was 60 3/4 inches in height and weighed 87 pounds. A proctoscopic examination showed an area of contraction just distal to the dilated portion which relaxed after about three minutes to admit the proctoscope. The laboratory work revealed a normal urinalysis and 13.1 grams of hemoglobin. The barium enema study showed a dilated and redundant sigmoid. The dilatation started approximately at the rectosigmoid junction and extended proximally. The descending colon was approximately normal in caliber. Evacuation was complete. Therapy here included enemas, vitamins, cooked fruit, and liberal amounts of fluids. Specific medication was prostigmine which was started November 28. The initial dose was 7.5 milligrams three times daily, which was increased to 15 milligrams three times daily. Because of the improvement and spontaneous bowel movements on this medication, he was discharged on this dose of prostigmine and vitamins.

The second admission was from February 10, 1950 to February 20, 1950. He had used the medication

prescribed and had gained five pounds, had normal bowel movements daily, a good appetite, and more "pep." The physical examination showed him to be slightly underdeveloped for his age, slight protuberance of the abdomen, hyperresonance, no visible peristalsis, and some areas of increased bowel sounds. The laboratory work was negative. The barium enema studies revealed that there was greater redundancy in the sigmoid region and redundancy in the areas of the flexures, particularly in the region of the hepatic flexure. It was thought that the caliber of the bowel was less but the change was considered to be minor. Evacuation was "fair." Except for an episode of acute enteritis which resulted from a dose of castor oil and which subsided quickly, his treatment was as before, and he was discharged on the same medication to return in June, 1950.



R. W., 267854. Barium enema showing megacolon.



R. W., 267854. Spot film, taken with the patient in an oblique position, showing dilatation beginning abruptly in the area of the rectosigmoid junction. Distal to the dilatation, the rectum is approximately of normal caliber.

PART THREE

Chapter XIII

CONCLUSION

A discussion of congenital megacolon including a new concept of etiology has been presented, together with thirteen case summaries from the records of Wisconsin General Hospital, Madison, Wisconsin.

The work of Whitehouse and Kernohan, Swenson and his group, Higgins, and others, is logical from a theoretical viewpoint and is based on well-controlled experimental data. It has been the basis of a successful surgical method of treatment. They believe congenital megacolon is a dilatation and hypertrophy of the colon secondary to a normal appearing but obstructing segment in the distal sigmoid and rectum which is nonfunctioning due to a congenital absence of the myenteric plexus. Up to the present time successful results have followed the resection of this segment.

The treatment of the cases from Wisconsin General Hospital presented in this paper has been primarily medical in all instances except one. Six different drugs which act specifically on the colon have been used, as well as sympathetic block and spinal anesthesia.

None of these has been consistently successful.

All six drugs, spinal anesthesia, and sympathetic block were tried on one patient over a period of years without permanent control of symptoms being achieved. He now has a colostomy preliminary to a rectosigmoidectomy which is to be done in the near future. One patient, who apparently was well-controlled with medical therapy for five years, expired suddenly with volvulus. Another patient expired suddenly after an enema and no specific reason could be found at the post-mortem examination. These cases stand as evidence that medical management is not the most desirable approach. Moreover, according to the literature, colectomy and sympathectomy have not been entirely successful; but success has been achieved in 34 cases following a rectosigmoidectomy. These 34 patients were having spontaneous bowel movements following this procedure. Perhaps rectosigmoidectomy is the answer to successful therapy in those cases where a nonfunctioning lower sigmoid and rectum exist.

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