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Original Article

Clinical Characteristics and Long-term Outcomes of Pediatric Ulcerative Colitis: A Single-Center Experience in Korea

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Background/Aims: Although pediatric ulcerative colitis (UC) has a different phenotype and clinical course than adult UC, its clinical features and outcomes are poorly defined, especially in Asian populations. This study investigated the clinical features and long-term outcomes of pediatric UC in a Korean population.

Methods: We retrospectively analyzed 208 patients aged <18 years diagnosed with UC between 1987 and 2013. The patient characteristics at diagnosis according to the Paris classification and the clinical course were analyzed.

Results: The male-to-female ratio was 1.3:1, and the median patient age was 15.5 years. At diagnosis, 28.8% of patients had proctitis (E1), 27.8%, left-sided colitis (E2); 5.2%, extensive colitis (E3); and 38.2%, pancolitis (E4). The cumulative probabilities of extension after 5, 10, 15, and 20 years were 32.7%, 40.4%, 52.5%, and 65.8%, respectively. Eighteen patients underwent colectomy, and three patients had colorectal cancer. The cumulative probabilities of colectomy after 5, 10, 15, and 20 years were 7.1%, 8.9%, 12.6%, and 15.6%, and those of colorectal cancer after 10, 15, and 20 years were 0%, 2.1%, and 12.0%, respectively. The disease extent, Pediatric Ulcerative Colitis Activity Index severity, and systemic corticosteroid therapy were significant risk factors for colectomy. The development of primary sclerosing cholangitis was significantly associated with colorectal cancer.

Conclusions: This study provides detailed information on the disease phenotype and long-term clinical outcomes in a large cohort of Korean children with UC. They have extensive disease at diagnosis, a high rate of disease extension, and a low rate of cumulative colectomy. **(Gut Liver 2022;16:236-245)**

Key Words: Pediatrics; Colitis, ulcerative; Outcome; Colectomy; Colorectal neoplasms

INTRODUCTION

Ulcerative colitis (UC) is a chronic and relapsing inflammatory bowel disorder localized to the colon, extending proximally from the rectum. The incidence of UC is increasing worldwide;[1,2] rapid increase in the incidence was reported in Korea and other parts of Asia, despite Asia being previously considered a low incidence area.[3-7]

Unlike Crohn's disease, UC is localized to the colon. Surgery is a treatment option. As UC may be complicated with the development of colorectal cancer, understanding the natural history of UC important. In Western countries, 10% to 25% adult UC patients undergo colectomy within the first 10 years after diagnosis. The relative risk of colorectal cancer is higher in UC patients than in the general population.[8] Some clinical features of pediatric UC differed from those of adult UC in Western studies.[9-11] In Europe and North America, pediatric UC is characterized by extensive and severe disease activity at diagnosis and an overall high disease extension rate at follow-up.[9-11]

Approximately 25% to 30% of pediatric UC patients are corticosteroid-dependent by 1-year,[12,13] and its aggressiveness is associated with a high early colectomy rate.[14]

Limited data are available regarding the long-term prognosis of pediatric UC patients in Asia. This study aimed to investigate the clinical characteristics and long-term outcomes of pediatric UC patients at a single tertiary center in Korea and evaluate the risk of cumulative colectomy and colorectal cancer and the factors associated with pediatric UC in Korea.

MATERIALS AND METHODS

1. Patients

UC patients aged <18 years diagnosed between March 1987 and December 2013 were enrolled and registered at the Inflammatory Bowel Disease (IBD) Center and the Children's Hospital of the Asan Medical Center, a tertiary-care referral hospital. The diagnosis was based on conventional clinical, upper and lower endoscopic, radiological, and histological criteria. Indeterminate colitis, infections, or other recognized causes of intestinal inflammation were excluded.[15] Patients who met the Pediatric Inflammatory Bowel Disease-class 1 features of the revised Porto classification were diagnosed with Crohn's disease and were also excluded.[16]

2. Study design

All data were collected from the IBD registry of the Asan Medical Center or obtained by retrospective medical record review. Baseline patient demographic and clinical characteristics, including age, sex, age at diagnosis, time between symptom onset and UC diagnosis, family history of IBD, extraintestinal manifestations (EIMs), and location, were recorded.

