THE SURGICAL TREATMENT OF CONGENITAL MEGACOLON*

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SINCE CONSIDERABLE EVIDENCE has been recorded that the abnormal physiology of congenital megacolon consists primarily of a lack of propulsive capacity of the rectum or rectum and lower sigmoid, the logical method of cure is a removal of the distal nonpropulsive segment.

The disease is of congenital nature, apparently due mainly to a diminution of parasympathetic ganglion cells in the wall of the recto-sigmoidal segment. As has been pointed out in the previous paper on the abnormal physiology of congenital megacolon, there is a delicate balance between the distal obstruction and the compensatory mechanism for overcoming that obstruction, *i.e.*, hypertrophy and dilatation. The severity of the hypertrophy and dilatation and the severity of the obstruction are dependent upon each other, so that an increase or decrease in one results in an increase or decrease in the other. If the propulsive capacity of the proximal hypertrophied and dilated segment is increased, there is a gradual increase in the obstruction as well, due probably to increased hypertrophy in the proximal portion of the nonpropulsive segment, thereby re-establishing the balance. If the distal obstruction is diminished by relaxation of the nonpropulsive segment, there is a proportionate loss in the compensatory mechanism of hypertrophy and dilatation, and again the balance is gradually re-established.

On the basis of this concept, there was little to expect from drug therapy, lumbar sympathectomy, or partial resection of the distal nonpropulsive segment. In our experience with treating several megacolon patients with Mecholyl, Prostigmin, and Syntropan, none of these drugs had a more than temporary beneficial effect. The poor results from lumbar sympathectomy have been previously reported,[1, 2] and our experience with inadequate resection will be discussed later.

Therefore, in order to cure congenital megacolon, the entire distal nonpropulsive segment has to be removed. Our first attempts to effect this surgical cure were inadequate, since there was no known procedure for complete removal of the rectum that was not associated with a high morbidity. In the first patient operated upon, only the lower one-half of the sigmoid and upper one-third of the rectum were removed with an end-to-end anastomosis. This resulted in marked clinical improvement, but only a moderate reduction in the hypertrophy and dilatation in the colon proximal to the remaining rectal segment.

Our next four attempts took the form of a lower sigmoidal resection with an anterior resection of the rectum, the mid-sigmoid being anastomosed to a rectal stump 4 to 6 cm. long. This resulted in an almost normal ability of these patients to have bowel movements. However, there was in all four cases a persistence of the megacolon in a degree proportionate to the amount of rectum left intact.

At this point, a method of total removal of the rectum and lower sigmoid devised by Swenson[3] appeared in the literature. Dr. Swenson, in conjunction with Newhauser,[4] had noted from radiologic examination (in the form of barium enemata) of patients

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with congenital megacolon what he described as "benign spasms of the recto-sigmoid." His concept was similar to ours in that he emphasized that the hypertrophied and dilated segment was capable of normal propulsive action and presented conclusive evidence of a recto-sigmoid dysfunction producing obstruction.[5] The procedure he recommended was then used on our next patient with excellent results. However, in our hands the procedure was somewhat cumbersome, and we therefore revised it so that it could be done with more ease and practically no contamination. Briefly, the operation consists of a surgical intussusception of the entire rectum and lower two-thirds of the sigmoid through the anus, with an end-to-end anastomosis of the anus and sigmoid in one stage without proximal decompression.

