Endorectal Pull-Through Procedure for Hirschsprung's Disease with and without Primary Anastomosis

By Scott J. Boley, Dennis J. Lafer, Sylvain Kleinhaus, Bertram D. Cohn, Ascher L. Mestel and Peter K. Kottmeier

THE BREAKTHROUGH in the treatment of Hirschsprung's disease occurred in 1948 when the distal improperly innervated segment was identified as the site of obstruction, and Swenson and Bill successfully applied a pull-through form of resection of the nonpropulsive segment.[1] Wide experience has proved the efficacy of this operation, and it remains the yardstick against which all new procedures must be measured. Numerous reports, however, have revealed significant mortality and morbidity figures, and Swenson himself describes the technic as "extremely difficult" and "long and tedious."[2] Good results can be expected when the operation is performed by able and experienced surgeons, but 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 or 2 cm. in length), and a low end-to-end anastomosis have prompted a continuing search for new procedures.

In 1956 Duhamel reported the retrorectal transanal pull-through procedure to solve the problem of megacolon in early infancy.[3] This technic has been employed successfully in neonates, avoiding the need of a preliminary colostomy, but recently several authors have again recommended colostomy, with delay of the definitive procedure. Duhamel's operation and its numerous modifications, substitute exclusion, or bypass, of the distal segment for resection, and create a long side-to-side rectocolonic anastomosis. This obviates the extensive pelvic dissection and retains the sensory side of rectal reflexes. As originally described, normal innervated colon was brought down to the mucocutaneous junction, but incontinence prompted modifications which retain

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From the Pediatric Surgical Services, Department of Surgery, State University of New York, Downstate Medical Center, Brooklyn, N. Y. and Montefiore Hospital and Medical Center, Bronx, N. Y.

Scott J. Boley, M.D.: *Attending-in-charge of Pediatric Surgery, Montefiore Hospital and Medical Center, Associate Professor of Surgery, Albert Einstein College of Medicine.* Dennis J. Lafer, M.D.: *Instructor of Surgery, State University of N. Y., Downstate Medical Center. Adjunct-in-charge Pediatric Surgery, Jewish Hospital and Medical Center of Brooklyn.* Sylvain Kleinhaus, M.D.: *Chief Resident in Surgery, Jewish Hospital and Medical Center of Brooklyn.* Bertram D. Cohn, M.D.: *Lecturer in Surgery, State University of N. Y., Downstate Medical Center, Surgeon-in-charge Pediatric Surgery, Maimonides Medical Center.* Ascher L. Mestel, M.D.: *Clinical Assistant Professor of Surgery State University of N. Y., Downstate Medical Center. Attending-in-charge Pediatric Surgery, Brookdale Hospital Center.* Peter K. Kottmeier, M.D.: *Associate Professor of Surgery and Chief, Pediatric Surgical Service, State University of N. Y., Downstate Medical Center.*

all or part of the internal sphincter below the anastomosis. Satisfactory results have been reported from many centers but problems have arisen from the rectocolonic spur and from fecalomas forming in the large rectal pouch. The need for prolonged immobilization while the spur is being crushed is an undesirable feature of the technic.

Several years ago Soave described the application of an endorectal bypass of the aganglionic rectum without an anastomosis[4] and one of us proposed a similar approach with a primary anocolic anastomosis.[5] In both of these operations the mucosa of the aganglionic rectum is removed, the aganglionic muscular cuff with its sensory reflexes is retained, and normal propulsive colon is brought through this cuff and down to the mucocutaneous junction. The internal sphincter remains intact for continence but is bypassed from within by normal bowel. Since the entire pelvic dissection is performed within the musuclar sleeve, possible damage to the pelvic nerves is avoided.

Since 1964 we have performed 13 endorectal pull-through operations for Hirschsprung's disease. Eight children, 8 months to 3 years of age, had primary anastomoses as proposed by Boley, 2 as an initial procedure, the others preceded by a colostomy. Four children, 5 months to 5 years of age, had a Soave procedure, 2 as an initial operation and 2 after a preliminary colostomy. In one child a Soave operation was used after an unsuccessful Duhamel procedure. A three-stage procedure was used in the first 2 patients having primary anastomoses, retaining the colostomy for protection, but experience showed this to be unnecessary and subsequent patients have had one-stage or two-stage procedures. In 11 patients only the rectum and rectosigmoid were involved, while long-segment involvement required hemicolectomy in 2.

The endorectal pull-through procedures consist of an abdominal and a perineal phase. Abdominal mobilization of the colon, preparation for resection, establishment of level of aganglionosis, and taking down of the colostomy (if present) are performed in the same manner as for a Swenson procedure, except for careful preservation of the blood supply to the rectum and rectosigmoid. The resection of the rectal mucosa is begun 4–8 cm. above the peritoneal reflection and carried down as far as possible. The perineal phase is then begun through a transanal approach. An incision is made 0.5–1 cm. above the pectinate line and the rest of mucosal stripping completed. (If the Soave nonanastomotic operation is to be used, the rest of the perineal phase follows his described technic. Except in unusual cases, the primary anastomosis is now preferred.)

