New modification of the surgical treatment of Hirschsprung's disease

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A surgical landmark was passed in 1948 when Swenson and Bill[11] established the pathophysiology of Hirschsprung's disease and successfully applied the "pull-through" operation of Toupet in its treatment. During the 15 years since that time, many surgeons have satisfactorily utilized this procedure in hundreds of cases, but time has proved that it is an operation of great magnitude requiring a tedious and meticulous dissection by an able, patient, and experienced surgeon. Swenson has emphasized that complications will diminish and results improve with the experience of the surgeon, and this indeed has been the author's observation. The problems associated with extensive pelvic dissection, excision of the rectum and its sensory receptors, preservation of a segment of aganglionic bowel below normal colon (though it be only 1 to 2 cm. in length), and a low end-to-end anastomosis have, however, prompted a continuing search for a superior operation. The more limited abdominal resection of State[10] and the rectosigmoid myotomy of Weiss and Hollender[13] have received little acceptance, though of at least temporary value in reported cases. The operation described by Duhamel[3] in 1956, and later modified by Grob[4] and Martin and Altemeier,[7] has stimulated great interest both in Europe and the United States, and a wide experience with this procedure is being reported.

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Editor's Note: For discussion of this technique and its utilization, see article by Soave in this issue.

Duhamel's technique[3] obviates the extensive pelvic dissection, creates a large side-to-side anastomosis, retains the sensory reflexes of the rectum and, as originally described, had normally innervated colon right down to the anocutaneous junction. It can be further utilized on cases of "failed" Swenson's operation and is performed in the neonatal period. These advantages would appear to make this operation an almost perfect solution to the problem of Hirschsprung's disease, but complications of incontinence and the occurrence of fecalomas in the retained rectum have been reported. Grob[4] modified the procedure to decrease incontinence by starting his side-to-side anastomosis above the internal sphincter. Martin and Altemeier[7] further modified it by extending the anastomosis the entire length of the retained rectal pouch. The need for continuing modifications and the difficulties being reported with Duhamel's operation have encouraged further exploration for a better surgical approach to the cure of congenital megacolon.

The author has long considered the possibility of solving the problems of Swenson's operation by bringing proximal normal colon through the lower portion of the aganglionic rectum. This would preserve the sensory reflexes of the rectum, bring normal propulsive colon down to the anocutaneous junction, and would not require any pelvic dissection. A stumbling block has always been the need for retaining the mucosa of the distal rectum which was considered necessary for preservation of the anorectal reflex.

However, recent studies by Karlan[6] and Brody and co-workers[1] have established that this reflex persists after removing the mucosa. These reports suggested that a procedure utilizing a modification of Ravitch's anal ileostomy[9] would permit the application of the principle of bringing the normal colon through the aganglionic rectum. The successful use of a similar technique in the treatment of imperforate anus with rectourethral fistulas[8] has encouraged me to propose the following surgical approach for a trial in the treatment of Hirschsprung's disease.

PROPOSED PLAN OF THERAPY

1. Preliminary transverse colostomy and establishment of level of aganglionosis. This step will be necessary to decompress the distended ganglionic colon and facilitate bringing it through the normal-sized aganglionic segment. Since, following proximal colostomy, it is impossible to estimate the transition area, biopsies can be done at the time of colostomy and the level of later resection determined and marked. This procedure could be carried out through a right paramedian incision, reserving the left side for the definitive operation.

2. Definitive resection and pull-through procedure. This operation would consist of three major steps and could be performed through a left paramedian incision. Step 1 would be the resection of the aganglionic colon above the peritoneal reflection and mobilization of the normal colon to permit anastomosis to the anus without tension. The transverse colostomy would not be disturbed.

Step 2 would consist of removal of the mucosa of the distal rectum. This could best be carried out by combining the techniques described by Ravitch[9] and Nicolai and Rehbein.[8] The latter begin their dissection from above, establishing the correct plane from the open end of the rectum just below the peritoneal reflection. After carrying this as far as can be done easily, the remainder of the dissection can be left for the perineal portion of the procedure.

Step 3 would be the completion of the mucosal resection and the anocolonic anastomosis. The child being positioned for an abdominal perineal procedure, the perineum would then be prepared. Dilatation of the anus and rectum would be done and, according to Ravitch's technique,[9] an incision would be made through the mucosa at the anocutaneous line and the remainder of the dissection of the mucosa completed. After removing the mucosa, the rectal remnant would be dilated vigorously and the proximal colon brought through it and anastomosed to the anus in the manner described by Ravitch. A layer of sutures between the serosa of the normal colon and the upper end of the rectal muscular shell would further fix the bowel.

3. Closure of the transverse colostomy. This would be performed soon after recovery from the definitive operation and requires no discussion.

DISCUSSION

The advantages of this proposed operation are: *(A)* The absence of any pelvic dissection; *(B)* the preservation of the rectum and its sensory receptors; *(C)* the presence of normal propulsive colon right down to the anus; *(D)* the diminished expectation of anastomotic leaks in this type of operation; *(E)* the preservation of all sphincters; and *(F)* the absence of the rectal pouch (as in the Duhamel procedure) in which fecalomas might form.

The disadvantages are: *(A)* The need for preliminary colostomy, *(B)* the mucosal dissection (which is unnecessary in Duhamel's operation), *(C)* the presence of an end-to-end anastomosis with its potential of stenosis, and *(D)* the theoretical danger that the aganglionic rectal-muscle shell will act as a block by contracting.

The preliminary colostomy might prove unnecessary in neonates in whom proximal dilatation has not occurred, but Nicolai and Rehbein's[8] experience with their operation in this age group has not been good. However, colostomy is also required in infants awaiting Swenson's operation, and the majority of cases are now being diagnosed soon

after birth. The possibility of stenosis can be prevented by dilations, and this complication was not a problem in Ravitch's cases.

The major question in this operation is the action of the retained rectal muscular cuff. As stated there is ample evidence that this segment retains its sensory receptors but its contractile activity can only be determined by clinical trial. We have tested resected portions of aganglionic colon for distensibility and the presence of fibrosis. No fibrosis was seen and the segments could be distended easily but not to the diameter of the dilated segments. The latter findings would be expected of course. The term "spastic segment," which has been applied to the aganglionic colon, raises the picture of this lower segment contracting around the normal colon brought through it. Studies by Swenson and associates,[12] Hiatt,[5] and Davidson and co-authors[2] have shown that the motility pattern of the resting aganglionic segment is indistinguishable from the normal rectum. Swenson, using a balloon-bymographic technique, reported diminished to absent activity in the "contracted" segment. The mass contraction described when a peristaltic wave reaches this area should not occur when the rectum is taken out of continuity.

The absence of aganglionosis in the experimental animal, with the exception of mice, necessitates trial of new procedures in human subjects. The technical aspects of the operation proposed have all been previously established both in the laboratory and clinically in conditions other than Hirschsprung's disease. It is the author's feeling that this operation offers potential advantages over both the Swenson and Duhamel procedures and is worthy of clinical evaluation. We hope to employ it on our next suitable patient and would hope that other surgeons who see merit in this technique will similarly investigate its potential value.

SUMMARY

A new operative approach for the treatment of Hirschsprung's disease is proposed. The essence of the procedure is the bringing down of normal colon through a retained segment of aganglionic rectum and an anocolic anastomosis within this muscular tube. The rectal mucosa is entirely resected. The advantages and disadvantages, apparent and theoretical, are discussed.

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