To evaluate long-term disease outcomes, we also noted the initiation of various medications, changes in disease extent, and cumulative probabilities of colectomy and colorectal cancer and analyzed the risk factors for colectomy and development of colorectal cancer. Additionally, to evaluate changes in baseline characteristics, treatment policies, and prognosis of UC, we divided the patients into three sequential subcohorts based on the date of diagnosis (subcohorts: 1, 1986–2000; 2, 2001–2005; and 3, 2006–2013), as used in our previous study on pediatric Crohn's disease patients.[17] Age at diagnosis was divided into three groups (A1a, 0–9 years; A1b, 10–16 years; and A2, ≥17 years) according to Paris classification.[18] Baseline and clinical characteristics were compared among the groups.

The disease extent was assessed using the Paris classification: proctitis (E1, disease limited to the rectum), left-sided colitis (E2, disease distal to the splenic flexure), extensive colitis (E3, disease with distal hepatic flexure), and pancolitis (E4, disease proximal to the hepatic flexure). We assessed the disease activity at diagnosis using the Pediatric Ulcerative Colitis Activity Index (PUCAI).[19]

This retrospective analysis was approved by the Institutional Review Board of Asan Medical Center (IRB number: 2015-0125). We received informed consents from all patients enrolled.

3. Statistical analysis

We used the chi-square or the Fisher exact tests to

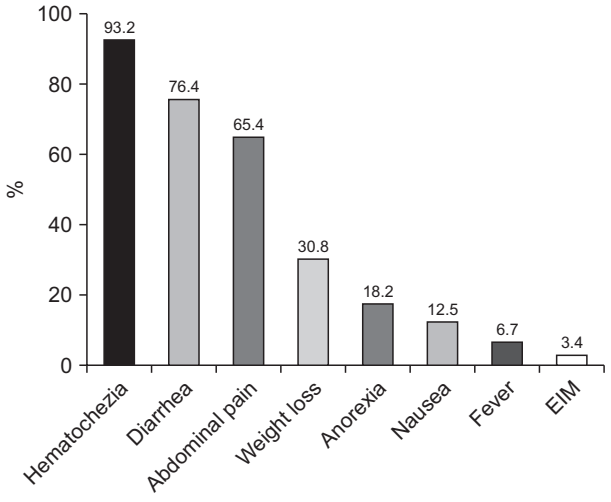

Fig. 1. Clinical manifestations at diagnosis.
EIM, extraintestinal manifestation.

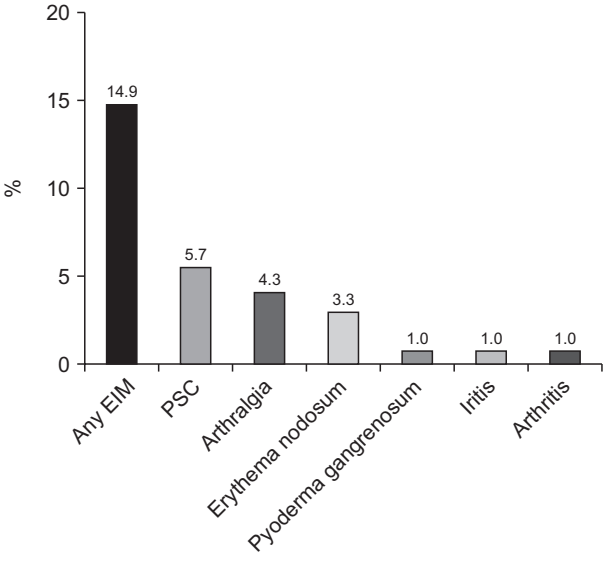

Fig. 2. Extraintestinal manifestations.
EIM, extraintestinal manifestation; PSC, primary sclerosing cholangitis.

compare the binominal variables. The cumulative risks of disease extension, colectomy, and colorectal cancer were calculated using the Kaplan-Meier method. Differences between the Kaplan-Meier curves were compared using log-rank tests. Hazard ratios could not be obtained and multivariate analysis could not be performed owing to the small numbers of incident cases. p-values <0.05 were considered statistically significant. Statistical analysis was performed using IBM SPSS Statistics for Windows, version 20.0 (IBM Corp., Armonk, NY, USA).

