

The Importance of Early Surgery in Children with Ulcerative Colitis

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Abstract

Background: Ulcerative colitis begins in early childhood in 4% of cases. Medical therapy is non-specific, and as many as 70% of children will ultimately require surgery. The dynamic growth, physical and psychological changes that characterize childhood are severely compromised by the complications of ulcerative colitis and its therapy.

Objective: To review the outcome of children undergoing early surgery for ulcerative colitis at a tertiary medical center in Israel.

Methods: A retrospective review was conducted of all children operated on following failure of medical therapy for ulcerative colitis during a 5 year period.

Results: Eleven children underwent a J-pouch procedure with ileo-anal anastomosis in one to three stages. Postoperative complications included recurrent pouchitis in 5 patients, intestinal obstruction in 3, fistula with incontinence in one, stricture in one, and wound infection in 4. Follow-up revealed that most of the patients have three to four soft bowel movements daily. All currently enjoy normal physical activities and a rich social life.

Conclusions: The quality of life in children with ulcerative colitis was markedly improved following J-pouch surgery. This procedure was not associated with major complications. We recommend early surgery as an alternative to aggressive medical therapy in children with this disease.

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Ulcerative colitis is primarily a disease of the rectal and colonic mucosa. The disease begins in 4% of patients before the age of 10, and in 18% between the ages of 10 and 20. The disease is more severe in children than in adults, and is often associated with more overt symptoms.

The onset of disease is acute in 15% of children and usually presents as fulminant bloody diarrhea accompanied by severe abdominal pain. Fever, and occasionally sepsis, may be the initial symptoms. Extra-intestinal manifestations such as growth failure, arthralgia, skin

lesions, failure of sexual maturation, anemia, nephrolithiasis, uveitis, erythema nodosum and pyoderma gangrenosum may precede gastrointestinal disease by several years.

Medical therapy is non-specific and provides only symptomatic relief. Although such modalities are preferred as the initial treatment of choice, 5% of patients will undergo urgent operation for toxic megacolon or severe bleeding. Regardless of medical therapy, as many as 70% will ultimately require surgical intervention [1].

Childhood is characterized by a highly dynamic state of growth and physical change, associated with a constantly evolving psychological status. Normal puberty is a key milestone in the physical and psychological development of the child. Since medical treatment may interfere with normal growth and complicate psychological maturation, early surgical intervention should be considered. Non-operative treatment is based on measures to provide symptomatic relief, and may include sulfasalazine for mild symptoms and corticosteroids for acute exacerbations. Although corticosteroids may alleviate symptoms, they are associated with growth failure and osteoporosis in children who may already be small for their age. Osteoporosis and its associated bone pain may prevent the child from participating in sports, while cushingoid features will impact on body image at this crucial age. Steroid enemata will further contribute to psychological trauma.

When steroids are unsuccessful, immunosuppressive agents such as azathioprine, 6-mercaptopurine, cyclosporine or methotrexate may be added. These drugs may produce pancreatitis or hirsutism, the latter particularly disturbing for young girls. Multiple blood transfusions carry additional hazards, while repeat hospitalizations are incompatible with normal life at this age. It is noteworthy that 15% of patients have family members with inflammatory bowel disease, each requiring intensive medical and psychosocial care [2].

Complications following surgery increase with the duration of disease and prior medical therapy [3,4]. Carcinoma of the colon or rectum develops in 3% of patients during the first 10 years of disease, and increases by approximately 10-15% during each subsequent decade. Cancer is more common in patients with

pancolitis and in those whose symptoms begin during childhood [5–7].

Methods

A retrospective 5 year review of all children with ulcerative colitis referred and treated in the Pediatric Surgery Service of a tertiary hospital was conducted. Cases in which surgery was performed following failure of intensive medical therapy were analyzed for the status of patients before and following corrective surgery.

Results

Eleven children with ulcerative colitis were referred following unsuccessful medical therapy [Table 1]. The patient group consisted of 7 girls and 4 boys with a mean duration of illness of 25.2 months (range 6–52 months). The youngest was 7 years old at diagnosis. All had received long-acting sulfa compounds, corticosteroids, 6-mercaptopurine, azathioprine and cyclosporine. Three of the cases had additional family members with inflammatory bowel disease.

All patients were suffering from abdominal pain and bloody diarrhea and had required multiple blood transfusions. Their physical appearances were similar, with 'moon facies', hirsutism and abdominal striae. Several complained of back pain, myalgia and bone pain. One patient had experienced a compression fracture of T5 and

had evidence of severe osteoporosis. Two boys had been treated for drug-induced pancreatitis, one requiring cyst gastrostomy for a pseudocyst. Two girls presented with pyoderma gangrenosum that resolved following surgery.

Complications included recurrent pouchitis (5 patients), intestinal obstruction (3 patients), fistula and incontinence (one), stricture (one), and wound infection (4 patients). Most of the patients in this study have been followed for at least 4 years following surgery, and report three to four soft bowel movements daily.

