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Surgical treatment of ulcerative colitis and polyposis in childhood.

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Summary

Since 1978, 64 children with colitis or polyposis underwent colectomy with endorectal ileal pullthrough in two stages with temporary ileostomy. In 59 of the children, a lateral ileal reservoir was used as the primary procedure. Of five children with a straight pullthrough without reservoir, four required reoperation with construction of a reservoir. Eighteen of the 64 children required reoperation for complications. Only three children have required a permanent ileostomy. Of the last 45 patients, 44 are functioning well with a mean of 5.8 bowel movements per day. A short reservoir with removal of all rectal mucosa produces the fewest complications and best results.

Key words: *Endorectal ileal pullthrough, ulcerative colitis.*

Although medical therapy may provide transient remission for over 90 percent of children with severe ulcerative colitis, proctocolectomy is eventually required in over half. Children with chronic ulcerative colitis and severe recurrent symptoms frequently experience growth retardation, during adolescent years. The combination of chronic colonic inflammation, poor appetite and chronic use of corticosteroids often causes a delay in growth and maturation. As a result, the child may develop feelings of inferiority and a lack of desire to participate in many social and physical activities. Since the majority of children with severe ulcerative colitis continue to have symptoms into adulthood and eventually require proctocolectomy, serious consideration should be given to removing the colon and curing the disease during childhood. Children with severe chronic ulcerative colitis are rarely cured without opearation. Catch-up growth, and rehabilitation socially, and physically have often been dramatic after removing the diseased colon and rectum during childhood.

A major concern of children with severe colitis who have been recommended for colectomy is that they will be required to wear an ileostomy appliance for life. Although the care of an ileostomy is usually easily mastered by a child, the presence of a stoma appliance often serves as a source of embarrassment during physical and social activities. Furthermore, there is great concern among adolescent boys that proctocolectomy carries a risk of impotence. Although postoperative impotence and/or bladder dysfunction after proctocolectomy in childhood are uncommon, these major concerns have caused many children, parents, and physicians to defer the operation until they are severely debilitated by the colitis and/or steroid therapy and may manifest irreversible systemic complications.

Colectomy for ulcerative colitis in children with retention of the diseased rectum does not relieve the patient of many of the symptoms of the diasease. Although troublesome diarrhea and abdominal cramps are relieved, one frequently does not experience improved growth, often requires steroid enemas, and suffers the unpleasant aspects of a cutaneous ileostomy. Experience with the continent ileostomy (Kock pouch) has been limited in childhood because of the high incidence of complications requiring reoperation and the need for frequent stomal catheterization and instrumentation to remove solid food particles.

During the past 12 years, colectomy with muco-

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sal proctectomy and endorectal ileal pullthrough has been used with increasing frequency for the definitive surgical treatment of ulcerative colitis that is refractory to medical therapy [1,3]. The vast majority of published reports regarding the ileal pullthrough operation with or without ileal reservoir, have reviewed the clinical experience with adult patients. This report focuses on the clinical experience with the endorectal ileal pullthrough operation in children and directs attention to some aspects of management that are more specific for patients in this age group.

Much of the pioneering work in developing the endorectal pullthrough operation was performed by pediatric surgeons. This procedure completely cures the colitis, obviates the need for a permanent ileostomy, and almost always assures against the patient's developing impotence.

Operative Technique

In 1974, Ravitch and Sabiston developed the initial prototype for the endorectal ileal pullthrough procedure for patients with ulcerative colitis [4]. The pullthrough operation did not gain extensive clinical application until 1964 when Soave refined it in conjunction with a colonic pullthrough procedure for Hirschsprung's disease [5]. Removal of the entire rectal mucosa down to the dentate line was shown not to interfere significantly with anorectal sphincter function and the ability to discriminate between gas and liquid or solid contents. It also became clear that ulcerative colitis and familial colonic polyposis are diseases of the mucosa that can be cured by colectomy and mucosal proctectomy.