RESULTS

1. Basic patient characteristics and clinical features

From 1986 to 2013, we identified 208 UC patients (119 males [57.2%] and 89 females [42.8%], male-to-female ratio=1.3:1). The median age at diagnosis was 15.5 years (range, 1.3 to 17.9 years). The median follow-up duration was 6.5 years (range, 0.3 to 29.3 years), and the median interval from symptom onset to diagnosis was 2.3 months (range, 0 to 91 months). Nearly half of the patients (43.8%) were diagnosed within 2 months, and 25.0% had a diagnostic delay exceeding 6 months after symptom onset. The predominant symptoms at diagnosis were rectal bleeding (n=194), diarrhea (n=159), and abdominal pain (n=136). Anorexia (n=38), nausea (n=26), fever (n=14), weight loss (n=64), and EIMs (n=7) were also reported (Fig. 1). Twenty-three patients (11.1%) had a family history of IBD, including first-, second-, and third-degree relatives in 16 (7.7%), six (2.9%), and one (0.5%) patient, respectively. Five (2.4%), 158 (76.0%), and 45 (21.6%) patients were aged >10 years (A1a), 10 to 17 years (A1b), and >17 years (A2), respectively. EIMs were detected in seven patients (3.3%) at diagnosis and in 31 patients (14.9%) at follow-up, including primary sclerosing cholangitis (PSC; n=12),

Table 1. Baseline and Clinical Characteristics of the 208 Pediatric Patients with Ulcerative Colitis According to the Year of Diagnosis

Variable	Total (1986–2013)	Subcohort 1 (1986–2000)	Subcohort 2 (2001–2005)	Subcohort 3 (2006–2013)	p-value
No. of patients	208 (100)	51 (24.5)	44 (21.2)	113 (54.3)	
Male sex	119 (57.2)	30 (58.8)	25 (56.8)	64 (56.6)	0.982
Age at diagnosis, yr	15.5 (1.3–17.9)	15.1 (11.0–17.9)	15.4 (4.0–17.9)	15.1 (1.3–17.9)	0.985
Time between onset of symptoms and UC diagnosis, mo	2.3 (0–91.0)	3.0 (0–40.0)	2 (0–70.0)	2.1 (0–91.0)	0.543
Follow-up period, yr	6.5 (0.3–29.3)	18.1 (1.3–29.3)	9.5 (0.3–15.0)	4.0 (0.4–8.4)	
Age at diagnosis					0.723
A1a (0–9 yr)	5 (2.4)	0	1 (2.3)	4 (3.5)	
A1b (10–16 yr)	158 (76.0)	41 (78.4)	32 (72.7)	86 (76.1)	
A2 (≥17 yr)	45 (21.6)	11 (21.6)	11 (25.0)	23 (20.4)	
Family history of IBD	23 (11.1)	7 (13.7)	5 (11.4)	11 (25.0)	0.731
PUCAI score at diagnosis					0.344
Remission	5 (2.6)	0	1 (2.4)	4 (3.6)	
Mild	75 (38.5)	21 (50.0)	12 (28.6)	42 (37.8)	
Moderate	87 (44.6)	18 (42.9)	22 (52.4)	47 (42.3)	
Severe	28 (14.3)	3 (7.1)	7 (16.7)	18 (16.2)	
Extent at diagnosis (n=191)					0.931
Ulcerative proctitis (E1)	55 (28.8)	13 (31.0)	10 (25.0)	32 (29.4)	
Left-sided colitis (E2)	53 (27.8)	13 (31.0)	10 (25.0)	30 (27.5)	
Extensive colitis (E3)	10 (5.2)	1 (2.4)	3 (7.5)	6 (5.5)	
Pancolitis (E4)	73 (38.2)	15 (35.7)	17 (42.5)	41 (37.6)	
Maximum extent during follow-up (n=191)					0.625
Ulcerative proctitis (E1)	32 (16.8)	5 (11.9)	5 (12.5)	22 (20.2)	
Left-sided colitis (E2)	41 (21.5)	11 (26.2)	6 (15.0)	24 (22.0)	
Extensive colitis (E3)	15 (7.8)	4 (9.5)	4 (10.0)	7 (6.4)	
Pancolitis (E4)	103 (53.9)	22 (52.4)	25 (62.5)	56 (51.4)	
Appendiceal orifice inflammation	36 (17.3)	6 (11.8)	12 (27.3)	18 (15.7)	0.127
Backwash ileitis	14 (6.8)	3 (5.9)	3 (6.8)	9 (8.0)	0.955
Extraintestinal manifestations	31 (14.9)	13 (25.5)	6 (13.6)	12 (10.6)	0.045
Systemic corticosteroid	144 (69.2)	36 (70.6)	35 (79.5)	73 (64.6)	0.185
Colectomy	18 (8.7)	7 (13.7)	5 (11.4)	6 (5.3)	0.179
Colorectal cancer	3 (1.4)	3 (5.9)	0	0	0.023