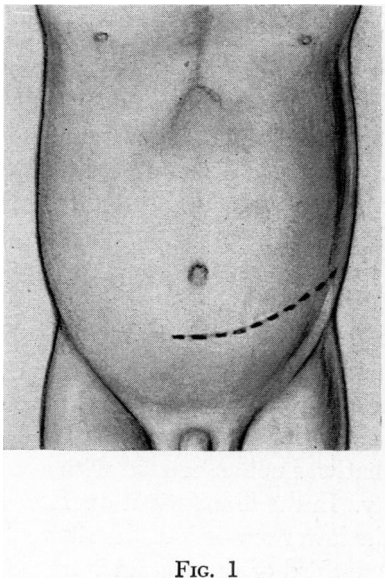


FIG. 1

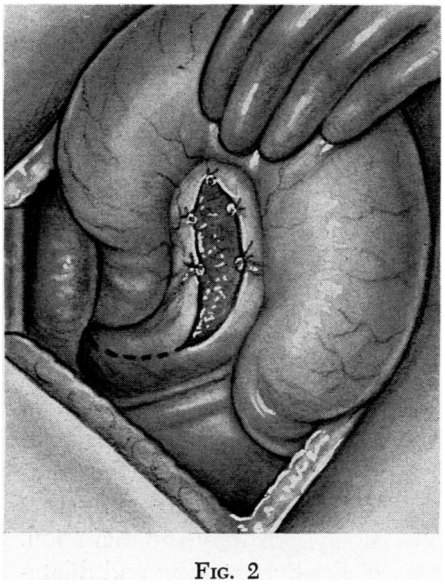


FIG. 2

FIG. 1.—Dotted line represents type of incision.
FIG. 2.—Incising of sigmoid mesocolon with ligation of superior hemorrhoidal and lower sigmoidal arteries.

In more detail, the procedure is as follows:

The patient is placed on the table in the recumbent position with legs abducted on special supports to allow easy access to the anus. A catheter is placed into the bladder.

A left lower oblique incision is made dividing the entire left rectus and one-half of the right rectus. The incision is carried laterally, far enough to allow mobilization of the descending colon and detachment of the splenic flexure if necessary. The aponeurosis of the external oblique is split in the direction of its fibers, and the internal oblique and transversalis muscle divided. Upon opening the peritoneum, the sigmoid is delivered into the wound and the remaining gut confined to the upper abdomen. The superior hemorrhoidal artery and vein are divided and ligated. The mesosigmoid is divided up to a point of election at least 20 cm. proximal to the distal end of the hypertrophied and dilated segment. Here a 6 cm. cuff of bowel is cleared of attached tissue. It is frequently necessary to mobilize the descending colon and occasionally to detach the splenic flexure to allow adequate mobility of the upper sigmoid. The dissection is then carried distally by opening the pre-sacral space beyond the coccyx and incising the pelvic peritoneum around the rectum. The lateral ligaments

