

Chronic Regional Enteritis *

A Survey of One Hundred Twenty-Six Cases Treated at the Massachusetts General Hospital from 1937 to 1954

BENJAMIN B. JACKSON, M.D.

*From the Surgical Services of the Massachusetts General Hospital
and the Harvard Medical School*

THE PROBLEMS in the management of regional enteritis are not completely solved. Regional enteritis characteristically involves the small bowel but it may involve the entire intestinal tract from the duodenum to the anus.¹ Remissions and exacerbations are significant features of this granulomatous disease. Regional enteritis may be as pernicious as any malignant neoplasm. All too vivid to the clinician is the picture of wasting, infection, partial obstruction, anemia, tetany, and avitaminosis.

This study represents a survey of 126 cases of regional enteritis treated at the Massachusetts General Hospital from 1937 to 1954. The follow up period ranged from two to 26 years. Ninety per cent of the cases had been evaluated within a one year period.

The etiology of the disease is unknown. It has been suggested that the granulomatous disease resulted from lymphatic obstruction⁷ occurring in the submucosal layer of the bowel wall. This obstruction seems to be directly responsible for the ulcerations on the mesenteric side of the intestinal wall. The lymph nodes become secondarily involved in this process of elephantiasis. Infection of the ulcers prevents healing and promotes anemia. As the disease progresses there is evidence of healing by heavy scar. The agent initiating endothelial hyperplasia has never been iso-

lated but may well be a toxic fatty acid in a constitutionally susceptible tissue.

Incidence and Recurrence

Van Patter *et al.* reported 600 cases from the Mayo Clinic.⁶ Crohn *et al.* tabulated 562 cases of regional enteritis over a 20-year period at the Mt. Sinai Hospital in New York.² This paper represents a survey of 126 cases of proven chronic regional enteritis treated at the Massachusetts General Hospital from 1937 to 1954. Fourteen different surgeons and nine internists and gastroenterologists participated in the treatment of those cases. There were 48 per cent males and 52 per cent females. The ages ranged from ten to 77 years. The average age was 24 years. No ethnic factors could be demonstrated.

The recurrence rate based on hospital admissions for treatment was 55 per cent. Only 1.6 per cent of the 126 cases revealed spontaneous remission on medical therapy without further evidence of the disease by clinical history or x-ray examinations.

Natural History

The stomach and the duodenum were not involved in this series of cases. There were skip areas in 25 per cent of the patients. The colon was involved in some way in 43 per cent of the patients. Fourteen cases of regional enteritis showed ulcerative colitis involving the entire colon occurring from three weeks to 19 years following the diagnosis of regional enteritis. Thirty-one

* Submitted for publication September 11, 1957.

TABLE I

	Number of Cases
Complications of the Disease	
1. Internal fistulae	35
2. External fistulae	20
3. Hemorrhoids	25
4. Rectal abscesses	21
5. Rectal stricture	4
6. Thrombophlebitis	8
7. Pulmonary infarction	6
8. Anal fissures	18
9. Microcytic hypochromic anemia	64
10. Tetany	15
11. Hypocalcemia	31
12. Renal stones	4
13. Malnutrition	59
14. Macrocytic hyperchromic anemia	4
Complications of the Treatment	
1. Fistulae	28
2. Wound abscess	32
3. Drug addiction	3
4. Dehiscence of wound	3

per cent of the cases of regional enteritis involved the cecum and ascending colon; 5 per cent had disease in the transverse colon; 5 per cent involved the descending colon; and 7 per cent manifested enteritis in the sigmoid. Fifty-five cases or 44 per cent had involvement in the terminal ileum in its distal 15 to 20 cm. The remaining 56 per cent had enteritis above this level in the terminal ileum and 33 per cent of this group had proximal skip-areas.

Sixty per cent of the patients demonstrated a right lower quadrant mass and 28 per cent gave evidence of internal fistulae by x-ray study.

Duodenal ulcer was present in 13 per cent of the cases but none were present in which ulcerative colitis was found involving the entire colon. There was a history of three ulcer perforations. Steroids had not been used in these cases. Carcinoma was not discovered in this group of 126 cases.