Data are presented as number (%) or median (range).
UC, ulcerative colitis; IBD, inflammatory bowel disease; PUCAI, Pediatric Ulcerative Colitis Activity Index.

arthralgia (n=9), peripheral arthritis (n=2), erythema nodosum (n=7), pyoderma gangrenosum (n=2), and iritis (n=2) (Fig. 2). Three patients had three EIMs (arthralgia and erythema nodosum, erythema nodosum and iritis, and erythema nodosum and PSC). Two patients with PSC underwent liver transplantation. The subcohort analysis showed no significant differences in the sex ratio, age, family history, PUCAI at diagnosis, UC extent at diagnosis or follow-up, or colectomy rate among the three subcohorts. EIMs were more frequent in cohort 1 (p=0.045), and all three colorectal cancer patients were in cohort 1 (p=0.023) (Table 1). In age group analysis, group A1b had a higher proportion of moderate to severe PUCAI (p=0.024) than the other groups, as well as the highest rate of systemic corticosteroid use (p=0.003). All three cancer patients were in group A1b (Supplementary Table 1).

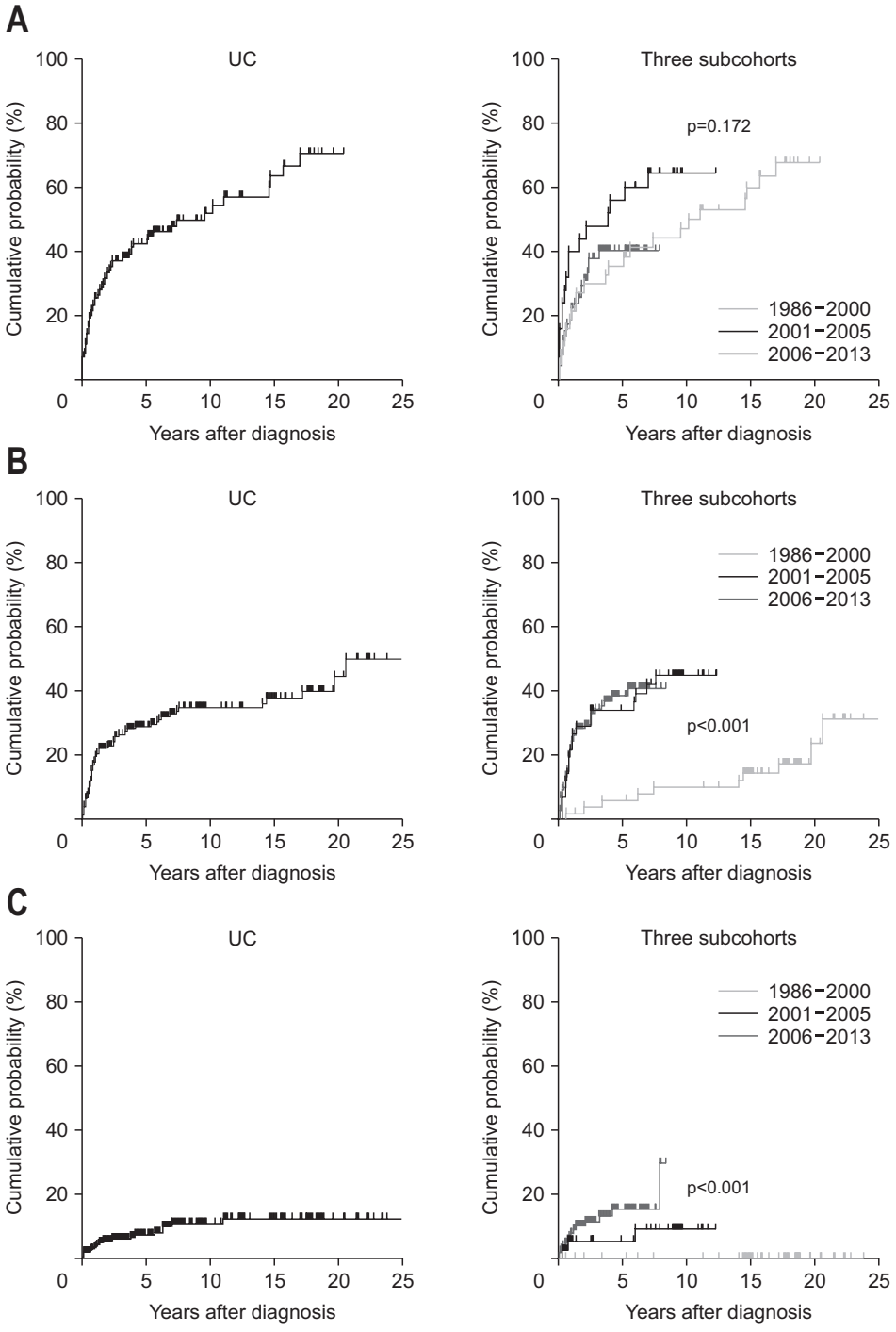

Fig. 3. Cumulative probability of medication uses in all 208 pediatric patients with ulcerative colitis (UC) and in the three subcohorts from the time of diagnosis. (A) Systemic corticosteroids. (B) Thiopurines. (C) Anti-tumor necrosis factor agents.

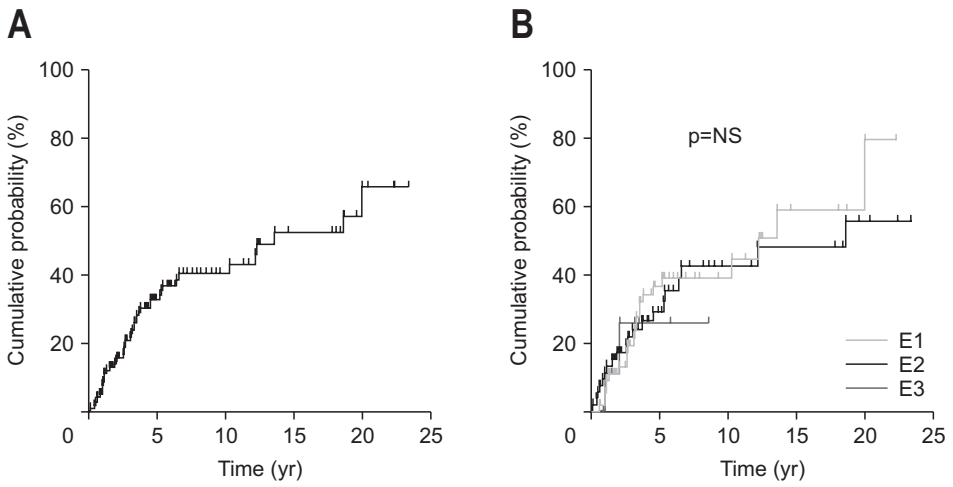

Fig. 4. Cumulative probability of disease extension in 109 patients with E1/E2 ulcerative colitis and three subcohorts classified according to the date of diagnosis. (A) Cumulative probability of extension in E1, E2 groups. (B) Cumulative extension rate in three subcohorts based on the date of diagnosis.
