Surgical Treatment of Ulcerative Colitis in Childhood*

TH. EHRENPREIS

From the Children's Clinic, Karolinska Sjukhuset, Stockholm, Sweden

As long as the aetiology of ulcerative colitis remains obscure, treatment of the disease must be confined to symptomatic relief measures. A wide variety of both medical and surgical methods of treatment have been tried over the years. During the past decade, our interest and expectations have been focused mainly on two principles of treatment, one medical and one surgical. The introduction of steroids in the treatment of ulcerative colitis brought about a wave of optimism among the medical profession. During the past few years, however, this optimism has proved unwarranted with regard to long-term results. The introduction of radical surgical procedures met with less enthusiasm, but has proved to offer considerably more than what is sacrificed. The value of radical surgery has been firmly established and the proportion of cases treated surgically is steadily increasing.

The pathology and clinical manifestations of ulcerative colitis in children are essentially the same as in adults, with the whole range of well-known disease patterns, from the acute fulminating disaster, to the chronic remittent manifestations of the disease. The same extracolonic manifestations are found and the same complications from the bowel lesions occur: haemorrhage with anaemia, protein and fluid losses with electrolyte imbalance, necrotizing ulcerations with perforations and peritonitis, fibrosis with stricture formation, and malignant degeneration.

In addition to these features many children show physical and emotional retardation from the detrimental effects of the disease upon the growing organism.

The response to medical treatment may be temporarily promising, but long-term results are unsatisfactory with regard to mortality and morbidity rates. In a series of 137 cases from the medical department of our clinic, Lagercrantz (1955) reported a mortality rate of 14%, the most common cause of death being cancer of the colon; 54% of the cases were more or less disabled for shorter or longer periods of time, the remaining 32% stated that they

had been free from symptoms for two years or more, but upon examination many of these patients still showed clinical or radiological evidence of the disease. The cancer incidence was 10% after 10 years of disease, and was higher after that time.

Similar results are found in other paediatric series. The seriousness of the situation is stressed by the fact that a remarkable increase in hospital admissions for ulcerative colitis in children has occurred in the Stockholm area (Fig. 1). These facts prompted us to adopt a more active policy, and in 1953 a team of paediatricians, paediatric surgeons, radiologists, and psychiatrists was established in order to mount a joint attack against ulcerative colitis. The following report is based upon the experience gained during the 12 years that have passed.

FIG. 1.—Number of children admitted for ulcerative colitis to all children's hospitals in Stockholm 1919-1953.

Present Approach

Every patient with ulcerative colitis admitted to the Children's Clinic has been subjected to complete clinical, psychological, laboratory, and radiological examination. Each patient has then been discussed by the full team in order to decide the mode of treatment. Initially, all patients were treated medically. Acutely ill patients were reviewed once or several

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times daily until remission of all threatening signs had occurred, or until emergency surgery had been performed. In chronic or relapsing forms of the disease, the patient's situation was re-evaluated periodically.

Indications for Surgical Treatment

Surgical treatment has been considered on the following indications, which from the beginning have formed the basis for our policy; (a) *emergency surgery* carried out for acute, life-threatening disease and/or complications (including profuse haemorrhage, perforation, obstruction, and malignancy); (b) *elective surgery* for chronic disablement.

The application of these basic principles has resulted in surgical treatment of 45 patients, representing about 20% of the total number of our series. This is in accordance with recent figures from other paediatric centres.

Sex distribution was about even in our series. The age at onset of the disease and at operation is shown in Table I.

TABLE I
Ages at Onset and at Operation

Age-group (yr.)	At Onset No. of Cases	At Operation No. of Cases
3-5	7	
6-8	7	
9-12	25	8
13-16	6	30
17-19		7
Total	45	45

Emergency operations were performed in 14 patients, representing about one-third of those treated surgically and 7% of the total of patients admitted. All of them followed a grave and rapidly progressive course with high fever, marked anaemia, and gross weight loss. In addition 5 developed signs of toxicity and peritoneal irritation. Radiological examination in these patients disclosed dilatation of the colon, fluid in the abdominal cavity, and/or deep ulcerations penetrating into the bowel wall. 3 patients bled profusely from the bowel, requiring almost continuous transfusions for replacement of blood losses which persisted until the diseased bowel had been removed. There were no bowel perforations, no obstructive strictures, and no malignancies. This group of patients had a history of ulcerative colitis dating back from 3 months to 8 years, the emergency situation being the first manifestation of the disease or developing as a severe relapse after insidious earlier symptoms.

