

Comparison of the clinical features of Crohn's disease and ulcerative colitis with onset in childhood or adolescence

William M. Michener, M.D.
Gregory Whelan, M.D.

Department of Gastroenterology

Richard L. Greenstreet, Ph.D.

Department of Biostatistics

Richard G. Farmer, M.D.

Department of Gastroenterology

Our recent publications have described the long-term prognosis of ulcerative colitis and Crohn's disease with onset in childhood and adolescence.¹⁻⁴ The purpose of this paper is to compare the clinical features of ulcerative colitis and Crohn's disease with onset in childhood or adolescence.⁵⁻⁹ Awareness of the differences in the clinical features of these two diseases should be helpful in treating a young person with either disease.

Patients and methods

Between January 1, 1955 and December 31, 1974, 858 patients 20 years old or younger at the time of diagnosis of inflammatory bowel disease were seen at The Cleveland Clinic Foundation. The records of 1050 patients with a possible diagnosis of inflammatory bowel disease from a wide geographic area were screened to obtain the group of patients with confirmed diagnosis.

The clinical, roentgenographic, and histologic features, when available, were reviewed, and according to established criteria patients were classified as having Crohn's disease or ulcerative colitis.¹⁰ The method of follow-up has been described.¹⁻² Trained interviewers contacted the patients by telephone to obtain information that we verified pertaining to complications and clinical course of the

diseases. Crohn's disease had been diagnosed in 522 patients and ulcerative colitis in 336. The data obtained on these patients were entered into a computer and for the purpose of this study the clinical features of 505 patients with Crohn's disease are compared with those of 333 patients with ulcerative colitis. Excluded were 20 cases with insufficient data for comparison. The percentages in *Tables 1 and 2* were compared with use of the chi-square test with the Yates continuity correction.

Results

Demographic features of these patients were noted (*Table 3*). The sex distribution among the patients with ulcerative colitis was virtually identical (168 males and 166 females), whereas 59% of patients with Crohn's disease were male (298 males and 207 females).

Table 1. Complications in inflammatory bowel disease (1955-1974)

No. of patients	Crohn's disease 505 (%)	Ulcerative colitis 333 (%)	
Complications	361 (71.5)	113 (33.8)	p < 0.001
Perianal disease	137 (27.1)	6 (1.8)	p < 0.001
Intestinal obstruction	123 (24.4)	11 (3.3)	p < 0.001
Internal fistulas	83 (16.4)	3 (0.9)	p < 0.001
Growth retardation	40 (7.9)	7 (2.1)	p < 0.001
Megacolon	32 (6.3)	10 (3.0)	p < 0.05
Chronicity	75 (14.9)	73 (21.9)	p < 0.01
Surgery	311 (62.8)	118 (35.6)	p < 0.01

Table 2. Extracolonic complications in inflammatory bowel disease (1955-1974)

No. of patients	Crohn's disease 505 (%)	Ulcerative colitis 333 (%)	
Arthritis	37 (7.3)	24 (7.2)	p > 0.9
Pyoderma	7 (1.4)	6 (1.8)	p > 0.4
Iritis	1 (0.2)	3 (0.9)	p > 0.3
Liver disease	3 (0.6)	5 (1.5)	p > 0.3
Erythema nodosum	14 (2.8)	11 (3.3)	p > 0.8

Table 3. Demographic data

	Crohn's disease	Ulcerative colitis
Sex		
Male	298	168
Female	207	166
Ten years or younger at time of diagnosis	5.6%	12.8%
Duration of follow-up	7.7 years	11.8 years
Died	13.0 (2.8%)	18.0 (5.3%)
Colonic cancer		9

At the time of diagnosis 12.8% of the patients with ulcerative colitis and 5.6% of those with Crohn's disease were less than 10 years of age. The distribution of patients more than 10 years of age was similar for both diseases.¹⁻² The duration of follow-up in the group of patients with ulcerative colitis was 11.8 years and in the group with Crohn's disease, 7.7 years.

