Follow up on 200 Patients Treated for Hirschsprung's Disease During a Ten-Year Period *

ORVAR SWENSON, M.D.
Boston, Massachusetts

From the Boston Floating Hospital for Infants and Children

TEN YEARS AGO it was postulated that Hirschsprung's disease was caused by dysfunction of a terminal segment of colon.[4] The basis of this concept was the observation that a transverse colostomy relieved the patient's abdominal distention. Manometric tracings from the left colon demonstrated normal or hyperactive peristalsis in the dilated colon and absence of propulsive contractions in the rectosigmoid.[5] The aperistaltic segment contained no ganglion cells, whereas, the proximal segment had normal Auerbach's plexus. These facts provided ample explanation for the persistent failures which were seen when the dilated colon was resected. It was further postulated that resection of the normal-appearing, but functionally defective terminal segment of colon with restoration of intestinal continuity would be beneficial. An operation was devised to remove all of the aganglionic segment except the internal sphincter.[6] The results in 200 patients who have been subjected to this type of procedure during the past ten years will be reviewed.

While the long-term results are the most important data for evaluation of a new operation, there are other aspects such as the intricacies of the procedure, the mortality rate, and complications which play a role in an appraisal of a new form of surgical treatment.

Resection of the aganglionic segment as we have accomplished it has proved to be a difficult operation, the success of which depends on meticulous attention to a number of details, such as careful dissection of the rectum and rectosigmoid, arrangement of the proximal colonic segment so that it is of sufficient length with an adequate blood supply, and the performance of an accurate anastomosis. While the procedure has proved to be fairly difficult, well-trained resident surgeons have repeatedly accomplished it with success, and most important, as the subsequent data will confirm, the excellent results more than compensate for the technical difficulties of the operation. All of the patients in this follow up study have been previously reported, except for a small number operated upon recently at the Boston Floating Hospital and New England Center Hospital.[7-10]

MORTALITY RATE

In 200 consecutive patients subjected to resection of the aganglionic segment there have been six postoperative deaths, none due to dehiscence of the anastomosis or sepsis in the pelvic cavity. This figure does not include deaths following colostomies which have been made in preparation for colonic resection. One infant died after a second resection which became necessary because of incomplete removal of the aganglionic segment. An ileostomy was performed to protect an inadequate anas-

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tomosis. Postoperative irrigations of the ileostomy resulted in peritonitis when the irrigating catheter perforated the intestinal wall. Another death followed a small bowel resection for intestinal obstruction. Actually this infant died of agranulocytosis. The third death was due to bilateral adrenal hemorrhage which unfortunately was not recognized until the postmortem examination was performed. Acute gastroenteritis was the cause of death of a fourth infant. A two-and-one-half-year-old boy died of a staphylococcic septicemia which may well have been related to pre- and postoperative antibiotic therapy. In these five cases the diagnosis was based on postmortem examinations. The cause of death in the sixth postoperative fatality was not verified by necropsy. In this patient a postoperative abdominal exploration identified multiple small bowel perforations, and this resulted in a proteus vulgaris peritonitis and wound infection. The cause of the multiple small bowel perforations may have been related to congenital defects in the intestinal muscular wall so that in minute areas the mucosa was adjacent to the peritoneum. This was the pathologic finding in a short segment of ileum examined after resection.

As the series of 200 operations included a number performed on ill and malnourished children, not infrequently with a history of previous surgery, a mortality rate of 3 per cent establishes the operation as acceptable from the standpoint of safety.

It is significant that four out of the six deaths were in infants, and this has prompted us to postpone resections until the children are 12 to 18 months of age, maintaining the patients with colostomy until this age. Further postponement is impractical, for the children at two or two and a half years of age are often in a phase of emotional development when the child's cooperation in colonic elimination may be lacking. The patient's cooperation is so essential to the operation's success that our experience has taught us to perform the resection either before or after the child goes through the period of uncooperativeness.

POSTOPERATIVE COMPLICATIONS

Another facet in appraising an operation is the frequency and severity of postoperative complications. The complications following 200 resections will be listed to answer this question.