Elective surgery was performed in 31 patients (about two-thirds of the surgical cases and 14% of the total patients admitted). Chronic disability was considered to be present when 1 year or more of total and continuous disability had elapsed after initiation of adequate medical treatment, or after 5 years or more of intermittent disease, causing more or less prolonged periods of disablement despite adequate medical therapy. Obviously, the indications for elective surgery are less clear cut than those for emergency operations. Besides the actual disablement caused by the disease, two additional considerations should be kept in mind for the choice of treatment in juvenile patients. Signs of physical and emotional retardation were common in our series: radical surgery stops retardation and promotes normal growth and development. The risks of malignancies are higher when the disease has started in childhood. Cancer may develop after many years of clinically inactive disease. The difficulties in diagnosing carcinoma in patients with chronic ulcerative colitis are reflected in the poor results of surgical treatment in these cases. A history of 10 years or more of ulcerative colitis is in itself a strong indication for surgical treatment, irrespective of the degree of clinical manifestations.

During the 12-year period covered by this study, there has been no official change in our indications for surgical treatment; however, there has been a trend towards earlier operations in the elective group. This was initiated mainly by the medical member of the team on the basis of his continuous observations on both medically and surgically treated patients in a follow-up clinic.

Pre-operative Treatment

Replacement of blood, albumin, fluid, and electrolyte losses are the main aims of the preparation of patients for operation. In emergencies, full replacement may not be attained before surgery has to be performed. The most obvious losses should be dealt with first. It is often astonishing to witness the improvement brought about by initiating repletion of these patients. A fulminating disease picture may change within 24 or 48 hours into one of modest symptoms, not requiring further emergency measures.

Pre-operative bowel cleansing by mechanical measures should be omitted in these patients. Antibiotics should be given in accordance with the routine of each clinic. We have used oxytetracycline for 48 hours before operation and 3 to 4 days after, in dosages of 20 mg./kg. If steroids have been administered at any time and in any way before surgical treatment is instituted, the patient should

receive adequate steroid support in connexion with the surgical procedure.

Psychological preparation of the patients as well as of their parents is of utmost importance, especially if a permanent ileostomy is planned. The reasons for surgical treatment, the choice of operation as imposed by the extent of the bowel lesion (see below), and its implications for the future status and care of the patient should be explained in simple and understandable terms. The best way to convince patients as well as parents of the benefits of colectomy, even at the price of a permanent ileostomy, is to introduce them to one of our earlier ileostomy patients and his parents for free questioning and discussion.

Choice of Operation

Today, temporary ileostomy alone has been abandoned by most surgeons in the treatment of ulcerative colitis. The reasons for this are unequivocal. Healing of the bowel lesion does not occur even after prolonged diversion by an ileostomy. The risks of malignant degeneration remain unchanged as long as diseased bowel remains. Paradoxical as it may seem, the operative mortality rate from ileostomy alone is higher than that of radical procedures: the diseased bowel remains as a source of toxicity and of continued losses of blood, albumin, etc. When severe fluid losses from the ileostomy are added to these pre-operative disturbances, a disastrous situation may develop.

All evidence points toward the necessity of more radical procedures. It is obvious that radical excision of the entire diseased bowel with restoration of bowel continuity would be the ideal method of surgical treatment. Unfortunately, the extent of the bowel lesion does not permit the fulfilment of both these requirements in the majority of cases. Segmental colitis is rare; the rectum is unanimously stated to be involved in 90 to 95% of all cases. These facts force the surgeon to renounce one of his two ideal requirements—either total excision or bowel continuity. The choice is not an easy one.

The majority of the leading surgeons prefer radical excision (pan-coloproctectomy) combined with a permanent ileostomy. Methods that compromise with a radical operation are condemned as ineffective and dangerous. Residual symptoms from proctitis forced these authorities to perform secondary removal of the rectum with ileostomy in a large proportion of their cases of ileo-rectal anastomosis. The dangers of malignant degeneration within the remaining rectum are stressed.

Relatively few surgeons have insisted on restoration of bowel continuity, either by ileo-anal or ileo-rectal anastomosis. Ileo-anal anastomosis has been

used in a small series of patients with ulcerative colitis. The results have generally been disappointing, mainly because of incontinence of the anal ileostomy. Today, this procedure seems to have been largely abandoned.

Aylett (1954, 1963) has been the most enthusiastic exponent of total colectomy and ileo-rectal anastomosis for ulcerative colitis. This procedure has been used by him for all patients except those presenting ano-rectal strictures, abscesses or fistulas. According to a recent report of 213 cases over a 10-year period, there were 11 operative deaths and 7 patients died later, from 1 to 8 years after operation. Secondary excision of the rectum with ileostomy had to be performed in 13 patients (6.4% of survivors). Cancer of the rectum developed in 3 patients. Excellent results are claimed in 190 patients in this series. Regression of rectal disease after ileo-rectal anastomosis is reported as common. This experience has recently been corroborated in an interesting report on a long-term follow-up study of Wangenstein and Toon's 1948 series with the addition of patients operated upon in more recent years. Altogether, 45 patients with ileoproctostomy were reviewed, and good results are reported in 25. The degree of rectal involvement at the time of operation played a significant role in the outcome. 2 patients developed carcinoma in the retained rectum.