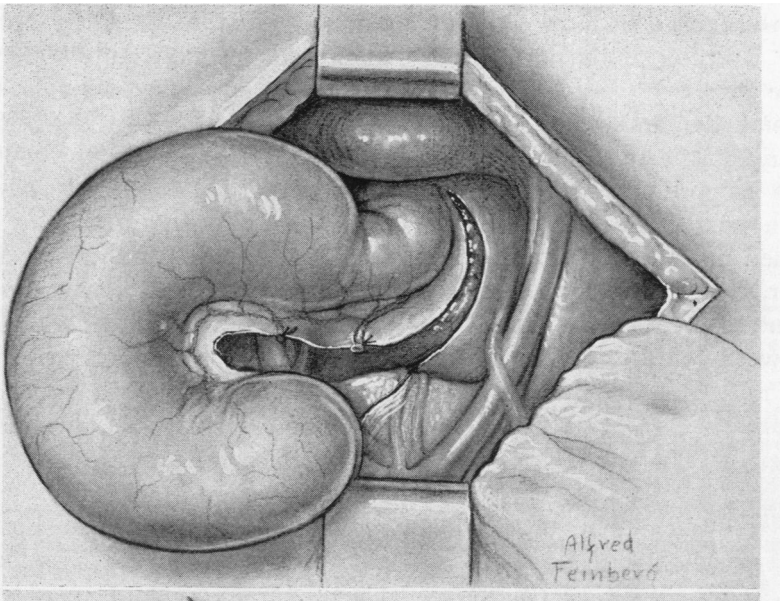


FIG. 3

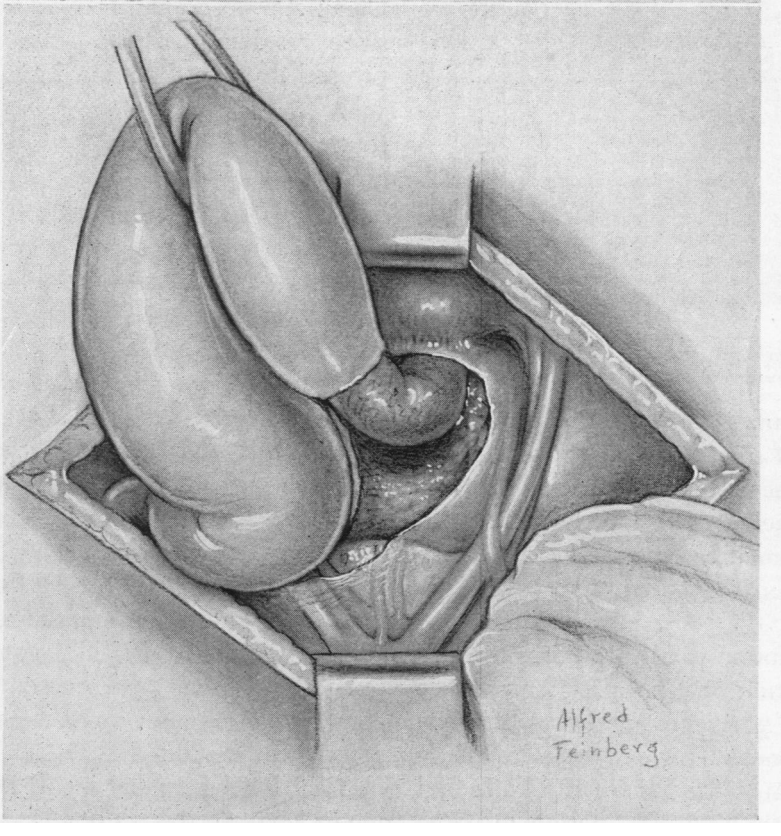


FIG. 4

FIG. 3.—Incising of sigmoid mesocolon with ligation of superior hemorrhoidal and lower sigmoidal arteries.
FIG. 4.—Exposure of presacral space which is open to the perineum.

of the rectum are divided and the rectum separated from the bladder and prostate or vagina anteriorly. This dissection is carried distally to the insertion of the levator ani muscle into the anus.

At this point the assistant inserts an Allis clamp through the anus into the mid-rectum where a portion of the wall is grasped. With gentle traction the lower rectum is turned inside out. Repetition of this maneuver easily effects a complete eversion or intussusception of the entire rectum and lower two-thirds of the sigmoid

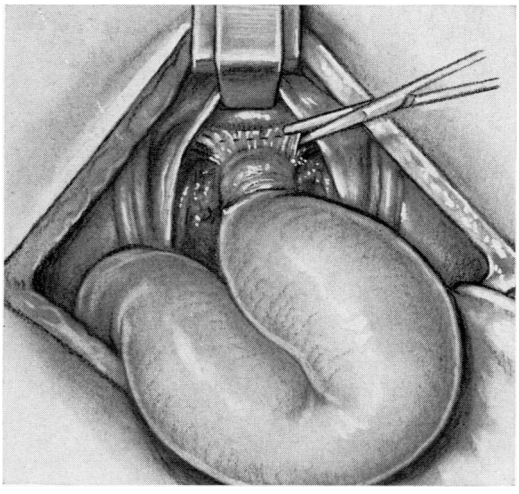


FIG. 5.—Dissection of rectum from the prostate and urethra or vagina.

through the anus, bringing the previously cleared cuff of sigmoid down to the level of the anus. The pelvic peritoneum is then loosely re-attached with interrupted silk to the newly-formed rectum, and the abdominal wall closed without drainage in layers with fine silk.

We have found it advantageous to have a Penrose drain in the pre-sacral space through the perineum midway between coccyx and anus. This can be effected by tying the drain to the handle of a knife and inserting it through the perineum from above before beginning the rectal eversion.

The patient is then placed in the lithotomy position and an anastomosis effected between the anus and the inlying sigmoid. The rectum is detached from the anus by a circumferential incision, the muscularis of the anus being sutured to the seromuscular layer of the inlying sigmoid with interrupted sutures of silk. The sigmoid is divided just distally and the mucosa of the anus approximated to the mucosa of the sigmoid with continuous chromic catgut. Upon completion of the anastomosis, it is withdrawn into the anus by the intact levator ani muscle.

The preoperative preparation of these patients consists of adequate doses of phthalyl sulfacetimide by mouth for four days with correction of any blood or electrolyte abnormality that may be present. A reasonable attempt at colonic decompression should be made by saline enemata. Complete decompression is not essential since residual gas and feces can be expressed through the anus during operation.

A transfusion during operation is given routinely.