The Crohn's group of patients had a higher rate of complications than the ulcerative colitis group (71.5% versus 33.8%) (*Table 1*). This difference in the rate of complications is predictable in light of previous studies, e.g., perianal disease (27.1% versus 1.8%), internal fistulas (16.4% versus 0.9%), intestinal obstruction (24.4% versus 3.3%), growth retardation (7.9% versus 2.1%). Indeed, the expected differences tend to support the validity of the classification system used to separate these two diseases.¹⁰⁻¹¹ Two features worthy of note, however, are the higher prevalence of megacolon (6.3% versus 3.0%) and a lower prevalence of chronicity (14.9% versus 21.9%) in the patients with Crohn's disease.

As might be anticipated, more operations were performed on patients with Crohn's disease (62.8% versus 35.6%). The extracolonic complications of these diseases are listed in *Table 2*. The less common complications of pyoderma, iritis, liver disease, and erythema nodosa occurred with similar frequency in each disorder.

Thirteen of the patients with Crohn's disease died before 1970; the deaths were related to perforations, abscesses, peritonitis, and sepsis. In the group with ulcerative colitis, 18 patients died, 9 of carcinoma of the colon, 1 of lymphoma, 5 of the complications of chronic disease, and 2 of unrelated causes. All patients with ulcerative colitis and cancer had disease involving the entire colon. The average duration of disease to diagnosis of cancer was 14.55 years, with a standard deviation of 3.36 years.

Comparison by decades

During the 20 years of the study, cases of inflammatory bowel disease were diagnosed as either Crohn's disease or ulcerative colitis by established criteria.¹⁰⁻¹¹ Also, during this period, programs of therapy and indications for surgery were still evolving. Therefore, it seemed appropriate to review the data for the two decades, 1955-1964 and 1965-1974, to determine any trends in prognosis or complications. These data are listed in Table 4. The number of patients with ulcerative colitis seen in the second decade increased by 52%; the number with Crohn's disease increased by 243%. Although there were slight differences in the prevalence of perianal disease and growth retardation over the two decades, the most remarkable change appears to be the rate of surgery

in the two conditions. In Crohn's disease, 59.5% of patients underwent operations in the second decade compared with 74.1% in the first decade, and in patients with ulcerative colitis the rate fell from 49.2% to 26.6%.

Discussion

This study offers an opportunity to review our experience with the two major types of inflammatory bowel disease in children and adolescents referred to a tertiary care institution for treatment. After review of each case with use of current definitions of Crohn's disease and ulcerative colitis, it is evident that the problems of perianal disease, internal fistula, and intestinal obstruction are similar to those observed in other large series.⁹⁻¹¹ That megacolon does occur with a higher frequency in Crohn's disease than in ulcerative colitis is not generally appreciated. Whether this complication will be confirmed by future studies or is merely unique to the type of patient referred to the Cleveland Clinic for treatment cannot be determined at this time, but it is noteworthy that this complication has occurred with similar frequency in both diseases during both decades studied.

The most significant observation in this study is the fall in the surgical rate during the second decade (ulcerative colitis 49.2% to 26.6% and Crohn's dis-

Table 4. Complications in inflammatory bowel disease

Number	1955-1964		1965-1974	
	Crohn's disease 114(%)	Ulcerative colitis 132(%)	Crohn's disease 391(%)	Ulcerative colitis 201(%)
Prevalence of complications	86 (75.4)	54 (40.9)	275 (70.3)	59 (29.2)
Perianal disease	43 (37.7)	1 (0.8)	94 (24.0)	5 (2.5)
Intestinal obstruction	24 (21.1)	7 (5.3)	99 (25.3)	4 (2.0)
Internal fistulas	15 (13.2)	2 (1.5)	68 (17.4)	1 (0.5)
Growth retardation	3 (2.6)	1 (0.8)	37 (9.5)	6 (3.0)
Megacolon	8 (7.0)	3 (2.3)	24 (6.1)	7 (3.5)
Chronicity	19 (16.7)	36 (27.3)	56 (14.3)	37 (18.3)
Surgery	83 (74.1)	65 (49.2)	228 (59.5)	53 (26.6)