NS, not significant.

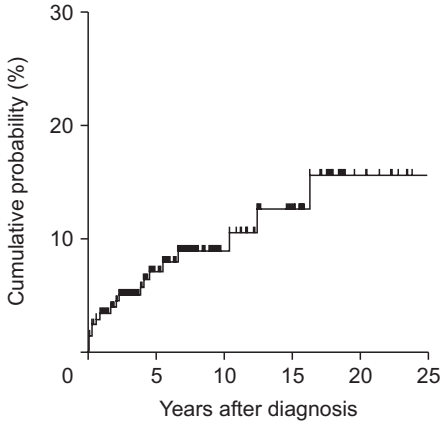

Fig. 5. Cumulative risk of colectomy in 208 pediatric patients with ulcerative colitis.

2. Medical treatment

The proportions of patients who used systemic corticosteroids, thiopurines, and anti-tumor necrosis factor (TNF) agents at diagnosis or during the follow-up were 69.2% (n=144), 34.6% (n=72), and 9.6% (n=20), respectively. The median intervals from the diagnosis of UC to the initiation of systemic corticosteroids, thiopurines, and anti-TNF agents were 0.1 months (range, 0 to 203.6 months), 43.4 months (range, 0 to 247.5 months), and 15.9 months (range, 0.4 to 133.7 months), respectively. The cumulative probabilities of medication use at 1, 5, 10, and 15 years after diagnosis were 54.6%, 64.8%, 70.4%, and 77.1%, respectively, for corticosteroids; 19.3%, 30.1%, 35.8%, and 38.9%, respectively, for thiopurines; and 3.9%, 7.8%, 11.3%, and 12.8%, respectively, for anti-TNF agents (Fig. 3). The cumulative probability of prescription of systemic corticosteroids did not differ significantly between the subcohorts. The cumulative probability of thiopurine prescription was the lowest in subcohort 1 (p<0.001). The cumulative probability of prescription of anti-TNF agents increased over time from subcohort 1 to subcohort 3 (p<0.001). Some patients received other immunomodulators, such as cyclosporine (n=7) and tacrolimus (n=2). Almost all patients (n=198) took 5-aminosalicylates and probiotics, but the exact rate of probiotics usage could not be determined.