Postoperatively the patients are given nothing but sips of water by mouth and are placed on streptomycin, penicillin, and phthalyl sulfacetimide for a four-day period. Then they are allowed out of bed and their diet is progressed rapidly. During this period it is important to maintain the blood and electrolyte values at normal level by infusions and transfusions if necessary.

Since December 1947, 30 patients at the Columbia-Presbyterian Medical Center, New York City, have been operated upon for congenital megacolon. Of the 23 treated by the method described, the follow-up results have been uniformly good. (In the remaining seven, as discussed before, the result was largely dependent upon the amount of rectum left intact.) They all initially have frequent small bowel movements because the residual hypertrophy of the newly-formed rectum is powerful enough to overcome the sphincter mechanism. This hypertrophy quickly subsides,

so that within a month the patients are having between two and five bowel movements a day with only minimal soiling between movements. As a rule these patients become aware of the impulse to defecate within two weeks after operation.

The postoperative complications have all emanated from the same underlying cause, *i.e.*, infection of fluid collections in the presacral space. Seven of the 23 patients in whom a total rectal removal was done developed this complication. In four, the infection resolved spontaneously with drainage into the newly-formed rectum, but the remaining three required a transverse colostomy to bypass temporarily the fecal stream. Our fear of this complication has been minimized by the institution of proper drainage technics as described in the procedure, plus the routine use of penicillin, streptomycin, and phthalyl sulfacetimide for four days postoperatively, but it still remains a threat. A transverse colostomy should be resorted to without delay should this complication be severe.

There was one death in the series in an extremely debilitated and critically ill 18-months'-old child, eight hours postoperative, from aspiration of vomitus which took place when the nurse was temporarily away from the bedside.

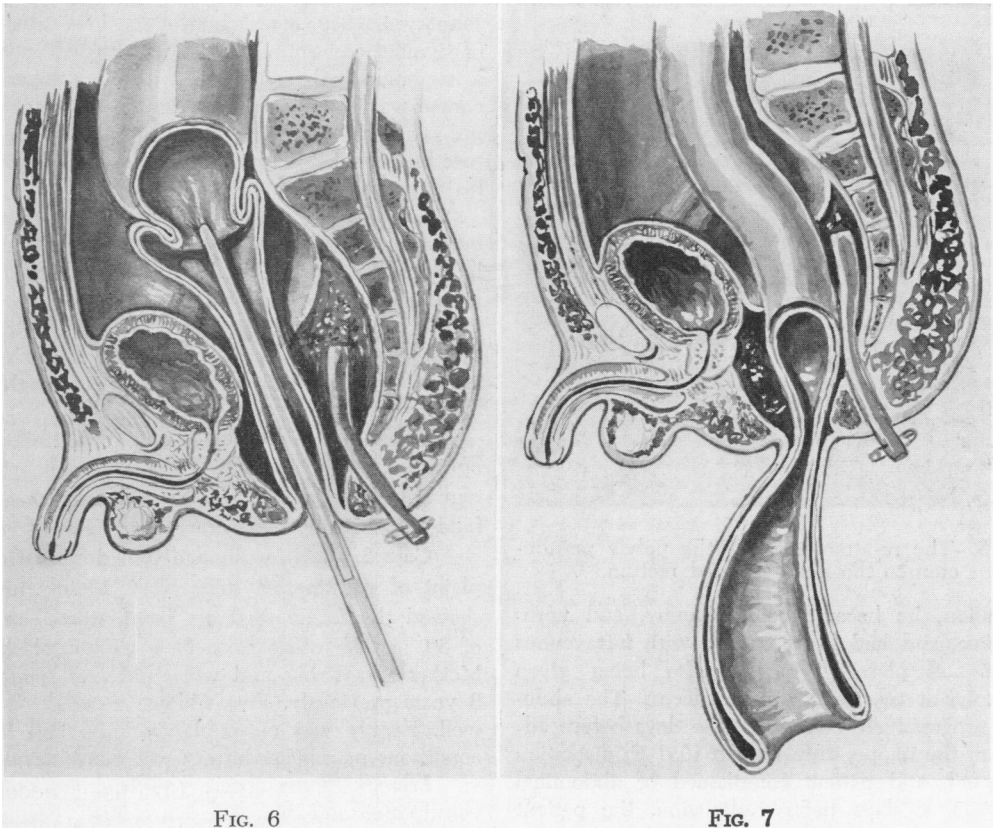


FIG. 6 FIG. 7

FIG. 6.—The ideal limit of dissection to the insertion of the levator ani muscle.
FIG. 7.—A cross-section of the pelvis at the completion of the rectal eversion.