ease 74.1% to 59.5%). There are several possible explanations. One is that these patients were referred earlier in the course of their illness, and hence were not as ill and responded better to medical therapy. In the group of patients with ulcerative colitis, 65% had had symptoms for more than 6 months, a rather long period before the diagnosis was established. As far as can be determined, no evidence exists to suggest that referral patterns have been altered to include large numbers of patients not as sick. Has there been a change in the medical therapy program resulting in fewer patients undergoing surgery? Certainly, no new drugs had been introduced during these two decades. Progress in our understanding and better supportive care utilizing corticosteroids, antibiotics, hyperalimentation, and principles of intensive care may account for some of the decrease in operative rates. It is tempting to attribute much of the decrease to an improved response to medical therapy, but this cannot be determined from the data. Undoubtedly, during these decades, the development of more specific indications as to when and when not to operate also contributed to the decrease in surgery during the second decade.

Conclusion

From 1955 through 1974, 838 patients (505 with Crohn's disease and 333 with ulcerative colitis) with inflammatory bowel disease diagnosed before age 21 years received medical and surgical therapy at the Cleveland Clinic. During this period a chronic course was the only common (>10%) complication seen in those patients with ulcerative colitis, whereas in addition to chronicity, perianal disease, intestinal obstruction and internal fistulas all occurred with a prevalence of greater than 10% in those patients with Crohn's disease. Megacolon

was more common in Crohn's disease, remaining steady at 6% to 7% in both decades. The most important observation noted was the fall in surgical rate in the second decade in both groups of patients.

This study reaffirms the chronic and recurrent nature of inflammatory bowel disease with onset in childhood and adolescence but notes a marked divergence in the clinical course of patients with Crohn's disease and ulcerative colitis.

References

1. Michener WM, Farmer RG, Mortimer EA. Long term prognosis of ulcerative colitis with onset in childhood or adolescence. *J Clin Gastroenterol* 1979; **1**: 301-5.
2. Farmer RG, Michener WM. Prognosis of Crohn's disease with onset in childhood or adolescence. *Dig Dis Sci* 1979; **24**: 752-7.
3. Farmer RG, Hawk WA, Turnbull RB Jr. Clinical patterns in Crohn's disease; a statistical study of 615 cases. *Gastroenterology* 1975; **68**: 627-35.
4. Farmer RG, Hawk WA, Turnbull RB Jr. Indications for surgery in Crohn's disease; analysis of 500 cases. *Gastroenterology* 1976; **71**: 245-50.
5. Farmer RG, Michener WM, Mortimer EA. Studies of family history among patients with inflammatory bowel disease. *Clin Gastroenterol* 1980; **9**: 271-7.
6. Korelitz BI, Gribetz D, Kopel FB. Granulomatous colitis in children; a study of 25 cases and comparison with ulcerative colitis. *Pediatrics* 1968; **42**: 446-57.
7. Ament ME. Inflammatory disease of the colon; ulcerative colitis and Crohn's colitis. *J Pediatr* 1975; **86**: 322-34.
8. Poley JR. Chronic inflammatory bowel disease in children and adolescents. Part I. *South Med J* 1978; **71**: 935-48.
9. Poley JR. Chronic inflammatory bowel disease in children and adolescents. Part II. *South Med J* 1978; **71**: 1123-33.
10. Lockhart-Mummery HE, Morson BC. Crohn's disease (regional enteritis) of the large intestine and its distinction from ulcerative colitis. *Gut* 1960; **1**: 87-105.
11. Lennard-Jones JE, Ritchie JK, Zohrab WJ. Proctocolitis and Crohn's disease of the colon; a comparison of the clinical course. *Gut* 1976; **17**: 477-82.