When dealing with children and adolescents, the desire to preserve bowel continuity is naturally greater than in adult patients. Most parents and children have a strong prejudice against permanent ileostomy. This has not failed to influence our own approach to the problem. Our selection of surgical procedure has been a compromise (Table II) between radical excision and preservation of bowel continuity, based upon the degree of rectal involvement as evidenced by proctoscopy, radiological examination, and operative findings. This approach is in close agreement with the views recently expressed by Griffen, Lillehei, and Wangenstein (1963).

Total colectomy and ileo-rectal anastomosis has been performed in all patients with slight or moderate signs of rectal involvement. 19 patients out of a total of 45 were treated according to this method. All showed slight or moderate signs of proctitis at proctoscopy. In 10, however, no signs of rectal involvement or periproctitis were observed radiologically, and the remaining 9 patients showed slight changes only. In 2 of these patients secondary excision of the rectum and ileostomy had to be performed. They were probably misjudged initially, and should have been treated primarily by pan-coloproctectomy and ileostomy. They illustrate

TABLE II
Indications and Types of Operation Performed

	Pan-coloproctectomy + Ileostomy	Colectomy + Ileo-rectal Anastomosis	Colectomy + Ileostomy	Total
Emergency operations	3 (1 death)	8	3	14 (1 death)
Elective operations	14 (1 death)	11	6	31 (1 death)
Total	17 (2 deaths)	19	9	45 (2 deaths)
Secondary proctectomies		2	4	6

the difficulties sometimes encountered in the assessment of rectal involvement.

A second group of patients showed rectal involvement of a more advanced degree, which was judged to make these patients unsuited for primary ileo-rectal anastomosis. On the other hand, the degree of proctitis in these patients was still moderate. In order to maintain a possibility of later restoration of bowel continuity, the rectum was saved and closed in these patients and a total colectomy and ileostomy performed. This procedure was used in 9 patients. It was hoped that colectomy and faecal diversion would promote healing of the rectal lesion. These expectations have, however, proved to be unwarranted. In none has healing or regression of the rectal changes occurred during follow-up periods of from

3 to 8 years. In view of the risk of malignant degeneration, it seems inadvisable, in the long run, to leave the rectum behind. Secondary proctectomy has already been performed in 4 patients and will probably be done also in the remaining 5.

Pan-coloproctectomy and a permanent ileostomy have been performed in all patients with severe proctitis and proctitis. Lateral views of the rectum after barium enema have been especially helpful in assessing the degree of rectal involvement (Fig. 2). 17 patients out of a total of 45 were treated by pan-coloproctectomy and ileostomy. This is a radical procedure, removing the entire diseased part of the bowel. No secondary operations for the cure of the disease were required in this group of patients.

As a consequence of our negative experiences with

FIG. 2a.—True lateral view of normal rectum (note distance to sacrum). FIG. 2b.—True lateral view of rectum in a case of ulcerative colitis: advanced proctitis with stricture of the rectosigmoid and increased distance between rectum and sacrum (periproctitis).

TABLE III
Surgical Procedures in Paediatric Series of Ulcerative Colitis

Author	No. of Cases	Pan-colo- proctectomy + Ileostomy	Colectomy + Ileostomy	Colectomy + Anastomosis	Ileostomy + Other Operations	Deaths
King, Lindner, and Pollard (1959) ..	12	4	6	2	—	1
Platt, Schlesinger, and Benson (1960) ..	12	1	1	9	1	—
Santulli (1960)	29	9	9	7	4	4
Present series	45	17	9	19	—	2

the colectomy and ileostomy group, we have abandoned this operation. The choice should be made primarily between colectomy with ileo-rectal anastomosis, and pan-coloproctectomy with permanent ileostomy. When in doubt with regard to the degree of rectal involvement, it seems reasonable to give these patients a fair trial of living with an ileo-rectal anastomosis. This status permits conversion to an ileostomy and excision of the rectum at any time. Primary pan-coloproctectomy, on the other hand, is a definitive and irreversible procedure.

Whether radical surgical procedures should be performed in one, two, or even three stages is a matter of surgical judgement and temperament. We have preferred to complete the whole procedure in one stage.

A synopsis of operative procedures recorded in paediatric series of ulcerative colitis has been compiled (Table III). The terminology used by different authors varies, and the extent of the surgical procedure of 'colectomy' is not always clearly defined. The cases listed under the heading 'colectomy + ileostomy' may include some in which a pan-coloproctectomy was performed.