In order to clarify further this approach to congenital megacolon, three cases will be reviewed in detail. Each of the three cases will represent one of the three forms of congenital megacolon, *i.e.*, the classical type, the gradual transition type, and the megarectum type.

Case 1.—*G. F.*, male, age 4, had a chief complaint of distention, vomiting, and fever. Family history was noncontributory. The mother had a thyroidectomy in 1940. Past history was noncontributory.

Present Illness. Patient had been constipated since birth, seldom had a bowel movement without mechanical aid, but was never allowed to go more than 48 hours without defecating. The mother noted that after enemata he was usually weak, tired, and sleepy. After an enema one year before

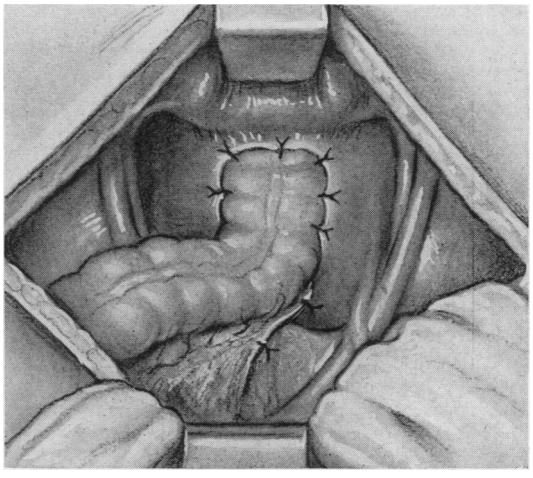


FIG. 8.—The re-attachment of the pelvic peritoneum to the newly-formed rectum.

admission, he became cold, clammy, and unresponsive, and had to be treated with intravenous saline and plasma. He was also being given Mecholyl at the time without success. The abdomen progressively enlarged. Five days before admission the temperature rose to 105° F, distention increased, and patient complained of abdominal pain. Three days before admission, the patient began vomiting and had no more bowel movements.

Physical Examination. T 103; P 90; R 26; and B.P. 130/90. The patient was a thin, pale, dehydrated male with a prominent abdomen, complaining of pain when the abdomen was touched. Head and neck were normal; and chest clear to percussion and auscultation. Heart was normal and abdomen protuberant and tympanitic with tenderness throughout. There was no spasm. There were occasional high-pitched peristaltic sounds, but no visible peristalsis. Rectal sphincter tone was increased, but otherwise normal. There were some hard feces in rectum. Extremities were negative.

Laboratory findings: Hgb. 8.5 Gm. WBC was 13,200 with normal differential. Urine was negative; chlorides 104 meq/L; hematocrit 29.4; ESR 62; Kline, negative. Roentgen ray of abdomen showed air in greatly dilated loops of large bowel. There was no gas in the rectum.

Course. On daily enemata and penicillin the patient's fever subsided and the distention and tenderness diminished. Barium enema revealed a tremendous dilatation of descending colon and upper two-thirds of the sigmoid with an abrupt transition to a small collapsed lower sigmoid and rectum. Patient was given two transfusions of whole blood as preoperative preparation, and on January 4, 1949, a resection of the lower one-half of sigmoid and entire rectum was carried out with a sigmoido-anal anastomosis. The postoperative course was complicated by the development of a pre-sacral abscess which drained through the suture line and healed promptly. Was discharged on his twenty-first postoperative day.

Pathology. An abrupt transition was revealed between the tremendously hypertrophied and dilated segment and the distal collapsed segment. Histologically there were numerous ganglion cells in the hypertrophied segment down to the transitional area, where they were absent. In the distal rectum ganglion cells could again be seen. Nerve fibers evident in all sections.

Follow-up Results. Sixteen months following operation the bowel function was normal, one to two bowel movements a day without leakage. A follow-up barium enema revealed a normal colon.

Case 2.—J. N., male, age 7, had a chief complaint of constipation since birth. Family history showed the father died of "heart attack" at age of 39, and he was said to be a chronic alcoholic. Mother was living and well, and had remarried 3 years previously. Five siblings were living and well. Family was on public welfare, and living conditions poor. Past history was noncontributory.