Diagnosis

The diagnosis of regional enteritis can be suspected from the clinical history. The

symptom complex usually consists of malaise, easy fatiguability, chills and fever, sweats, weight loss, right lower quadrant and periumbilical cramps, and usually frequent, foul, mushy stools. A right lower quadrant mass and/or rectal abscess, fissure, or fistula with the aforementioned symptoms point toward regional enteritis. The diagnosis can be confirmed by reflux of a barium enema into the terminal ileum if the disease is there. If regional enteritis is suspected, a small bowel study is invaluable in providing knowledge of the lesion and the proximal small bowel. A sigmoidoscopy should be done in all cases. Usually sigmoidoscopy, barium enema, and small bowel study should be employed in that order.

In 25 per cent of the cases the stool was alkaline, foul, mushy, foamy yellow to slate gray, and often contained increased fats, starch, and muscle fibers. The deficiency in fat absorption was usually demonstrable first; whereas carbohydrate metabolism was upset last. The stool was occasionally guaiac positive.

Complications

The overall picture of malnutrition was discernible in about one-half of the cases. Microcytic hypochromic anemia appeared in 51 per cent of the patients. Macrocytic hyperchromic anemia was observed in four cases in which a side to side ileo-transverse colostomy without transection of the bowel was performed eight, eight, ten, and 11 years previously. Hypocalcemia was seen in 25 per cent and tetany was noted in 12 per cent of the 126 cases. Frank avitaminosis was observed in 11 per cent of the patients despite what was thought to be adequate intake of vitamins.

Management

The great need for meticulous evaluation of these patients once the diagnosis is apparent should be stressed. The reason for this careful planning is readily seen if one

notes the recurrence rate of regional enteritis no matter how they are treated. An effort must be made to establish whether the picture of malnutrition is from poor absorption and/or hypermotility or an inadequate program for the patient. A total protein with A/G ratio, vitamin A absorption test, oral glucose tolerance test, carmine marker for hypermotility and a stool examination for occult blood, soaps, neutral fats, fatty acids, muscle fibers, and starch will help make the decision.

Once the period of formal evaluation was completed and treatment had been planned, medical therapy was employed unless the following indications for operation were present: hemorrhage, obstruction, perforation, fistula, or right lower quadrant mass. Medical treatment usually consisted of absolute bed rest, high calorie, low fat, low residue diet, mild sedation, intestinal antibiotics, and antispasmodics. Tween "80" has not been helpful. ACTH and cortisone were disappointing. X-ray treatments to the pelvis have not been helpful and could be harmful in the young female. No successes can be reported from vagotomy in three cases.

There is probably no disease in which it is more difficult to evaluate the role of surgery than in chronic regional enteritis. The indications for operation appear to be for the treatment of the complications, i.e., hemorrhage, obstruction, perforation, external fistulization, and right lower quadrant mass. The presence of a mass suggests internal fistulae or partial obstruction or both. Rarely will a right lower quadrant mass resolve completely in regional enteritis. In this series, 93 per cent of the cases of chronic regional enteritis operated upon, the indication for operation was partial small bowel obstruction. Hemorrhage was an indication in 2 per cent, perforation in 1 per cent and fistula in 4 per cent. The finding of regional enteritis by x-ray is by no means an indication for surgical intervention.

TABLE 2. *Deaths*

Four Deaths in 86 Resections
1. Mesenteric thrombosis.
2. Infectious diarrhea and lower nephron nephrosis.
3. Perforation and peritonitis.
4. <i>S. aureus</i> pneumonia.

Four Deaths in 57 Short Circuiting Operations
1. Rupture of suture line and peritonitis.
2. Rupture of suture line and peritonitis.
3. Hypocalcemia and tetany; marked inanition.
4. Hepatic vein thrombosis.