The most serious complication was slough of ten inches of terminal colon. The resection on this 17-year-old girl was complicated by dense intra-abdominal adhesions produced by a series of previous operations. The colonic slough was probably related to inadvertent ligation of the marginal vessels during reperitonealization of the pelvis. Following a hectic postoperative course this patient recovered. One year later a reconstruction was accomplished, and the patient has had normal colonic function with sphincter control for the year since the reconstruction was completed.

Another serious complication was in an 18-year-old boy who had excessive postoperative bleeding, probably related to intensive preoperative colonic preparation with antibiotics. Early in the postoperative course re-exploration relieved the abdominal cavity of a large amount of bloody fluid. Eventually a massive pelvic hematoma became infected and drained into the colon. A fistula developed from the infected hematoma in the presacral space to the colon which failed to close until a colostomy was made. This young man is now two years from closure of the colostomy, and he has normal colonic function.

Two gross leaks of the anastomosis early in the postoperative period required emergency colostomy. Both children have been normal since closure of the colostomies. One child developed a pelvic abscess ten days after operation, which drained into the colon and required a proximal colostomy.

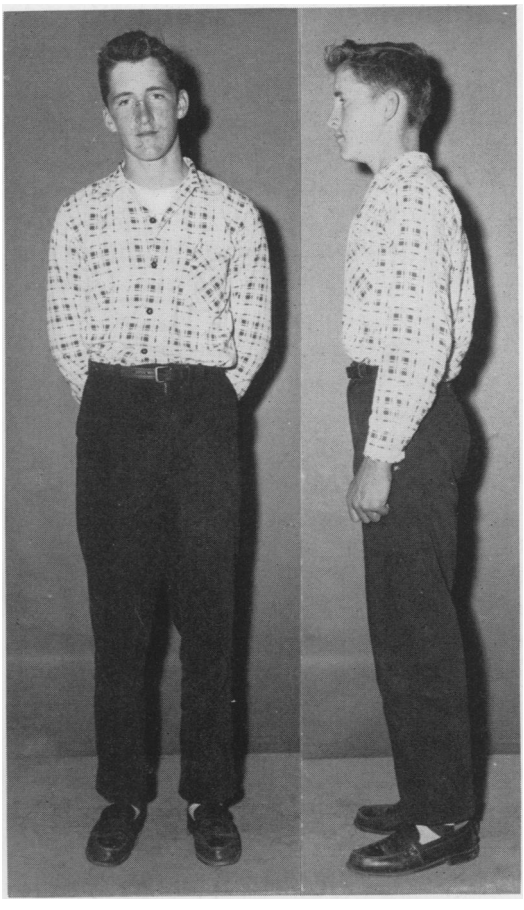

FIG. 1. Photograph of a 16-year-old boy 10 years after resection. He was the first patient in our series. From Swenson, O.: Pediatric Surgery. Appleton-Century-Crofts, Inc. In Press.

A minute rectovaginal fistula occurred in a small girl; it was probably related to a suture being placed too deep so that the vaginal wall was perforated when the anastomosis was being performed. A proximal colostomy was made. No procedure involving the fistula was required and the child has been perfectly well since closure of the colostomy four years ago.

Nine patients have had strictures at the colonic anastomosis. Six of these were in children who had had transverse colostomies, and over a period of time the deactivated colon had become smaller than normal in diameter. It was this small size rather than an inadequate anastomosis which resulted in strictures when the colon dilated to normal size. Two were in infants subjected to primary resection. Eight patients were treated with dilatation and all made good recoveries and have normal bowel function after operation. One stricture was severe and eventually was removed by a second resection. This boy is now perfectly well three years after the last operation.

One infant had been treated with antibiotics before admission to the hospital for resection and developed a Pseudomonas aeruginosa septicemia postoperatively which fortunately responded to neomycin. Two unexpected complications developed, both rather belatedly detected and treated. One infant developed some fever on the seventh postoperative day. Twenty-four hours later abdominal exploration was performed and a perforated appendix was removed. A second child was explored postoperatively with a diagnosis of intestinal obstruction and jejunal intussusception was discovered and reduced.

The remaining complications have been less serious and have consisted of five cases of gastroenteritis, four urinary tract infections, two wound infections, two drug reactions, one intraabdominal abscess, one paralytic ileus, one urethritis, and one transient hemolytic anemia.