Present Illness. There had been intermittent constipation and distention since birth. Diagnosed by roentgen ray as congenital megacolon at age 4. He was maintained on mineral oil and frequent enemata until 3 months prior to admission, when constipation and distention increased and child became malnourished and dehydrated. He was treated with Mecholyl and Prostigmin with only slight temporary success.

Physical Examination. The patient was moderately well-developed and well-nourished white male in apparent good health. Positive physical findings were confined to the abdomen, which was protuberant and round. Greatly enlarged sigmoid was easily palpable and full of feces. Rectal examination revealed only increased tone of the sphincter.

Laboratory findings showed blood count and urinalysis to be normal. Tuberculin test was nega-

tive to 1 mg. Barium enema was greatly enlarged sigmoid and descending colon with a gradual transition to a narrowed lower sigmoid and upper rectum.

Course. On the eighth hospital day operation was performed in which rectum and lower sigmoid were resected and an end-to-end anosigmoidostomy performed. Postoperative course entirely uneventful. He began having bowel movements on the first postoperative day, and was out of bed on the fourth postoperative day. Fever did not rise above 100.5°. The wound healed per primum but not absence of ganglion cells which extends far up into the hypertrophied segment. While clinically markedly improved, the patient still has some residual pathology.

Case 3.—H. L., female, age 23, had a chief complaint of severe constipation since birth, with intermittent swollen abdomen for 8 years. She was born in New York City, and had never been in the tropics. She was unmarried. She reported sleeping poorly and getting up every night because of hunger to "eat a snack." She smoked moderately while feeling well. Diet was adequate, but

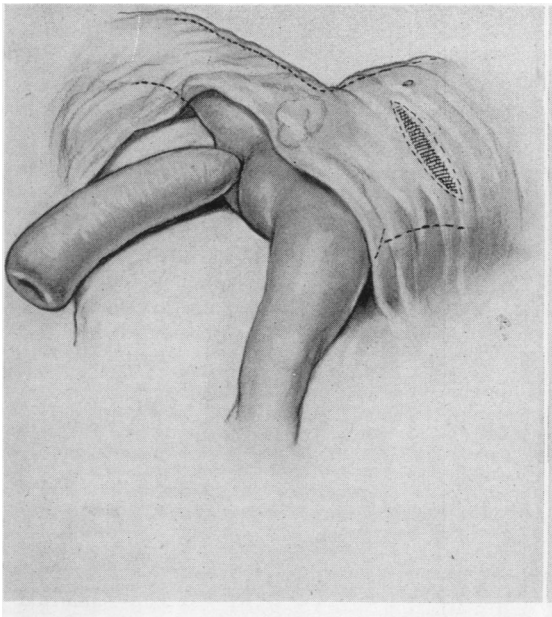


FIG. 9

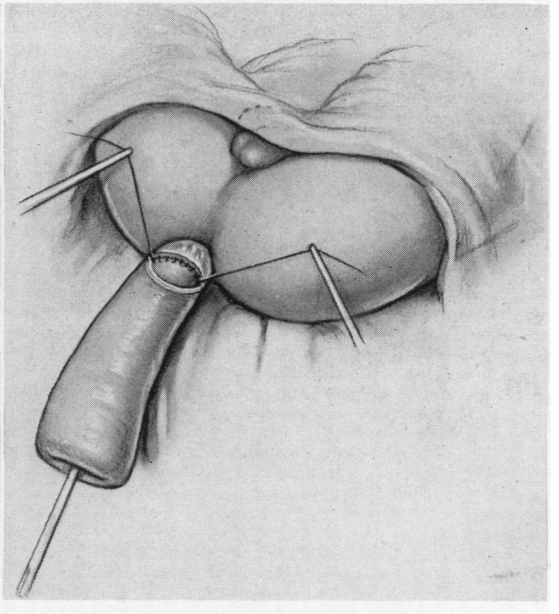


FIG. 10

FIG. 9.—The appearance of the surgical specimen at the completion of the rectal eversion.

FIG. 10.—Beginning of the ano-sigmoidal anastomosis, showing the first row of seromuscular sutures being placed.

and the patient was discharged on the twelfth postoperative day.

Pathology. There was a marked reduction of ganglion cells in all segments of the rectum and sigmoid resected, but not a complete absence.