One of the great problems in the surgical management of chronic enteritis is side tracking operations versus resections. One hundred and four patients in this study underwent operation for regional enteritis. The mortality in 57 short circuiting procedures was 7 per cent and 4.5 per cent in 86 resections. It seemed that morbidity could not be utilized to select the type of operation, for it was approximately the same in both groups. Natural factors such as age of patient, character of local lesion (acutely inflamed or cicatrizing) and location of lesions as well as careful preoperative evaluation and diligent postoperative care appeared to have as much to do with success or failure as the type of technical procedure performed.

At the time of operation the entire gastrointestinal tract is carefully examined for skip areas. A standard operation for all cases of regional enteritis will not suffice. There are occasions when either resection or short circuiting operations or both are advisable. If multiple skip areas are discovered, proximal transection of the small intestine and anastomosis with grossly normal bowel has given the best results. On occasion the disease may be so extensive that no type of operation can be rationally performed. If there is an isolated segment of enteritis in the small bowel or colon, resection with a wide margin of mesentery may be performed.⁵ If the enteritis was confined to the terminal ileum or the ileocecal region, a right colectomy and an ileo

TABLE 3. *Number of Operations per Patient*

Operations	Cases
One	56
Two	19
Three	16
Four	6
Five	4
Six	3

transverse colostomy was favored by the 14 surgeons who treated regional enteritis at the Massachusetts General Hospital. However, no particular effort was made by them to excise the lymph nodes. None of the cases in this series with more than ten feet of small bowel resected did well. The management of regional enteritis and diffuse colon involvement is discussed in another paper.⁴

If a short circuiting procedure is chosen because of severe disease in the terminal ileum, the distal end may be brought out of the inferior part of the wound as a mucus fistula to provide drainage. This seemed reasonable in view of the necessity of re-operating on two cases in which blind loop obstruction developed. If the disease in the defunctioned loop continues to progress, subsequent resection will be indicated. Resection was subsequently required in six of eight cases in which a side to side enter-enterostomy without division of the bowel was done. Twenty-three per cent of the cases with short circuited loops with division later required resection because of persistent symptoms. Fifty-two per cent had good results and 25 per cent of the short circuited loops with division had fair results.

A point at least 12 inches proximal to the diseased bowel is usually chosen for division of the bowel. It should be stressed that the anastomosis be made as carefully as possible no matter what type is elected. This becomes particularly significant when a study of this series revealed obstruction at the site of anastomosis in 14 per cent of

143 operations. Of this 14 per cent only 33 per cent proved to have regional enteritis at re-operation. The obstruction in the remaining 67 per cent was due to scar formation.

Hemorrhage was treated by resection. If possible internal fistulization into the bladder, vagina, duodenum, and sigmoid was managed by resection; otherwise a shunting operation was utilized.

If regional enteritis in the ulcerated or scarred stages be operated upon because of symptoms of acute appendicitis, the appendix can be excised without fear of a fistula only if the ileo-cecal junction and the proximal 30 cm. of the terminal ileum are free of enteritis. Fifteen per cent of this group had an appendectomy during the quiescent phase of the enteritis for acute nonperforate appendicitis and only one patient developed a fecal fistula. In this series of cases the drainage of an abscess adjacent to loops of small bowel involved in enteritis resulted in a fecal fistula in 24 of 30 cases. Drainage in regional enteritis must not be taken lightly. Definite indications for drainage should be present.

Seven cases underwent resection at the time of exploration for symptoms of acute appendicitis with a palpable mass. Six of the seven cases have done well. If multiple skip areas are discovered or the mass cannot be excised safely, a short circuiting operation with complete division is done. If division of the bowel is not done, Garlock and Crohn noted exacerbations as a result of inadequate defunctioning.³

Ano-rectal complications in the form of ischiorectal or perianal abscesses, anal fissures, or anal fistulae with intractability, severe pain, and irritating purulent drainage, were treated with sigmoid colostomy in four cases. Attempts to re-establish intestinal continuity after subsidence of ano-rectal symptoms in two of the four cases met with failure and ushered in the old symptoms.

