Changing concepts of inflammatory disease of the colon in children

Richard G. Farmer, M.D.

Department of Gastroenterology

William M. Michener, M.D.

Department of Pediatrics and Adolescent Medicine

Recent experience with inflammatory disease of the colon in adults has shown a striking increase in the number of patients with transmural colitis (Crohn's disease of the colon, granulomatous colitis) and a decrease in the number of patients with ulcerative colitis.[1] A review of cases of inflammatory bowel disease in children examined at the Cleveland Clinic from 1965 to 1970 documents this trend. Until recently, there has been much confusion concerning the differential diagnosis between ulcerative colitis and Crohn's disease and, thus some patients may have been misclassified. In children, the variable natural history of the disease, the hesitance to do repeated roentgenographic studies of the gastrointestinal tract, and the reluctance to subject children to proctosigmoidoscopy have contributed to misclassification of these two diseases. Disregarding the incidence of cancer, it is not known whether the long-term prognosis in the two diseases is widely different. Previous studies have indicated that the prognosis is poor for children who have the onset of inflammatory bowel disease before age 14. The condition frequently requires operations, and there is a greatly increased risk of cancer and a higher mortality than among patients in whom inflammatory bowel disease develops at a later age.[2, 3] A recent study from the Mayo

Clinic of the long-range prognosis of Crohn's disease following onset in childhood seems to indicate a poor prognosis, and an even higher risk of cancer than was originally thought.[4] Other investigators have not duplicated these results and the controversy continues. However, there does appear to be significantly less incidence of cancer among patients with Crohn's disease than among those with ulcerative colitis.

Table 1. Crohn's disease in children, 1965–1970

Age at diagnosis, yr	No. of patients
14	28
13	12
12	12
11	11
10	4
9	3
8	2
7	0
6	1
5	1
4	1
Total	75

Table 2. Ulcerative colitis in children, 1965–1970

Age at diagnosis. yr	No. of patients
14	10
13	10
12	10
11	7
10	3
9	1
8	5
7	2
6	0
5	2
4	1
Total	51

Although inflammatory bowel disease of both the ulcerative colitis and Crohn's disease types is regarded as a disease of young adults, it is relatively unusual in children younger than age 14. In a study of 615 consecutive new cases of Crohn's disease at the Cleveland Clinic during a 4-year period (1966–1969), it was found that 169 (27%) of these patients had the onset of Crohn's disease before age 20. By contrast, only 42 patients (7%) were younger than 15 at the time of the original diagnosis. Therefore, Crohn's disease can be regarded primarily as a condition affecting late adolescence.

At the Cleveland Clinic in the 6-year period (1965–1970), a new diagnosis of inflammatory disease of the colon was established in 126 children younger than age 14. There were 75 with Crohn's disease and 51 with ulcerative colitis. The majority of these children were between the ages of 12 and 14 at the time of diagnosis (52 had Crohn's disease and 30 ulcerative colitis). Fewer patients had onset before age 10 (8 had transmural colitis and 11 ulcerative colitis). *Tables 1 and 2* list the ages of the patients at the time of the original diagnosis. During this 6-year period, there was a gradual decrease in the number of patients with ulcerative colitis, and only 12 new diagnoses were established after 1967. In contrast, the number of patients with Crohn's disease increased each year. The diagnostic criteria were those previously documented to differentiate between the two diseases;[1] diagnoses may be difficult for patients with purely colonic disease. Under these circumstances, proctosigmoidoscopy was often the most valuable single diagnostic procedure for differentiation. For patients with ulcerative

colitis, the rectal mucosa was found to be uniformly granular and friable and without significant anal canal disease. For patients with Crohn's disease, there was a considerable degree of atypicality to the rectal mucosa, as well as a much higher incidence of changes in the anal canal and perianal region. Of the 51 patients with ulcerative colitis, eight had proctosigmoiditis only. The increasing incidence of proctosigmoiditis in patients with ulcerative colitis has also been observed among adults. In a study which we reported consisting of 276 patients with proctosigmoiditis treated during a 20-year period, it was found that the prognosis was favorable for 90% of these patients and progression of disease occurred in less than 10% of the entire group.[5]

The general observation that the diagnosis of ulcerative colitis is somewhat less frequent and that patients with ulcerative colitis have somewhat less severe illnesses, has also been observed by others.[6] In the group of patients with ulcerative colitis, five had rectal prolapse as a significant early manifestation. Rectal prolapse may be associated with inflammatory bowel disease or may closely mimic ulcerative colitis in children.[6] When associated with inflammatory bowel disease, rectal prolapse usually occurs in those children with severe muscle wasting.

Of these 51 patients, 11 required colectomy and four had toxic megacolon. Ileorectal anastomosis was performed satisfactorily for six of these patients.

Of these 75 patients with transmural (Crohn's) colitis, 27 required operation. Toxic megacolon developed in three; five underwent successful ileorectal anastomosis.

It is concluded that Crohn's disease in children, as in adults, occurs now considerably more frequently than ulcerative colitis. Further, the disease appears to be more severe, response to medical therapy is less predictable, and operation appears to be required more often than in patients with ulcerative colitis. Although our experience has not indicated an increased risk of cancer among patients in whom the onset of Crohn's disease was in childhood, long-term follow-up studies are clearly required to determine whether the prognosis in Crohn's disease is different from that of ulcerative colitis. Recent clinical experience seems to indicate that such is the case, but the long-term prognosis requires further clarification.

References

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