Follow-up Result. For 2 months the patient had frequent small stools without complete control. They gradually diminished to 3 a day with normal control. One year postoperative examination revealed a persistence of sigmoid dilatation. Rectal examination revealed a slightly tonic sphincter.

Comment. The follow-up and pathologic appraisal of the operative specimen revealed this patient to have had the gradual transition type of congenital megacolon in which there is a reduction

she had special cravings for foods. She had been occupied as a secretary, part time, for 6 years. Her weight averaged 84 pounds; maximum 89 in 1943.

Previously she had always felt well in spite of bronchiectasis and undernutrition. Whooping cough and pneumonia were reported occurring at age of 4, with "pneumonia" every winter since age of one. Operations reported were tonsillectomy at age of 5; there were no previous injuries.

Repeated episodes of pneumonia resulted in severe bilateral bronchiectasis with a productive cough. She required chemotherapy during every U. R.I. There was moderately increased exertional dyspnea, and orthopnea because of cough on lying flat. Nocturia was experienced 1 to 2 times O.N.

Only one menstrual period following "injections" in 1944.

The patient was withdrawn and shy, and very self-conscious of protuberant abdomen and lack of

Present Illness: The patient had severe obstipation since birth, requiring gradually increasing effort in the form of rectal irrigations to produce results. At the age of 14 her abdomen began

FIG. 11

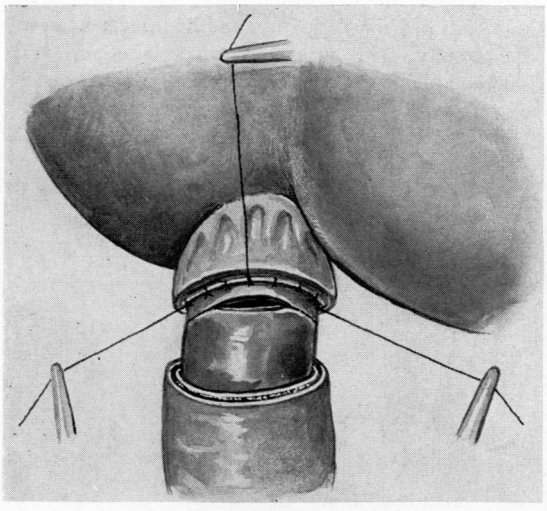
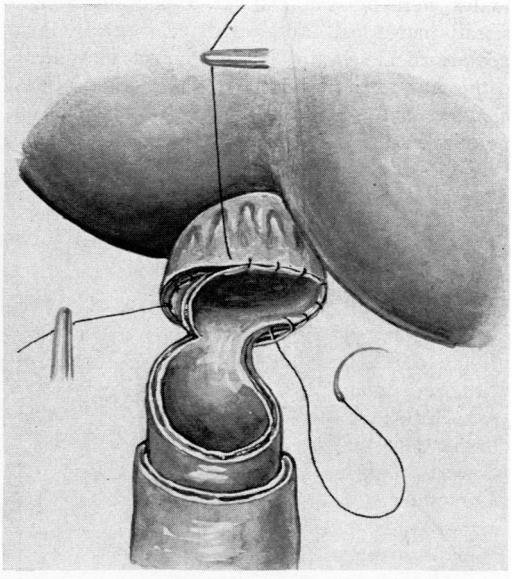
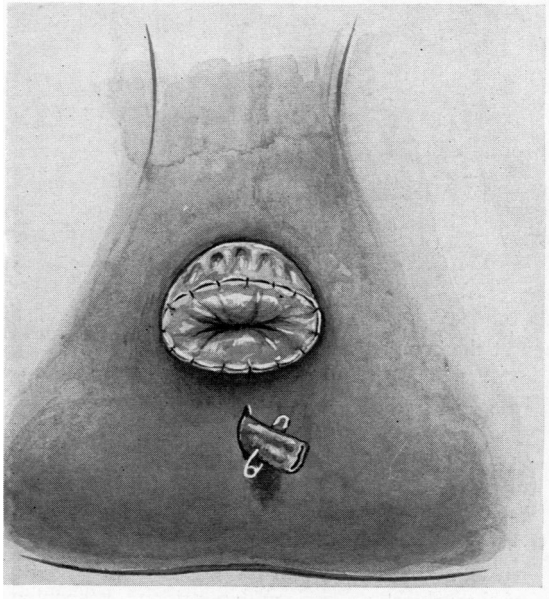