The proximal bowel is then pulled through the rectal muscular sleeve and out the anus. The site of anastomosis, previously chosen and prepared intra-abdominally, is positioned without tension. A Penrose drain is then placed between the colon and rectal muscular cuff and brought out through a vertical stab wound posterior to the external sphincter. Four to 6 fixation sutures are placed between the rectal muscular cuff and the seromuscular layers of the intussuscepted colon. The colon is then transected by quadrants and an anastomosis performed between the anal mucosa (0.5–1 cm.) and all layers of the colon.

The abdominal phase is completed by loosely approximating the proximal edge of the muscular cuff to the seromuscular layer of colon.

Results

There were no postoperative deaths in this series. Technically, the operations have proved to be relatively easy. The mucosal dissection is the only unfamiliar step and this is readily mastered. If the colon has never been hypertrophied, the mucosa can be stripped down to the anus with minimal blood loss and with few, if any, vessels requiring control.

The only complication related to the technique of the operation occurred in the group having the Soave procedure. The colon retracted after amputation of the stump on the twentieth day. The subsequent stricture responded to prolonged dilation. There have been no anastomotic leaks or strictures following primary anastomosis. Postoperatively, all children have had normal bowel movements except for a mentally retarded girl with a Soave procedure who continues to be constipated.

The postoperative care of those patients without anastomoses has provided some difficulties, and these children have usually been hospitalized for 3–4 weeks after operation. Prolonged dilations are necessary to prevent stricture formation.

The postoperative course following primary anastomosis has been exceptionally smooth, and the major drawbacks of the Soave procedure appear to be obviated. Prolonged hospitalization and care required by children awaiting amputation of the colonic stump are avoided. Repeated dilatations necessitated by the edema following the Soave procedure have not been necessary after primary anastomosis except where a proximal colostomy was left in place. In such cases, dilatation has been essential until the colostomy was closed.

Discussion

The basic technic of removing rectal mucosa while retaining the muscular cuff was first described by Ravitch and Sabiston.[6] Simonson, Habr and Gazal and others have employed the principle in acquired and congenital megacolon with reported good results.[7]

Our experience with the endorectal pull-through with primary anastomosis suggests that this is a simple, safe and physiologically sound approach to the treatment of Hirschsprung's disease. Since our average postoperative follow-up is only 1–2 years, a long-term evaluation is impossible, but the operative and early results are most encouraging.

Both the Duhamel and Swenson operations have the problem of how to handle the internal sphincter. If all or too much of this area is resected or bypassed, incontinence has resulted, while if too much is left behind in continuity, persistence or recrudescence of symptoms has ensued. Endorectal bypass avoids this problem by bringing propulsive colon to the mucocutaneous junction while preserving the integrity of the sphincters.

Several important theoretical objections to the endorectal technic have been raised. The danger that the aganglionic rectal muscular cuff would act as a

block by active contraction or fibrosis has not been realized. A biopsy done one year postoperatively revealed the muscle cuff to be viable and adherent to the enclosed normal colon. The failure of any contractile obstruction to materialize was anticipated as the distal aganglionic segment is not really "spastic" but rather has a normal resting motility pattern. Functional obstruction results from the absence of propulsive activity in the involved segment and perhaps by a mass contraction of the segment when a peristaltic wave reaches it. Such mass contraction should not occur when the rectum is taken out of continuity.

Another criticism of the procedure is that it removes the receptor area in the rectum which is necessary for coordinated defecation and continence. This objection is based upon the assumption that the afferent or sensory portion of the rectosphincteric reflex arc resides in the resected rectal mucosa. While the children in our series are too young to determine if the anorectal reflex is retained, there is equal or greater experimental and clinical evidence indicating that the sensory receptors are in the muscular layers rather than in the mucosa.[8-10] Based upon Swenson's criterion for sphincteric control in children not yet toilet trained, i.e., the mother's observation that the child is perfectly clean between bowel movements (2–3 times per day),[2] incontinence does not appear to be a problem in any of our patients. Soave has followed 24 patients for 5 or more years after endorectal pull-through and reports the "motility and function of the lower colon is excellent." He has now performed his nonanastomotic technic on 61 patients, none of whom have developed enterocolitis since operation.

We have not as yet employed the endorectal pull-through operation in the neonatal period, but the ease of the mucosal dissection when colonic hypertrophy has not developed has encouraged us to do so. Experience with stillborns indicates that the dissection is remarkably easy even in small infants. Certainly, if the definitive procedure is to be delayed, early colostomy is indicated to prevent enterocolitis and facilitate the later dissection. The colostomy is best placed in the most distal ganglionic bowel and is resected at the time of the endorectal operation.

Summary

The advantages of the endorectal pull-through procedure with primary anastomosis are: (1.) The absence of any pelvic dissection; (2.) the presence of normal propulsive colon right down to the anus; (3.) the preservation of the rectal muscular cuff and its sensory receptors; (4.) few, if any, anastomotic problems; (5.) the preservation of all sphincters and (6.) simple postoperative care. Although more time and experience are required to evaluate the procedure, we believe that its relative ease of performance, inherent safety, and physiologic soundness may make it the operation of choice for Hirschsprung's disease.

SUMMARIO IN INTERLINGUA

Le technica del "transtraction" endorectal – con e sin anastomose primari – esseva usate in dece-tres juveniles con morbo de Hirschsprung. Le resultatos immediatemente

chirurgic e brevitemporalmente catamnestic esseva satisfacentissime. Le facilitate del operation e le rationalitate physiologic de illo in combination con anastomose primari promitte facer de illo le tractamento de election in casos de morbo de Hirschsprung.

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