Post-operative Treatment

Blood lost during operation, including estimated loss of intracolonic blood, should be replaced. A nasogastric tube has been used routinely for decompression until bowel movements start, as a rule for 2 to 4 days. Fluid losses from the gastric tube, and later from the ileostomy or rectum, were

measured and replaced to about 80% by electrolyte solutions. Maintenance of normal blood volume, Hb, and serum protein levels by administration of whole blood, plasma, or serum albumin is essential. As a rule, fluid losses are considerable during the first week after operation. Treatment with antibiotics and steroids was continued as stated above.

Results

The post-operative course has been surprisingly mild in the majority of our patients, including those in whom an emergency operation was performed. Two deaths occurred in the present series.

As seen in Table III, operative mortality rates in other paediatric series were also low. So far, no late deaths have occurred in our series or been reported in the other paediatric series among patients treated by radical surgical procedures. With due reservation for relatively short follow-up periods, this compares favourably with mortality rates of medical treatment.

A number of complications are apt to occur in any series of cases of this kind. The present one is no exception to this rule. A total of 49 complications (Table IV) occurred in 27 of our patients: 18 made an uneventful recovery; 3 children were desperately ill and exhausted as a result of emergency operations, and required tracheostomy and respiratory treatment. The frequent occurrence of small bowel obstruction in this series as well as in other reports is not surprising in view of the pericolicitis, lymphadenitis, and peritoneal irritation regularly found at

TABLE IV
Post-operative Complications in Present Series

Complication	No.	Treatment	Death
Gangrene of terminal ileum, peritonitis	1	Laparotomy, resection	1
Thrombocytopenia, septicaemia	1	Corticosteroids	1
Respiratory failure	3	Tracheotomy, respirator	—
Small bowel obstruction (in 14 patients)	21	Lysis	—
Ileostomy dysfunction	5	Revision of stoma	—
Wound rupture or infection	13	Resuture for drainage	—
Cicatricial hernia	1	Hernia repair	—
Intra-abdominal abscess, profuse intestinal bleeding	1	Spontaneous perforation into bowel	—
Renal concretion	2	Pyelolithotomy	—
Duodenal ulcer	1	Conservative	—

operation, and the large denuded areas following total colectomy. Ileostomy dysfunction was due to stenosis, retraction, prolapse, or kinking.

Apart from the complications listed, convalescence was rapid and uneventful, and the patients were discharged in good condition.

Follow-up

The 43 surviving patients have been followed for from 8 months to 12 years after operation.

The general condition of all patients has been excellent. Weight gain after operation was rapid and as a rule corresponded to pre-operative weight loss. The majority of patients gained between 10 and 20 kg. during the first 6 months after operation. Signs of physical and emotional retardation ceased, and development subsequently became adjusted to normal proportions. Anaemia disappeared. The stools were of varying consistency, mostly moderately loose or semisolid, depending on dietary variations. Patients treated by colectomy and ileo-rectal anastomosis have from 2 to 6 bowel movements a day, and only rarely in the night. None of them has been incontinent. Total regression of slight pre-operative proctitis occurred in 2 instances, the remaining patients in this group showing evidence of persistent slight or moderate proctitis. Ileostomy patients change their bag two to four times daily and only rarely during the night. Adjustment to ileostomy life has been surprisingly good in view of what might be anticipated in a child or adolescent. No obvious differences in results were found between the ileostomy group and the group with ileo-rectal anastomosis, except for the natural advantages of restored bowel continuity. As mentioned before, proctitis showed no signs of regression in those ileostomy patients in whom the rectum had been left behind at the primary operation. Secondary proctectomy has already been performed in 4 patients and will probably be performed also in the remaining 5 patients. All of these children and adolescents are reasonably healthy and happy, leading a normal life with regard to school work or professional activities, sports, and other diversions.

Obviously, the evaluation of results is somewhat arbitrary and strictly temporary. Prolonged follow-

up is required for final assessment of the results of any method of treatment in ulcerative colitis. In particular, we would like to stress the importance of regular and life-long surveillance of patients with remaining rectum, whether it has been anastomosed to the ileum or not. With these reservations in mind, however, we feel justified in stating that the results of radical surgery in the treatment of ulcerative colitis in children are extremely gratifying and promising.

Summary and Conclusions

Indications for surgical treatment of ulcerative colitis should be more liberal in children than in adults. In patients with slight or moderate evidence of rectal involvement, total colectomy and ileo-rectal anastomosis can be performed safely, provided that regular surveillance is continued as long as the rectum remains in these patients. Advanced rectal changes indicate that a pan-coloproctectomy with a permanent ileostomy should be performed. Radical surgery carries a reasonably low mortality rate, and results in subsequent replacement of an acute life-threatening condition or a chronic disabling disease, by a state of health and functional fitness.

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