FIG. 12





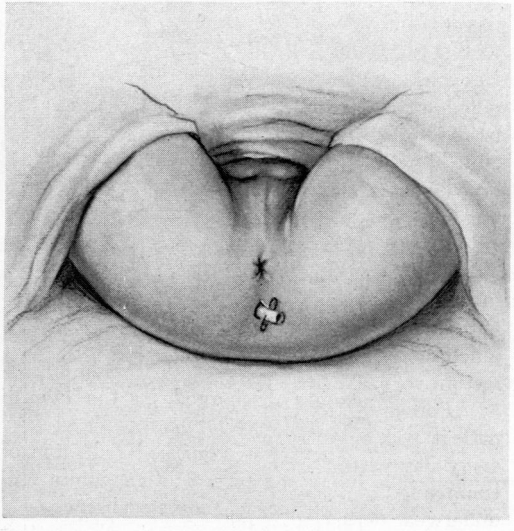


FIG. 13

FIG. 14

FIG. 11.—Completion of seromuscular layer with incision into inlying sigmoid.
FIG. 12.—The placement of continuous muscularis suture.
FIG. 13.—Completion of anastomosis.
FIG. 14.—The appearance of the anus when the levator muscle is allowed to retract the anus.

breast development. Social activities were greatly curtailed.

Family History. Mother and father and one sibling were living and well. One brother died of "pneumonia" at 3 months of age. There was no history of familial disease.

to enlarge in spite of rectal treatments. She developed severe anemia and intermittent ankle edema at age of 20.

Physical Examination. P 135; R 26; and B.P. 130/65. She was a small, pale, thin female appearing more the age of 13 than stated age of 23.

She was active and alert. Eyes, ears, nose and throat examination was essentially negative. The neck was supple, and the thyroid not enlarged. There was no venous distention. There was marked increase in chest A-P diameter. Respiratory excursions were limited. Coarse rhonchi and medium râles were present over lower half of chest. The heart had a regular sinus rhythm, with moderate enlargement to left. There was marked tachycardia and a systolic murmur over entire precordium, loudest over pulmonic area. The abdomen was protuberant and tympanitic; with no shifting dullness. There was occasional visible peristalsis, and a large movable mass in the lower abdomen. Extremities showed marked clubbing of fingers and toes. There was no sacral edema. Reflexes were physiologic. Rectal examination showed a hypertrophied sphincter mechanism and small anal orifice.

Laboratory Findings. R.B.C. 5.2; W.B.C. 7,900; and Hgb. 11.5. The Kline test was negative. Serum protein was 7.8; alb. 5.0; and globulin 2.8. Urine was negative. Barium enema showed a tremendous dilatation of the sigmoid and rectum down to the anus. Evacuation was very poor. Chest x-ray showed marked increase in bronchovascular markings. Rounded shadow, in each lower lung field were suggestive of bronchiectasis cavities.

Course. On May 24, 1949, resection of the rectum and lower half of the sigmoid removed with a sigmoido-anal anastomosis was performed. Her postoperative course was stormy because of poor pulmonary reserve and the development of a mild pre-sacral infection. The latter subsided quickly with catheter irrigation and systemic streptomycin and penicillin. She was discharged on her fifteenth postoperative day.

Pathology. The operative specimen revealed an absence of ganglion cells in the wall of the rectum for a distance of 6 cm. proximal to the anus with an apparent reduction up to the recto-sigmoid junction. The muscularis of the sigmoid and entire rectum revealed tremendous hypertrophy.

Follow-up Results. One year after operation patient was having one to two bowel movements every day with a normal impulse to defecate. She complained only of occasional slight soiling of underwear with fecal fluid. A follow-up barium enema revealed the complete disappearance of the megacolon with normal evacuation. We were surprised also to find that she no longer had a productive cough. For this we have no adequate explanation.

SUMMARY

The principle of total removal of the nonpropulsive segment of rectum and sigmoid for cure of congenital megacolon is discussed.

A modification of the operative procedure devised by Swenson, embodying this principle, is described.

The results of operative treatment of 30 cases, 23 by the method described, are presented.

These results indicate that the method described is more effective in restoring normal bowel function than previous procedures.

Three cases are presented in detail to illustrate the three types of congenital megacolon.

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