There was little enthusiasm for clinical use of the endorectal ileal pullthrough operation until Martin in 1977 [6] reported a good outcome in 15 of 17 children with a straight ileal pullthrough operation after colectomy and mucosal proctectomy. Although many modifications in the technique have been made during the ensuing years, it is now generally accepted as one of the most desirable options for the surgical treatment of ulcerative colitis and familial polyposis. Appealing features for this operation include the absence of a permanent stoma, the lack of repeated catheterization as needed with the Kock pouch, and the development of a near-normal pattern of defecation.

Although the initial operations used a straight endorectal ileal pullthrough with long rectal muscle cuff and retained a short segment of rectal mucosa, these features have all been changed during ensuing years. The long rectal muscle cuff causes commpression of the reservoir and angulation with partial obstruction. Retaining any rectal mucosa leads to development of recurrent colitis with frequent disruption of the ileoanal anastomosis and does not noticeably improve continence. Attempts to distend the ileal pullthrough segment with a balloon catheter to create a small reservoir have been cumbersome and generally unsuccessful.

Patients with a straight endorectal ileal pullthrough, even with a short rectal muscle cuff, in most cases will experience persistent stool frequency and urgency, and most will have some incontinence. The pullthrough segment of ileum retains peristaltic contractions, generating spike pressures down to the anal anastomosis. Most surgeons performing the ileal pullthrough procedure have, therefore, constructed an ileal reservoir above the ileoanal anastomosis to reduce peristalsis in the distal ileum and to provide an area for fecal storage. Regardless of the type of reservoir technique performed, as long as the lower 4 cm of the rectal muscle is not damaged, the anal sphincter resting pressure and the anal sphincter squeeze pressure will be close to normal within six weeks [7].

The early surgical experience with pullthrough operations indicated clearly that a completely diverting protecting ileostomy for at least three months is necessary to minimize the risk of pelvic infection. Children with chronic ulcerative colitis who receive chronic corticosteroids are often malnourished and frequently have a suppressed immune response.

Three basic reservoir types have been used clinically: the S-shaped revervoir as described by

Parks [8], the J-reservoir described by Utsonomiya [9], and the lateral isoperistaltic revervoir as initially described in our surgical unit [10]. The S-reservoir as initially must be hand sutured and requires a longer operating time than the other pouches. The blood supply to the lower ileum may be partially obstructed by bending the mesentery, particularly if it is thick. Pouch stasis is common and an irrigating catheter is often necessary for adequate emptying. After several months, the reservoir often enlarges and the spout elongates. Surgical revision is difficult because of the complex vascular pedicle.

The J-shaped reservoir has been constructed in most cases with a stapling instrument. A major advantage is the placement of the lower end of the reservoir close to the anus. A drawback with this reservoir is that it is sometimes difficult to bring the side of the ileum down to the anus without tension, particularly in heavy or tall patients. The lateral ileoanal anastomosis is also more difficult technically than an end anastomosis and has produced a somewhat higher incidence of ileoanal strictures.

In 1980 we reported our clinical experience with a lateral isoperistaltic ileal reservoir for treatment of ulcerative colitis and polyposis [10]. Although initially the procedure was performed in two stages with the first operation placing the ileum to the anus as a straight pullthrough with a diverting ileostomy, and the second, taking down the ileostomy to construct a long lateral reservoir, this technique was modified approximately five years ago.

Our current technique entails dividing the ileum within two cm to the ileocecal valve and mobilizing the mesentery up to the origin of the superior mesenteric arter. It is rare for active ulcerative colitis in children to extend into the terminal ileum. The colon and omentum are removed down to the level of the pelvic rim, and the specimen is examined by the pathologist to be sure that the child does not have Crohn's disease or malignancy. The rectum is extensively irrigated with antibiotic solution. The peritoneal reflection is incised circumferentially and the rectum is mobilized with cautery dissection down to within four cm from the dentate line, where the full thickness of rectum is divided. The anus is dilated and a circumferential incision is made through the mucosa at the level of the dentate line with electrocauter. The mucosa is elevated from the underlying muscularis using scissor dissection, facilitated by injecting dilute epinephrine solution between the two layers. Care is taken to cause infection and/or bleeding. Thorough hemostasis is achieved with electrocauter, and the pelvis is irrigated with antibiotic solution.