RESULTS

The relief of symptoms over a long period postoperatively is the most important data in evaluation of an operation. All but four of the 200 patients subjected to resection have had a follow up study. A majority have been seen and examined by the author. In the remaining patients, examinations by local physicians and reports from the patient's family have been relied upon. Since the time interval from operation is important, the patients will be divided into groups. There are 73 patients followed from five to ten years, 64 from

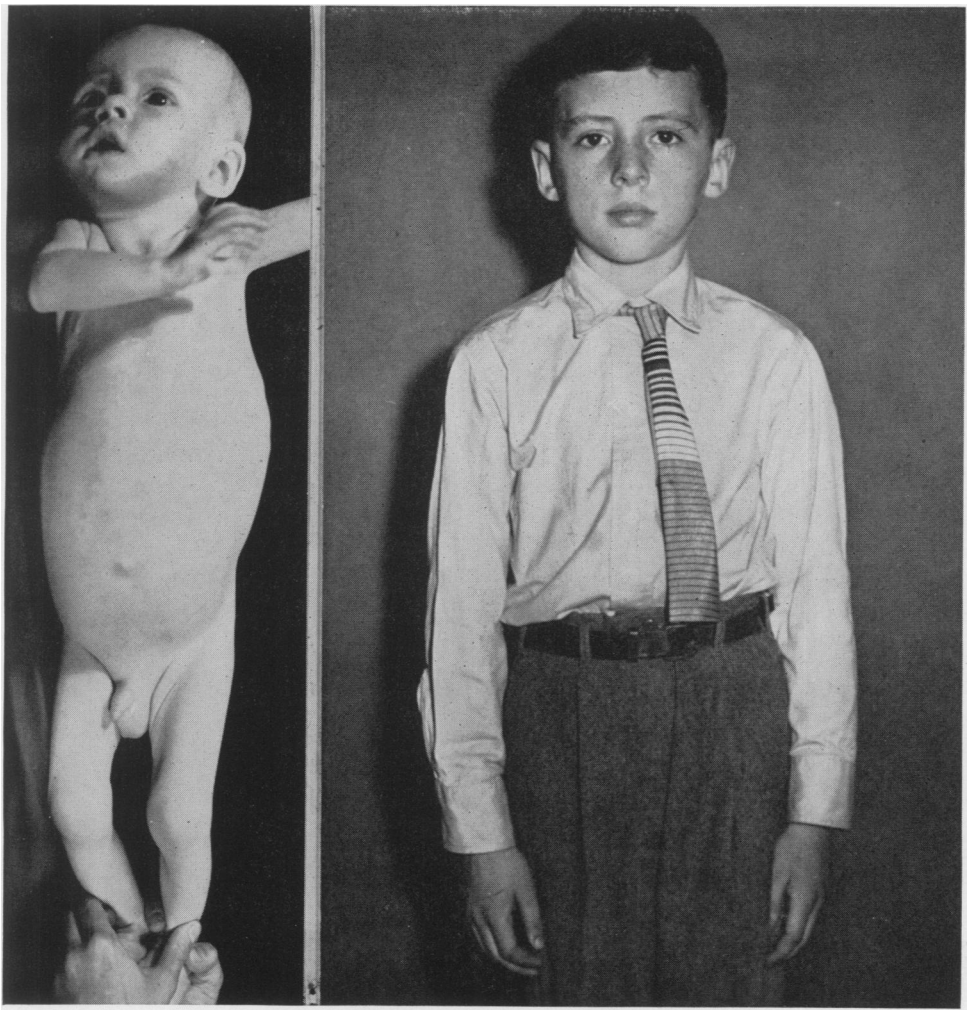

FIG. 2A. Photograph of an infant with Hirschsprung's disease. B. Photograph of same patient 9 years after resection. He is a normal 10-year-old boy.

two to five years, and 52 less than two years.

In the first group of seventy-three patients followed from five to ten years, all but one have been completely relieved of their symptoms and are normal individuals (Figs. 1, 2, 3, and 4). One boy is classed as unsatisfactory because of recurrent attacks of gastroenteritis. While attacks of gastroenteritis have occured in several patients, particularly in the early postoperative course, this child is different in that the attacks still occur at two to three month intervals eight years after operation. The boy's general health is excellent.