Even though some of the patients with

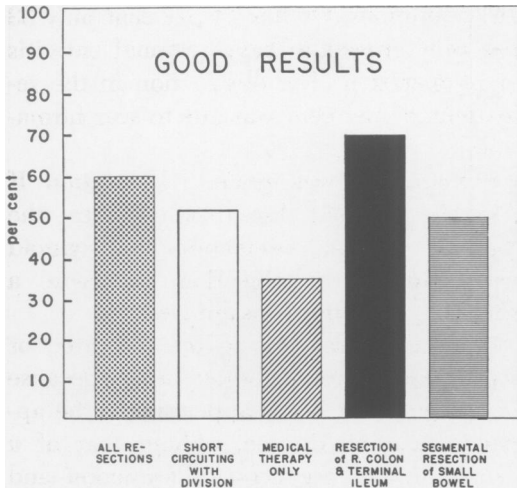


FIG. 1. Results of operations.

quiescent defunctioned loops have displayed healed small bowel at the time of operation for other reasons, it has not been thought wise to re-enter those loops into the intestinal circuit.

Follow up

The follow up period ranged from two to 26 years. Ninety per cent of all patients had been evaluated within a one year period. Recurrences were based on hospital admissions only for treatment. A recurrence has been defined as the return of clinical signs and symptoms with confirmatory x-ray examinations. The over-all recurrence rate was 55 per cent. Resection gave good results in 60 per cent of 86 operations. Excisions of the terminal ileum and the right colon when the diseased area was entirely confined to this region gave good results in 70 per cent; whereas segmental resection of other areas of small bowel produced only 50 per cent good results. Short circuiting procedures with complete transection of the bowel gave good results in 52 per cent of 48 operations. Medical treatment alone provided 35 per cent good results in 22 cases.

The results from both medical and surgical therapy were evaluated in the follow-

ing fashion:

Recurrences within Periods from
 0 to 1 year —poor
 1 to 3 years —fair
 4 or more years—good

In almost all of the cases that required operation and who survived did well for nine months to a year. If their disease returned, it was apt to be between nine to 12 months. If they were able to pass over this peak, it was often three to four years before a recurrence of enteritis was observed. However, just because the disease has not flared in a four year period is no indication that it will not. We have seen four cases that recurred after a ten to 15 year interval and one after 20 years.

The results of all types of treatment of patients with chronic regional enteritis was further analyzed by dividing them into age groups by decades. This analysis is not to be confused with the evaluation of the types of operation, for a patient may have had a poor initial recovery with one operation but finally had an excellent result with another operation. The following table was made to demonstrate those results.

It seems that the most virulent form of regional enteritis occurs within the second

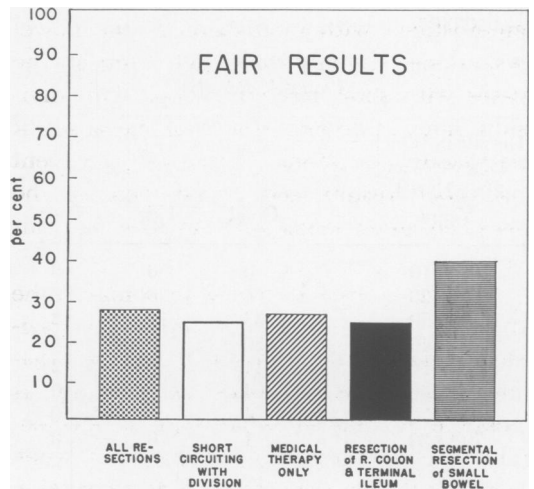


FIG. 2. Results of operations continued.

and third decades of life and the poorest results from all types of therapy occur in the same period. Skip area in this series were most commonly found in the younger age group in contrast to the absence of skip areas in the nine cases after the age of fifty. There were nine cases of chronic regional enteritis first noted after age fifty and of this group eight were treated for partial small bowel obstruction by short circuiting with transection or resection: all with good results. One of the nine cases was treated with a medical regimen also with excellent results. All of the nine cases past 50 years of age had the disease confined to the distal terminal ileum with occasional bridging into the cecum. This group of nine cases is too small to make any valid comments about, but it seems that the course is more tractable in the older age groups.