The ileum is transected approximately 15 cm from the and, with care taken to preserve the vascular supply to the distal segment. The proximal ileum is advanced into a position adjacent to the distal segment and a lateral isoperistaltic anastomosis is performed between the antimesenteric sides of each using the GIA stapling device. A second layer of continuous absorbable suture is placed circumferentially around the entire anastomosis. A spout of approximately 1.5 cm extends between the distal end of the reservoir and the open end of the ileum. The ileal revervoir is brought through the pelvis and rectal muscle canal such that the end of the ileum extends to the anus (Figure 1). The full thickness of the open end of the ileal spout is stitched to the anoderm and underlying muscularis circumferentially with interrupted absorbable sutures. The ileum is transected approximately 10 cm proximal to the end of the reservoir with the GIA stapling device. The proximal end of the divided ileum is fashioned into an end-ileostomy stoma. The pelvis is drained through a Silastic catheter extending through the abdominal wall for approximately four days.

During the postoperative period, oral feedings are usually started within 48 hours and the child is given instruction regarding ileostomy care by the enterostomal therapist. Intravenous steroids are tapered down rapidly after operation and oral Prednisone can usually be discontinued within there weeks. Most children are discharged from the haspital by the seventh postoperative day. During the ensuing three months the patient is examined on at least two occasions with particular empha-

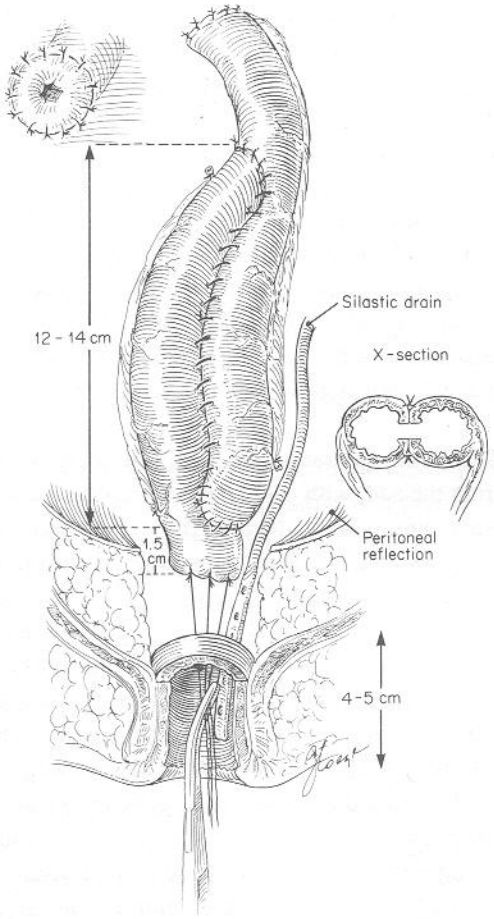

Figure 1. Lateral ileal reservoir of 12-14 cm length with 1.5-2 cm ileal spout. Rectal muscle cuff is 4-5 cm. Completely diverting ileostomy is placed approximately 8-10 cm above the reservoir.

sis given to identifying an ileoanal stricture at an early stage in order to permit dilatation as soon as possible. A Gastrografin enema is performed within three months to assure that the ileal reservoir has healed securely and that there are no leaks or sinus tracts. Most children resume full physical activities within three weeks.

Approximately four months following the first operation, the child is rehospitalized and the ileostomy is closed with an end-to-end anastomosis performed through the midportion of the previous laparotomy wound. A large Foley catheter is left in the rectum to drain ileal contents for four days. The child is started on oral feedings on the third day and gradually advanced to a modified regular diet, omitting chocolate, salad dressings, spicy foods, and given certain citrus fruits. Intravenous steroids and antibiotics are given for four days following which oral Prednisone and Metronidazole are started and gradually tapered downward. Small amounts of Metamucil are given to increase fecal bulk for the first few weeks. Medications to reduce peristaltic activity are rarely necessary. The patient is usually discharged from the hospital within seven days postoperation.