3. Disease extent

The most common Paris classification grade for extent at diagnosis among 191 patients with known extent at diagnosis was E4 (n=73, 38.2%). Fifty-five (28.8%), 53 (27.8%), and 10 (5.2%) patients had E1, E2, and E3, respectively. With respect to the maximum extent during follow-up, 32 patients (16.8%) had E1, 41 (21.5%) had E2, 15 (7.8%) had E3, and 103 (53.9%) had E4 (Supplementary Fig. 1). The cumulative probabilities of extension rates (except for the E4 group) after 5, 10, 15, 20 years were 32.7%, 40.4%, 52.5%, and 65.8%, respectively. Among patients with E1 at diagnosis, the cumulative probabilities of extension after 5, 10, 15, and 20 years were 36.6%, 44.6%, 59.0%, and 79.5%, respectively. Among patients with E2, the cumulative probabilities of extension after 5, 10, 15, and 20 years were 29.2%, 42.5%, 48.3%, and 55.7%, respectively. Among patients with E3, the cumulative probability of extension after 5, 10, 15, and 20 years was 25.9%. With respect to disease extension, there were no significant differences among patients with E1, E2, and E3 (Fig. 4).

4. Colectomy

Eighteen patients (8.7%) underwent colectomy soon after diagnosis or during follow-up: 13 patients underwent total proctocolectomy with ileal pouch-anal anastomosis and five patients underwent total colectomy with ileostomy. The cumulative probabilities of colectomy were 7.1%, 8.9%, 12.6%, and 15.6%, at 5, 10, 15, and 20 years after

Table 2. Risk of Colectomy in 208 Pediatric Patients with Ulcerative Colitis Analyzed by the Log-Rank Test

Variable	No. of patients	Colectomy	p-value
Age group			0.385
A1a (0–9 yr)	5	1	
A1b (10–16 yr)	158	15	
A2 (≥17 yr)	45	2	
Sex			0.794
Male	119	11	
Female	89	7	
Family history of IBD			0.352
No	185	17	
Yes	23	1	
Extent at diagnosis			0.007
Ulcerative proctitis (E1)	55	2	
Left-sided colitis (E2)	53	2	
Extensive colitis (E3)	10	0	
Pancolitis (E4)	73	12	
Maximum extent during follow-up			0.002
Ulcerative proctitis (E1)	32	0	
Left-sided colitis (E2)	41	0	
Extensive colitis (E3)	15	0	
Pancolitis (E4)	103	16	
ANCA			0.268
Negative	92	8	
Positive	101	5	
PSC			0.549
Negative	196	16	
Positive	12	2	
EIM			0.835
Negative	174	15	
Positive	31	3	
PUCAI at diagnosis			<0.001
Remission	5	0	
Mild	75	1	
Moderate	87	8	
Severe	28	8	
AOI			0.336
Negative	172	17	
Positive	36	1	
Systemic corticosteroid			0.028
Negative	64	1	
Positive	144	17	
Immunosuppressant			0.273
Negative	132	10	
Positive	72	8	
Anti-TNF-α			0.155
Negative	188	15	
Positive	20	3	

IBD, inflammatory bowel disease; ANCA, antineutrophil cytoplasmic antibody; PSC, primary sclerosing cholangitis; EIM, extraintestinal manifestation; PUCAI, Pediatric Ulcerative Colitis Activity Index; AOI, appendiceal orifice inflammation; TNF, tumor necrosis factor.

diagnosis, respectively (Fig. 5). There were no significant differences in the probability of colectomy among the three subcohorts. Disease extent at diagnosis (p=0.007), maximum extent during follow-up (p=0.002), PUCAI (p<0.001), and use of systemic corticosteroids (p=0.028) were significant risk factors for colectomy, based on the log-rank tests. Age at diagnosis, sex, family history, antineutrophil cytoplasmic antibody, PSC, EIM, and use of anti-TNF-α agents were not associated with the risk of colectomy (Table 2).