Incontinence has not been a problem. In the early postoperative course there have been several cases with soiling which subsided generally before the child was discharged; rarely, the symptoms persisted for two to three weeks after discharge. One boy who was perfectly normal for two years after operation began to soil. In addition, he began to dribble urine and also to stutter. Since soiling developed two years after operation and was associated with other

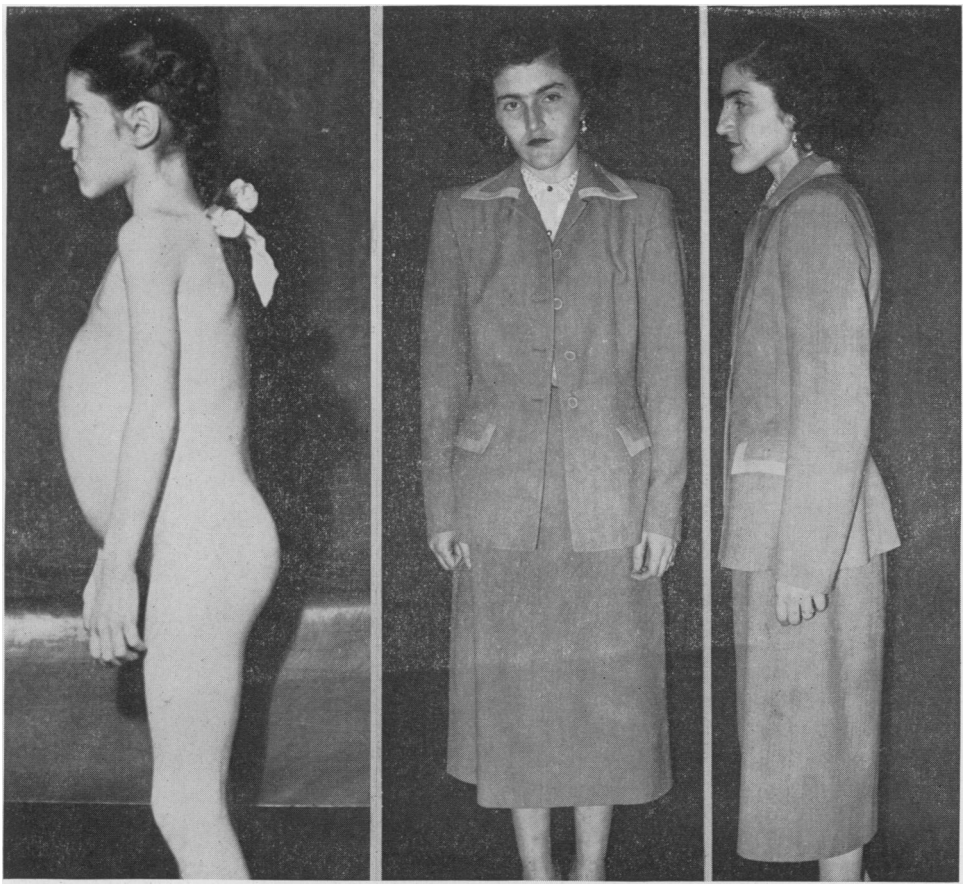

FIG. 3A. Photograph of a 10-year-old girl with Hirschsprung's disease. B. Photograph of same patient 7 years after resection. The patient is married and the mother of 2 children. She has regular colonic function and no abdominal distention.

symptoms of emotional disturbance, the child was treated in a habit clinic and has not soiled for three years. He is classed as a good result. A second boy has had this same type of problem. He had essentially little trouble for a year postoperatively, then began to have occasional soiling. Next he began to wet himself and exhibit infantile behavior. As in the preceding patient, this appeared to be an emotional problem, and with appropriate treatment he is making a satisfactory recovery. Soiling has stopped, but he still wets himself occasionally. He is now classed as a satisfactory result.

Sixty-four patients have been followed two to five years and all except one are perfectly normal. The unsatisfactory result is in a child who did well for one year, then began to have chronic diarrhea and slight distention. He has improved and is now doing well. Since his recovery is still recent he is classed as an unsatisfactory result.

There are 52 patients who have been operated on during the past two years. All are good results except three. One child continues to have some constipation. There is no associated abdominal distention. Enemas are given several times a month.