Results

During the past 12 years, 64 children 18 years of age or younger, 58 with ulcerative coilitis, and 6 with polyposis, have undergone colectomy, mucosal proctectomy, and endorectal ileal pullthrough at the UCLA Medical Center. Operation was performed on six patients under 10 years, 24 patients between 10 and 14 years, and 34 patients between 14 and 19 years of age. During the same period an additional 148 patients 19 years of age or older with ulcerative colitis or polyposis underwent the endorectal ileal pullthrough procedure.

In 59 of the children, a lateral ileal reservoir was used as the primary procedure. Twenty-eight of the children had the ileal reservoir constructed at the second operation when the ileostomy was closed. Thirty-one children, all during the past four years, had the reservoir constructed at the first operation, as decribed earlier. Five children had a straight endorectal pullthrough without reservoir. Each of the children had a protecting ileostomy for a mean period of 3.8 months after the pullthrough procedure. Only nine patients required blood transfusions intraoperatively, all of whom were anemic before surgery. Each of the 58 children who underwent the pullthrough operation for colitis experienced marked clinical improvement following colectomy.

Fourteen of the 59 patients with a lateral reservoir and four of the five patients with a straight endorectal pullthrough procedure required subsequent reoperation (29.7 percent). The most common complication following the pullthrough operation was chronic reservoir inflammation

which appeared to be related to difficulty in emptying the reservoir, either due to outlet obstruction from stricture, long rectal muscle cuff or long ileal spout. Reservoirs exceeding 20 cm in length appeared particularly susceptible to distention and elongation, causing diarrhea, stool frequency and urgency, increased gas, bacterial overgrowth, and abdominal discomfort. In seven patients elongated reservoirs were shortened during the ensuing five years to a length of 15 cm or less by resecting the upper end of the reservoir and anastomosing the end of the ileum to the remaining reservoir. Three patients had simultaneous shor-tening of the elongated rectal muscle cuff to alleviate the reservoir outlet obstruction. Mild to moderate stenosis of the ileoanal anastomosis occurred in six patients which was corrected by dilatations on an outpatient basis. Reservoir inflammation rarely occurred in patients who had the ileal reservoir constructed at the same time as the pullthrough procedure and in whom the reservoir was less than 15 cm in length and the ileal spout distal to the reservoir was less than 1.5 cm. Nine children had intestinal obstruction secondary to adhesions, to internal hernias, or to intussusception. Two patients developed a small sinus tract extending up from the ileoanal anastomosis between the rectal muscle cuff and the ileal pullthrough segment which healed after marsupialization into the rectal lumen under general anesthesia. A rectovulvar fistula requiring operative repair occurred in one patient.

In summarizing the complications, three of the 64 children were returned to a permanent ileostomy (4.7 percent) with removal of the reservoir; one of these patients eventually proved to have Crohn's disease instead of ulcerative colitis. Six of the 64 children were returned to a temporary ileostomy because of various complications and/or to encourage catch-up growth during the adolescent years; four of the ileostomies heve subsequently been closed without sequelae. Four of the five children with straight pullthrough procedures exqerienced severe stool fequency and urgency and required reoperation with conversion to a lateral revervoir. Thus 58 of the 63 children currently have a functioning lateral reservoir and one a straight pullthrough. None of the children died following either the first or second stage operations.

Fifty-nine of the 64 children (92 percent) are currently progressing well with the pullthrough procedure. The majority have between three to six continent bowel movements per 24 hours (mean 5.2/24 hours). Less than 15 percent experienced daytime soiling within three months although 35 percent had an occasional soiling episode during nocturnal hours. By six months less than 18 percent experienced nocturnal soiling. By 24 months, seepage or incontinence is very rare. Thirty-four of the patients are participating in active or vigorous athletic activities. None of the male patients has experienced impotence or evidence of retrograde ejaculation. One of the patients, age 17 at operation, delivered a healthy full-term male baby two years postoperation, vaginally. Detailed review of the last 35 children shows that all but two are currently functioning well, indicating the decreased number of complications as our clinical experience has increased.