Discussion

Ulcerative colitis can be cured by surgical resection of the diseased colon and rectum. Surgery may be either elective or emergent. Available procedures vary from proctocolectomy with permanent ileostomy, to various forms of ileal pouch with ileo-anal anastomosis.

Forty years ago the procedure of choice was proctocolectomy with ileostomy. Unfortunately, this required a permanent appliance, which represented punishment to the pediatric patient and caused embarrassment during physical and social activities. Koch [8] introduced the continent ileal reservoir in 1969. In 1964, Soave [9] described an endorectal pull-through technique for Hirschsprung's disease that was subsequently used for the definitive treatment of ulcerative colitis. This procedure is still

Table 1. Surgery for ulcerative colitis in children

Case	Age/Gender	Duration of disease (mo)	Complications of medical therapy	Staged surgery	Postoperative complications	BM per day / continence
1	16/F	24	Back pain, arthralgia, hirsutism	II	Mild ileo-anal stricture dilatation	3 / yes
2	16/M	52	Pancreatitis	I	Fistula *	Many / no
3	16/M	24	Pancreatitis, pseudocyst, cyst, gastrostomy	II	Pouchitis	4 / yes
4	15/M	24	Moon face, hirsutism	II	Pouchitis, SBO	5 / yes
5	14/M	13	Myalgia, moon face, arthralgia	II	Pouchitis, SBO, wound infection	4 / yes
6	13.5/F	36	Moon face, hirsutism, compound fracture T5, osteoporosis	III	Pouchitis	4 / yes
7	16/F	12	Hirsutism, pyoderma gangrenosum	II	Wound infection	3 / yes
8	15/F	6	Myalgia, moon face, hirsutism, pyoderma gangrenosum	II	Pouchitis	4 / yes
9	14/F	36	Headaches, moon face, arthralgia	I	Wound infection	4 / yes
10	9/F	36	Short stature, hirsutism, myalgia, moon face	II	None	4 / yes
11	14/F	12	Moon face, short stature, hirsutism	**	Stricture, pelvic abscess, fistula, wound infection	**

* Later found to have Crohn's disease

** Permanent ileostomy

SBO = small bowel obstruction

used successfully by some authorities in the field, often in conjunction with multiple vertical linear myotomies to the distal 10–15 cm, proximal to the ileal-anal anastomosis [10]. Since frequent bowel movements often follow this operation, surgeons have continued to search for alternative procedures.

In 1980 the ileal reservoir pouch was introduced for familial polyposis, and later for ulcerative colitis. Pouches may be lateral isoperistaltic, S-, W- or J-shaped reservoirs. Since S- and W- pouches must be hand sutured, longer operating time is required. Lateral isoperistalsis and J-pouch techniques employ stapling techniques.

We favor the J-pouch procedure using double staples. First described by Utsunomiya et al. [11], this technique is the simplest and most commonly used, and is associated with the lowest incidence of long-term complications in children and adults. If the lower 4 cm of rectal muscle is not damaged, the resting and squeezing pressure of the anal sphincter will approach normal values within 3 months regardless of the type of reservoir used [2].

While 30–60% will develop complications following ileo-anal pouch surgery, the mortality rate is less than 1% [12,13]. Pouchitis (reservoir inflammation) is the most common complication during the first 2 years following surgery and occurs in 10–50% of patients. This process may be related to outlet obstruction due to narrowing at the pouch-anus anastomosis site. Stelzner et al. [14] attribute this condition to the length of the reservoir and short rectal muscle cuff. The condition improves in most patients after 6 months on a regimen of metronidazole or ciprofloxacin. Other complications of pouches include anastomotic leak with pelvic abscess, strictures and late fistulae. The incidence of postoperative pelvic infection is 20%, while those of leaking and late fistula are 4% and 10%, respectively. Rectovaginal fistulae occur in 1–6%, intestinal obstruction develops in 10–25%, severe incontinence in 2–6%, and wound infection in 5–15% [2].

Pouch removal and construction of a permanent ileostomy will be required in 6–15% of patients [6]. Among these, 30% will ultimately be found to have Crohn's disease. During the first 3 months after surgery, children experience five to nine bowel movements daily, and three to four by the end of 6 months. Nocturnal staining occurs in 15% during the first 3 months, decreasing to 5% by 6 months. Many patients participate in vigorous

physical activities, including marathon races, swimming and football [2]. Following surgery, functional results and quality of life are considered to be good to excellent [12].

Although our patients experienced many of the above complications, all are presently engaged in normal sports and activities, enjoy a rich social life, and participate in the perioperative psychological support of other children requiring surgery in our service. In view of these positive results and an impressive body of favorable literature, we highly recommend early surgery for children with ulcerative colitis as an alternative to aggressive medical therapy.

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