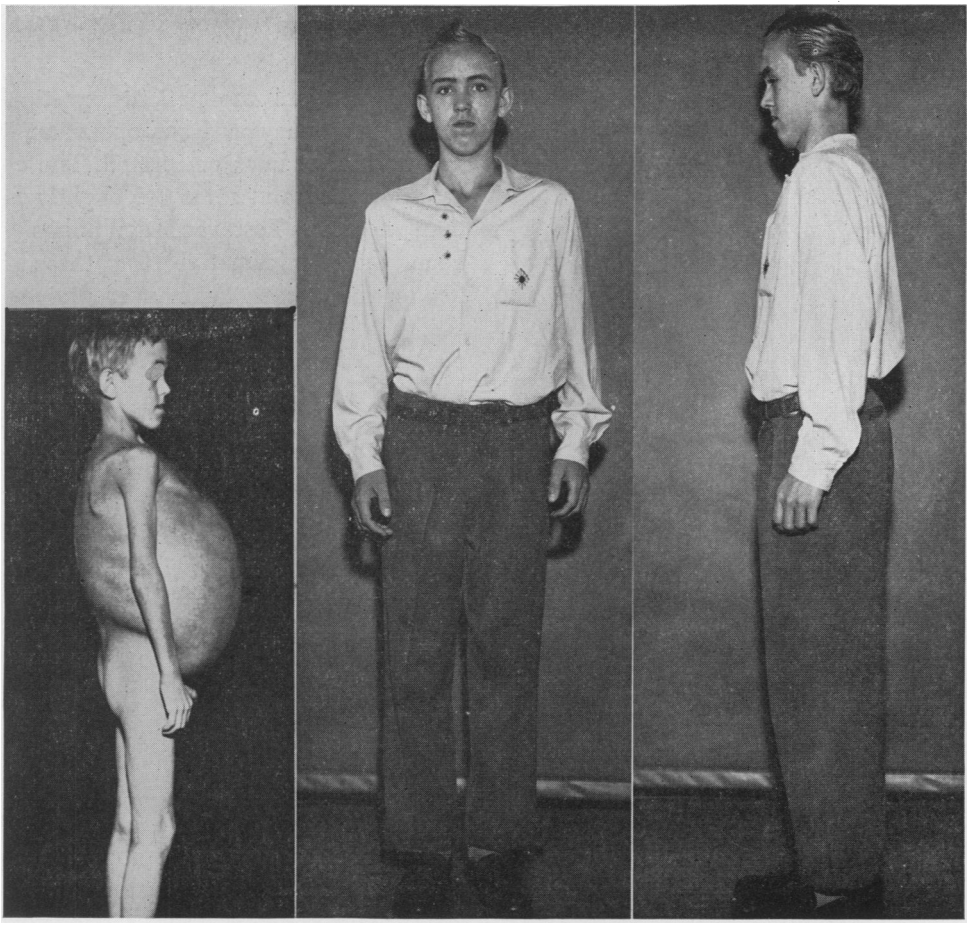

FIG. 4A. Photograph of a boy with advanced Hirschsprung's disease. From Swenson, O.: Pediatric Surgery. Appleton-Century-Crofts, Inc. In Press; and Swenson, O.: J. A. M. A., 154: 651, 1954. B. Photograph of same patient 6 years after operation. He has normal colonic function, and the massive abdominal distention has disappeared. From Swenson, O.: Pediatric Surgery. Appleton-Century-Crofts, Inc. In Press.

A second boy has had recurrent diarrhea and has failed to respond greatly to treatment. A third boy has recurrent diarrhea. Despite the diarrhea, the two children are in excellent health with normal growth and development. These three are considered poor results.

In summary of the patients followed, all are good results except five. Our conception of a good result is a child who is normal in growth and development, who has regular bowel movements unaided by enemata or laxatives, and who has no abdominal distention. It is perfectly true that on rare occasions such as illness, there may be transient constipation just as in any normal child. The majority of the children have had no laxatives or enemata postoperatively. The cause of the chronic recurrent diarrhea is perplexing. Children with this complication have extremely tight sphincters, and this may be related to the complication.