Summary and Conclusions

1. This study represents a survey of 126 cases of regional enteritis treated at the Massachusetts General Hospital from 1937 to 1954. There was colon involvement in 43 per cent of the cases. Only 44 per cent of the patients had regional enteritis of the distal 15–20 cm. of the ileum and 33 per cent of this group had proximal skip areas. There were skip areas in one-fourth of the 126 cases.

TABLE 4. *Results*

Decade in Which Disease Was First Noted, Years	Good	Fair	Poor
0 to 10	0	0	0
10 to 20	6	0	8
20 to 30	33	11	8
30 to 40	20	9	3
40 to 50	11	6	2
50 to 60	4	0	0
60 to 70	4	0	0
70 to 80	1	0	0
Totals	79	26	21

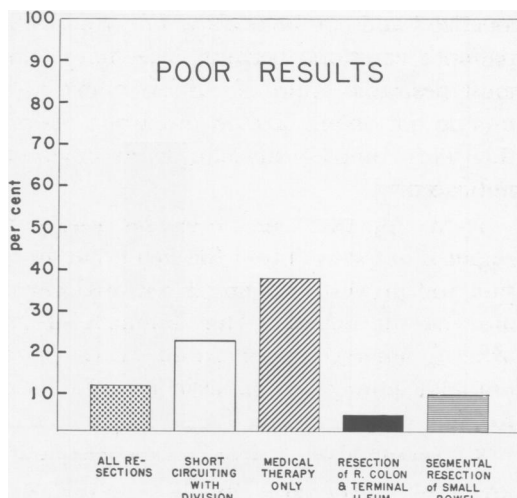


FIG. 3. Results of operations continued.

2. The etiology of the disease is unknown but it has been postulated that regional enteritis is due to lymphatic obstruction secondary to a toxic fatty acid in a constitutionally susceptible tissue.

3. The diagnosis of regional enteritis can be suspected from the clinical history if constantly kept in mind when abdominal disease is a question. It can be confirmed by small bowel motor meal and barium enema.

4. The recurrence rate in this group of cases was 55 per cent when all types of therapy are averaged together. Spontaneous remissions were observed in 1.6 per cent of 126 cases. These facts stress the great need for a meticulous evaluation before any therapy is contemplated.

5. The indications for surgery are the complications of the disease, namely hemorrhage, obstruction, perforation, fistulization, and right lower quadrant mass. The presence of a mass is interpreted as evidence of internal fistulization or partial obstruction.

6. One hundred and four patients of this group of 126 cases underwent surgery for regional enteritis. The mortality in 57 short circuiting operations was 7 per cent and 4.5 per cent in 86 resections. If the enteritis is

localized and can be excised safely and the patient's condition permits, resection is the most desirable approach. If the above factors do not obtain, a short circuiting operation with complete division of the bowel is indicated.

7. An appendix can be excised safely in regional enteritis only if the ileo cecal junction and proximal 30 cm. of terminal ileum are free of disease. The drainage of an abscess adjacent to loops of bowel with regional enteritis frequently led to a fecal fistula.

8. Intractable ano-rectal complications are most satisfactorily handled with a permanent sigmoid colostomy.

9. The follow up period ranged from two to 26 years. Ninety per cent of all patients had been evaluated within a one year period. Resection gave good results in 60 per cent of 86 operations. Excisions of the terminal ileum and right colon had the best prognosis when the disease was confined entirely to this region. Short circuiting operations with complete transection of the bowel gave good results in 52 per cent in 48 operations.

10. Patients may be free of enteritis for 15 to 20 years and then have a recurrence

but patients free from disease longer than four years may have a permanent cure.

11. It seems that the cases of chronic regional enteritis past age 50 have a more benign course than the younger age group.

Acknowledgment

The author gratefully acknowledges the counsel of Dr. Leland S. McKittrick, who carefully reviewed the manuscript.

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