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The Surgical Management of Ulcerative Colitis, Familial Polyposis and Total Colonic Aganglionosis in Infants and Children

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Abstract

The endorectal pull-through, a sphincter-saving operation, has become the procedure of choice for the definitive management of ulcerative colitis and familial polyposis in both children and adults for the past fifteen years. Controversies exists as to whether this procedure should be combined with a reservoir when the entire colon has been resected. At our institution, we have performed a total colectomy, a mucosal proctectomy and a straight ileo-anal and pull-through for the above two diseases without a pouch or reservoir because the functional results are similar to those with a pouch and the complications are far less. This same operation has also been carried out in our institution for the management of total colonic Hirschsprung's Disease.

This manuscript focuses on the operative technique for performance of the total colectomy and ileo-anal endorectal pull-through and the results of this operation in 116 patients with ulcerative colitis and 30 patients with familial polyposis. In addition, our experience with 18 children with total colonic Hirschsprung's Disease is also reviewed.

The final portion of the paper compares our results with the straight endorectal pull-through with those of the endorectal pull-through combined with a reservoir and demonstrates that the clinical and functional outcomes are not different.

Introduction

Martin's landmark report in 1977 ushered in a new era in the management of ulcerative colitis. Most centers initially used the straight pull-through method for patients with ulcerative colitis and familial polyposis. However, disenchantment with high stool frequencies and a significant incidence of complications promoted these various groups to develop a variety of reservoirs to increase rectal capacitance and, thereby, decrease stool frequency. In essence, a Kock-type pouch was added to the pull-through. The theoretical and physiologic advantages of a reservoir have been reviewed by Wong et al. With the introduction of the reservoirs, however, come a number of complications not seen with the straight endorectal pull-through (ERPT), such as pouchitis and a higher incidence of pelvic sepsis.

Our experience with the straight ERPT at the University of Michigan Medical Center has been quite different than the experience reported by other centers, with a stool frequency comparable to that seen with reservoirs and an incidence of complications lower than that reported with the various pouches. Because this group of patients represents the only large series of straight ERPTs in children and adults, we have reviewed our experience with this procedure so that our results can be compared with the many...
series using different types of reservoirs.

Materials and Methods

Between July 1977 and June 1996, 164 children and adults (116 with ulcerative colitis, 30 with familial polyposis, and 18 with total colonic Hirschsprung's disease) underwent total colectomy and straight ileoanal ERPT under the direction of the author. Patients who had not yet undergone closure of their temporary loop ileostomy were excluded from this review. The mean age at operation was 20.6 ± 9.8 years (range, 1 to 48 years). The mean age of the children with Hirschsprung's disease at the time of operation was 17 months and the mean age of the children and adults with ulcerative colitis and familial polyposis was 22.8 ± 10.0 years at the time of surgery. There were 95 female and 69 males patients.

In all patients the diagnosis was confirmed histologically after sigmoidoscopic or colonoscopic examination. In the children with Hirschsprung's disease, the diagnosis was made with a suction rectal biopsy.

The operative technique has been described in detail in previous reports. The mucosal proctectomy is entirely carried out from the abdominal approach, with removal of an intact mucosal and submucosal tube. A 4- to 5-cm rectal muscular cuff is left and the ileoanal anastomosis is performed 1 cm above the dentate line. The anastomosis is performed with absorbable polyglycolic acid sutures and the muscular cuff is drained with a sump suction catheter for 24 to 48 hours after operation. The top of the rectal cuff is tacked to the pull-through ileum with silk sutures. Before closure of the abdomen, a temporary loop ileostomy is created in the right lower quadrant of all patients with ulcerative colitis and familial polyposis. None of the children with Hirschsprung's disease underwent a diverting ileostomy. The ileostomy was closed 2 to 3 months after the ERPT.

Follow-up was complete in this group of 164 patients and involved monthly visits for the first 6 months, visits every 3 months for the remaining 6 months, and then yearly visits. Sigmoidoscopy was carried out on all patients with ulcerative colitis yearly and every 6 months in patients with familial polyposis; biopsies were taken to detect dysplastic changes in patients with ulcerative colitis and any evidence of new polyp formation or malignant degeneration in the case of familial polyposis.

Stool frequencies were compared at various time intervals and between age groups and diagnosis using...
Table 4  Stool Frequency (Per 24 Hours)

- All Patients-7.4
- Familial Polyposis-7.0
- Ulcerative Colitis-7.4

the paired Student's t test.