Comment

As clinical experience with the endorectal ileal pullthrough operation has increased, it has been well documented that anorectal sensory and continence mechanisms can be preserved and function well if anal sphincter and low rectal muscle are not disturbed, even if the entire rectal mucosa is removed down to the dentate line. The legth of rectal muscle retained above the ileoanal anastomosis need not be longer than four cm, shorter muscle cuffs may increase the risk of retrograde ejaculation and impotence. Longer muscle cuffs cause reservoir compression, are prone the contracture, and may cause pouch angulation, and appear to provide no useful function.

Although the report by Morgan and associates [11] indicates that the straight pullthrough produces continence with similar stool frequency as when a reservoir is used, our clinical experience and that of most other authors has been quite different. Four of the five children with no reservoir had such severe frequency, urgency, and eventu-

ally and that an ileostomy be constructed. Each of the four patients who had an unacceptable result with a straight pullthrough underwent successful conversion to a lateral reservoir with an average of seven stools per 24 hours within four weeks. Although it may be technically difficult to construct a reservoir and perform a pullthrough at the first operation in certain obese patients, or those with a thick and short ileal mesentery, a straight pullthrough can almost always be performed. At the second operation in four months, invariably the mesentery will be elongated and the straight pullthrough can be mobilized from the pelvis, a reservoir constructed, and a new pullthrough performed. Only rare patients in our experience will progress well without a reservoir.

Although a variety of reservoir are currently in clinical use, with somewhat comparable early results with regard to stool frequency and continence, it has become apparent that pouchitis is more common in larger reservoir that empty only partially with each defecation. In contrast to colonic mucosa, the ileal reservoir mucosa becomes inflamed when stasis and bacterial overgrowth occur, leading to diarrhea, urgency, frequency, and occasional incontinence. Although many patients will be relieved of symptoms by adherence to a constipating diet and the use of antidiarrheal medications, the symptoms often recur if only partial emptying occurs. Stenosis at the ileoanal anastomosis or lower end of the reservoir may appear to be a mild annoying problem, but can cause reservoir distention, stasis, and pouchitis if not corrected early.

Gradual enlargement of ileal reservoirs during the first year is anticipated regardless of the configuration. Since the daily fecal output for most patients with a pullthrough operation varies between 600 and 1.000 ml, reservoirs with a capacity of more than 350 ml would appear to be unnecessary and may be counterproductive.

Reservoir enlargement of sufficient magnitude to produce chronic pouchitis may not become evident for more than 18 months after operation; however, when symptoms persist despite therapy, operative shortening of the reservoir combined with relief of outlet stenosis or shortening of the ileal spout may relieve the pouchitis permanently as with seven patients in this report. The lateral reservoir lends itself particularly well to the shortening procedure with no major complications in any of the children.

An important observation from the present clinical review is that almost all complications following the endorectal ileal pullthrough procedure were surgically correctable when recognized and treated early. Temporary ileostomy may be necessary in certain patients particularly if growth and development during the adolescent years is not progressing at an optimal rate. After the reservoir obstruction or reconstruction has been completed and the child has returned to a steady progress upward on the growth curve, the ileostomy in almost all cases may be safely closed. Most complications occurred early in our clinical experience; none of the last 40 patients has the reservoir removed and had a permanent ileostomy constructed. Since the endorectal pullthrough procedure is somewhat complex and is usually performed on sick children with severe colitis who have received high dose corticosteroid therapy, a completely diverting end ileostomy for there months is essential to reduce pelvic complications.

During the past four years, operating time and hospital stay have been shortened significantly. Complications requiring reoperation have been reduced to seven percent. Reduced complications have been observed with the use of a short rectal muscle cuff, a short ileal reservoir, a short ileal spout, and removal of all rectal mucosa down to the dentate line. Long-term follow-up is necessary to recognize some of the more insidious complications of the endorectal ileal pullthrough with reservoir in children.

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