Hiatt [2] in New York, and a group at the Hospital for Sick Children in London,

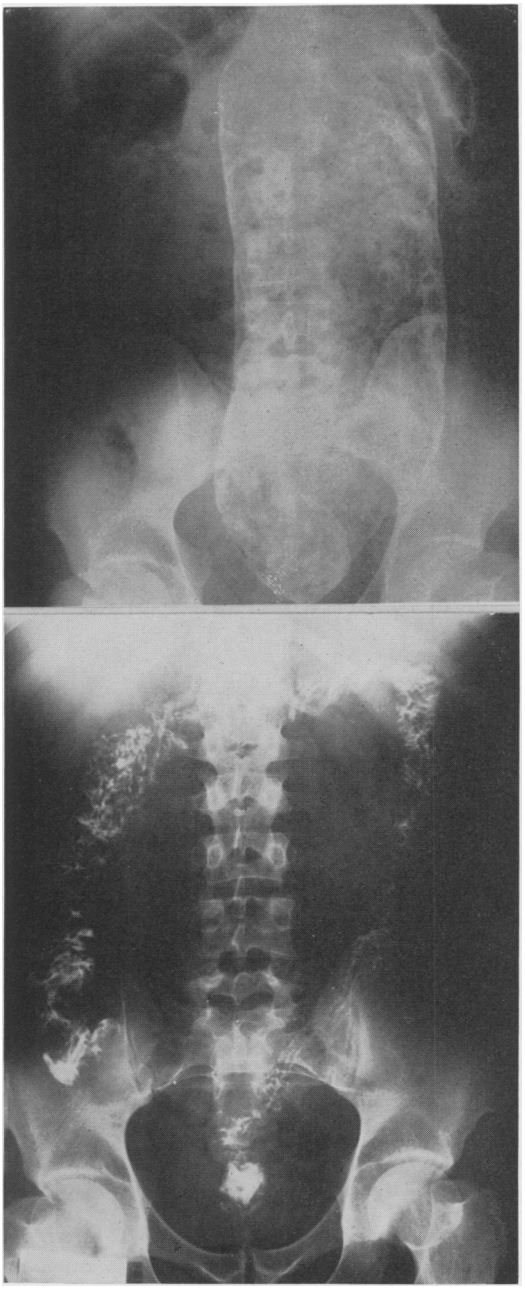

FIG. 5A. Roentgenogram of a young adult with severe Hirschsprung's disease. B. Follow up roentgenogram of same patient. Both roentgenograms were postevacuation. Note the completeness of emptying in the postoperative film. From Swenson, O.: J. A. M. A., **154**: 651, 1954, and Swenson, O., R. H. Segnitz and R. H. Shedd: Am. J. Surg., **81**: 341, 1951.

have used our operative procedure with slight modifications and have reported satisfactory results.[1, 2]

A majority of the patients, particularly those over one year from operation, have had barium enemas performed. The colons are shortened and some are slightly larger than normal. Postevacuation films have been most informative for these have demonstrated satisfactory emptying. Inability to empty has been a constant preoperative finding (Fig. 5).

The most distressing finding in this follow up study is that one to five years from operation seven children have died suddenly following illnesses of less than 24 hours' duration. In all but one a postmortem examination was performed, and infection with severe dehydration has proved to be the cause of death.

This experience plus the fact that such sudden deaths occur in patients prior to operation suggests that some infants, and to a less extent children, with Hirschsprung's disease have some defect in the normal mechanisms to combat infection. In addition, there may be abnormalities in the autonomic control of water balance and electrolytic concentrations to account for the extreme dehydration observed in the patients after illnesses of less than 24 hours. One older boy died of renal failure with atrophic malformed kidneys.

Seven patients have come under our care who have had segmental resections of the left colon with anastomosis of the transverse colon to the rectosigmoid at the level of the pelvic peritoneal reflection.[3] Following these operations there was persistence of symptoms, and they came for further treatment (Fig. 6). This type of operation has been advocated on the supposition that a more extensive operation as we have used would result in loss of ejaculation in males.