Results

The follow-up has ranged from 3 months to 20 years. There have been two deaths in the series (operative mortality rate, 1%). The first occurred in a teenager with ulcerative colitis and pericholangiolitic hepatitis who developed fulminate hepatic failure 8 months after surgery. The second was in a 13-year-old girl who had an initial diagnosis of ulcerative colitis and underwent a colectomy and an ERPT. After closure of her loop ileostomy, she developed severe diarrhea and intestinal obstruction requiring multiple operations. Eventually she developed multiple enterocutaneous fistulas and invasive sepsis and died 25 years after her ERPT. Histologic evaluation of her small intestine at autopsy revealed extensive Crohn's disease. Fourteen patients (8%) developed adhesive intestinal obstruction, seven (4%) of whom required an enterolysis. Two patients developed pelvic abscesses, which required operative drainage, and a third patient developed a pelvic phlegmon that resolved with intravenous antibiotics. Rectovaginal fistulas developed in two women and both healed with temporary intestinal diversion. There were five minor wound infections.

In six patients mild narrowing of the ileoanal anastomosis was found and this responded easily to early digital rectal dilatation. There were no anastomotic leaks encountered at the ileoanal anastomosis. No patients developed clinical or histologic evidence of pouchitis during the entire follow-up period.

Five patients (3%) were converted to a Brooke ileostomy. One additional patient with familial polyposis was found to have an anaplastic carcinoma at the anal margin of the mucosal proctectomy specimen on histologic examination and required abdominal perineal resection 1 week after the ERPT. One of the five patients, a 10-year-old girl, developed recurrent symptoms of inflammatory bowel disease after her ERPT and colectomy. The histologic diagnosis of the colon and terminal ileum specimens was ulcerative colitis. A repeat biopsy of her small intestine revealed Crohn's disease and she was converted to an end ileostomy. A second patient underwent a sigmoidoscopy at another institution shortly after closure of his ileostomy, at which time the pull-through ileum was separated from the rectal cuff. He developed a huge rectoperineal fistula that was treated with an end ileostomy. Only three patients (2%) were converted to an ileostomy because of dissatisfaction with their stool frequency (between 10 and 15 per 24 hours). All three patients were continent during the day and at night.

Daytime continence was achieved in all but two patients within 3 months after ileostomy closure, and reached 100% 1 year after operation. Seven patients (4%) experienced minimal nighttime soiling during the first year after ileostomy closure; this nocturnal leakage disappeared 2 years after surgery.

Stool frequency progressively declined after operation, reached a value of 7.4 ± 4.3 per 24 hours 3 years after the pull-through (Fig. 1). All patients were able to evacuate spontaneously immediately after ileostomy closure. Stool frequency was also analyzed by age groups and diagnosis. There were no statis-
Figure 1  The stool frequency for the entire series decreased during the 3-year period to a value of 7.4 ± 4.3 per 24 hours.

Figure 1

A review of the world literature in 1985 revealed that more than 500 ERPTs had been performed for ulcerative colitis and polyposis, the large majority of which involved a reservoir25. During the decade from 1985 to 1995, probably over 5,000 cases have been done worldwide, but mostly in North America26-51. Because no institution has done enough straight ERPTs to adequately evaluate and compare with their reservoir procedure, and because almost all the straight ERPTs were done during each center's early experience with the entire procedure, when the learning curve was steep, it is impossible to equitably compare the two procedures. The stool frequencies reported with the various reservoirs have ranged from 4 to 9 per day (5 to 7 for the J pouch23-41, 16-21, 23, 37-39, 51-53, 59-60, and 6 for the lateral ileal reservoir25-46). We have evaluated the various reservoir procedures with the straight ERPT in an experimental model and found that there were no differences in any of the clinical and physiologic parameters studied between the various reservoir procedures and the straight ERPT25.

The incidence of pelvic sepsis in the various series of reservoirs reported ranges from 3% to 20%2, 3, 8, 9, 13, 15, 17, 18, 20, 21, 23, 33, 34. This high incidence is not surprising in view of the long suture line that must be placed within the rectal cuff with the various reservoir procedures. In the current series, pelvic abscess occurred in two patients (1%), both of whom required operative drainage; and a pelvic phlegmon, responsive to antibiotic therapy, occurred in a third patient.

Discussion

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Clearly the most serious complication of the reservoirs is pouchitis. The incidence of this complication varies from 5% to 25%9, 10, 13, 15, 17, 18, 20, 21, 23, 33, 34. This is one complication that can be completely avoided with the straight ERPT; none of our patients developed any evidence of pouchitis. Nicholls found that 90% of the reservoirs that were subjected to biopsy showed evidence of chronic and acute inflammation. All of our patients have undergone an annual or semiannual biopsy of their pull-through ileum; none of these biopsies have shown any significant inflammation.