In our 200 resected patients there have been eight males who have married and

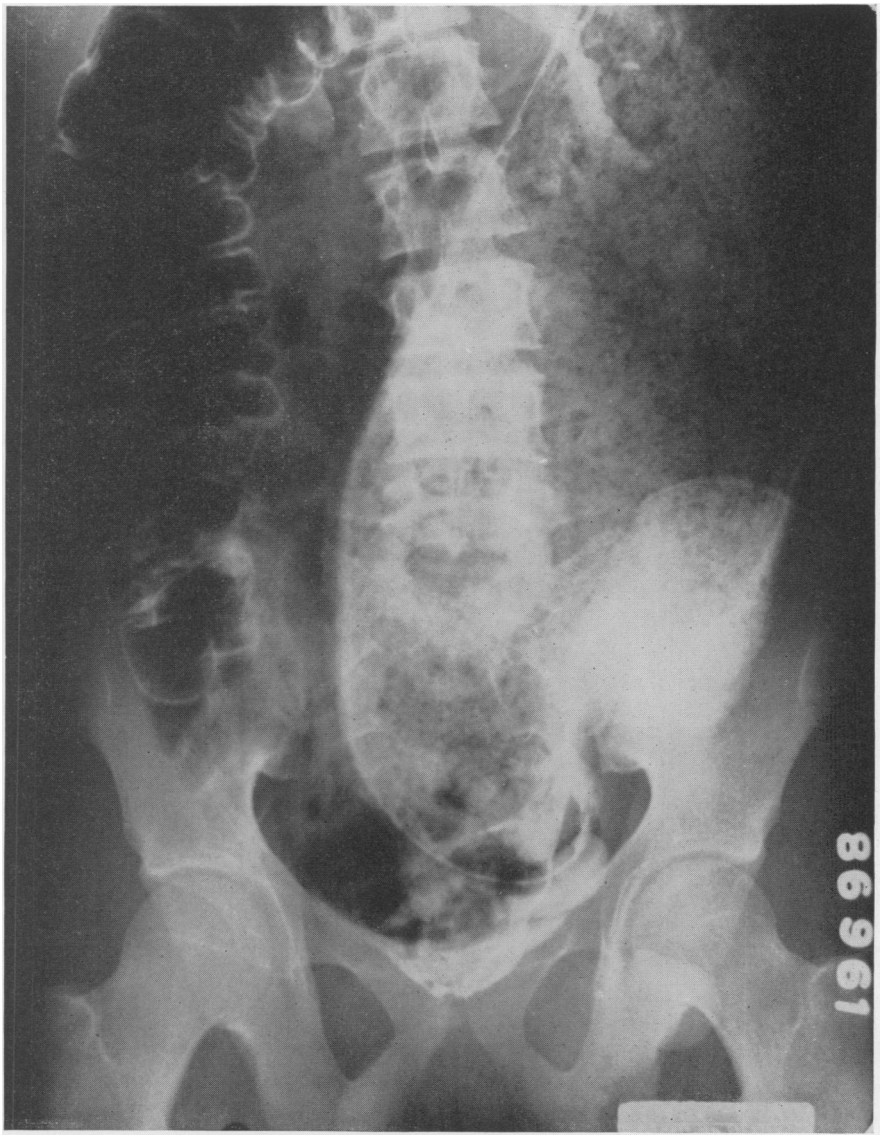

FIG. 6. Roentgenogram of a patient who had segmental resection with an anastomosis at the level of the pelvic peritoneal reflection. Symptoms persisted following this incomplete resection. From Swenson, O.: Pediatric Surgery. Appleton-Century-Crofts, Inc. In Press.

become fathers of children. There are six men over 20 who are not married. None claim a defect in ejaculation. Consequently this does not seem a valid reason for advocating segmental resection, particularly since there is incomplete relief of symptoms following such operations. Furthermore, resection of the remaining aganglionic colon has been difficult and reconstruction almost impossible, due to the short length of proximal colon. In several the right colon has been utilized, turning it into the pelvis for the reconstruction.

Since removal of the pathologic colon

down to within two centimeters of the mucocutaneous margin can be accomplished without disturbance of ejaculation or fecal continence, and since failure to accomplish this fails to relieve the patient's symptoms, it seems that resection of the aganglionic segment to within two centimeters of the anal canal is the operation of choice.

SUMMARY

1. In a group of 200 resections for Hirschsprung's disease, there were six postoperative deaths.

2. The postoperative complications in 200 patients have been listed.

3. In the follow up there are five out of a total of 200 patients who are classed as unsatisfactory results.

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