Adhesive intestinal obstruction is a significant complication in all the series reported, including our own. The incidence ranges from 8% to 35%, with a significant number of these patients requiring an enterolysis8, 10, 12, 13, 15, 17, 20, 22. Spontaneous evacuation has been noted to be a problem for patients who have undergone the S reservoir. One series reported a 53% incidence of inability to spontaneously evacuate, and another series reported an 11% incidence of this25, 26. Although blood was commonly used for the first 50 patients in this series, only 10 of the second 50 patients transfusions during the operative procedure and none of the recent 64 received blood intraoperatively. In all cases except one, only one unit of blood was administered.
Table 7: Summary of results and complications of the most recent series of ERPT’s for ulcerative colitis and familial polyposis.

<table>
<thead>
<tr>
<th>INSTITUTION</th>
<th>AUTHOR, DATE</th>
<th># OF PATIENTS UC/FP</th>
<th>RESERVOIR</th>
<th>STOOL FREQ /24 HOURS</th>
<th>INCONTINENCE (%) DAY</th>
<th>INCONTINENCE (%) NIGHT</th>
<th>INCIDENCE OF POUCHITIS (%)</th>
<th>CONVERSION TO ILEOSTOMY</th>
<th>POUCH REMOVAL (%)</th>
<th>COMPLICATIONS (%)</th>
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<tr>
<td>Mayo Clinic</td>
<td>Kelly, 92</td>
<td>971/62</td>
<td>J</td>
<td>7</td>
<td>3</td>
<td>10</td>
<td>31UC: 0FP</td>
<td>4</td>
<td>2</td>
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<td>U of Toronto</td>
<td>Cohen, 88</td>
<td>450/24</td>
<td>J257:224S:2W</td>
<td>7J: 5FP</td>
<td>0</td>
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<td>1</td>
<td>4</td>
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<td>Vedianheimer, 93</td>
<td>382/78</td>
<td>J94 : S5</td>
<td>6</td>
<td>10</td>
<td>10</td>
<td>18</td>
<td>3.5</td>
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<td>321/43</td>
<td>LR</td>
<td>5</td>
<td>0</td>
<td>5</td>
<td>11</td>
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<td>J</td>
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<td>25UC: 0FP</td>
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<td>19UC: 0FP</td>
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<td>25</td>
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<tr>
<td>U of Cinc</td>
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<td>200</td>
<td>S193 : J7</td>
<td>6</td>
<td>5</td>
<td>13</td>
<td>18</td>
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<tr>
<td>U of Br Col</td>
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<td>174/1</td>
<td>S95: J79</td>
<td>6</td>
<td>12</td>
<td>13</td>
<td>15</td>
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<td>Str49 : 172</td>
<td>8Str : 6J</td>
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<td>W</td>
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<td>J38: S35</td>
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<td>3</td>
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<td>SBO-12, PS-6, SD-2</td>
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<tr>
<td>U of Chicago</td>
<td>Michelassi, 93</td>
<td>50/0</td>
<td>J</td>
<td>6</td>
<td>16</td>
<td>32</td>
<td>50</td>
<td>-</td>
<td>-</td>
<td>SD-19</td>
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Str-Straight Pull-Through ; LR-Lateral Ileal Reservoir  
SBO-Small Bowel Obstruction ; PS-Pelvic Sepsis  
AC-Anastomotic Complication ; SD-Sexual Dysfunction

Finally the sole purpose of a reservoir is to increase rectal capacity and thereby decrease stool frequency. We have shown recently that the neorectum created by the straight ERPT progressively dilates during the first 2 years after operation and develops a normal appearance on barium enema (suggesting normal rectal capacity) between 1 and 2 years after surgery. This is consistent with the clinical observation that initially the stool frequencies on the average are somewhat higher with the straight ERPT than with the reservoir modification; however, these differences tend to disappear 1 to 2 years after the procedure.

The data from this series of patients confirm that the straight ileoanal ERPT is an appropriate
alternative procedure (compared to the ERPT with a reservoir) for children and adults with ulcerative colitis, familial polyposis, and total colonic Hirschsprung's disease, with comparable functional results and fewer complications than are seen with the reservoir procedures.

References


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15) Fonkalsrud EW, Stelzner M, McDonald N. Experience with the endorectal ileal pullthrough

Figure 2 Radiological appearance one year after ERPT in a 14 year old female with ulcerative colitis.