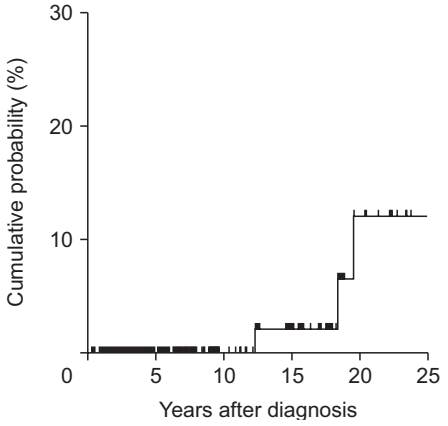

Fig. 6. Cumulative probability of colorectal cancer in 208 pediatric patients with ulcerative colitis.

5. Colorectal cancer

Three patients were diagnosed with colorectal cancer, which developed 12.4, 18.4, and 19.7 years after the diagnosis of UC, respectively. The cumulative risks of colorectal cancer after 10, 15, and 20 years were 0%, 2.1%, and 12%, respectively. No significant differences were observed in the cumulative probabilities of colorectal cancer among the three cohort groups (Fig. 6). Two cancer patients had PSC. Based on the log-rank tests, only PSC was observed to be significantly associated with colorectal cancer (p=0.002) (Table 3).

6. Mortality

By the last follow-up, four patients had died: one of colorectal cancer and one of sepsis after azathioprine-induced neutropenia; the other two causes of death were not related to UC.

DISCUSSION

This report on a large, single-center study applying Paris classifications to an Asian pediatric UC population provides detailed information on the disease phenotype and long-term clinical course. Disease extent at diagnosis, EIMs, and progression of UC extent were similar to the findings reported in Western studies, while the cumulative colectomy rate was lower than that reported previously.[10,20-22]

Table 3. Risk of Colorectal Cancer in 208 Pediatric Patients with Ulcerative Colitis Analyzed by the Log-Rank Test

Variable	No. of patients	Colorectal cancer	p-value
Age group			NC
A1a (0–9 yr)	5	0	
A1b (10–16 yr)	158	3	
A2 (17 yr)	45	0	
Sex			0.759
Male	119	2	
Female	89	1	
Family history of IBD			0.410
No	185	3	
Yes	23	0	
Extent at diagnosis			NC
Ulcerative proctitis (E1)	55	0	
Left-sided colitis (E2)	53	0	
Extensive colitis (E3)	10	0	
Pancolitis (E4)	73	3	
Maximum extent during follow-up			0.302
Ulcerative proctitis (E1)	32	0	
Left-sided colitis (E2)	41	0	
Extensive colitis (E3)	15	0	
Pancolitis (E4)	103	3	
ANCA			0.642
Negative	92	2	
Positive	101	1	
PSC			0.002
Negative	196	1	
Positive	12	2	
EIM			0.113
Negative	174	2	
Positive	31	1	
PUCAI at diagnosis			NC
Remission	5	0	
Mild	75	1	
Moderate	87	8	
Severe	28	8	
AOI			0.579
Negative	172	3	
Positive	36	0	
Systemic corticosteroid			0.359
Negative	64	0	
Positive	144	3	
Immunosuppressant			0.594
Negative	132	2	
Positive	72	1	
Anti-TNF-α			NC
Negative	188	3	
Positive	20	0	

IBD, inflammatory bowel disease; ANCA, antineutrophil cytoplasmic antibody; PSC, primary sclerosing cholangitis; EIM, extraintestinal manifestation; PUCAI, Pediatric Ulcerative Colitis Activity Index; AOI, appendiceal orifice inflammation; TNF, tumor necrosis factor; NC, not calculable.

Recent studies have reported on the association between pediatric UC and higher proportions of pancolitis.[23-25] In our study, 65.8% of patients without pancolitis had extension during follow-up and pancolitis became the most predominant type. Pediatric UC patients have more extensive disease, are more frequently treated with immunomodulators, have a higher frequency of steroid dependency, and have a more severe disease course than adult UC patients.[23] The presence of EIMs and high PUCAI scores at diagnosis are related to disease extension.[11,24] A family history of IBD and diagnostic delays >6 months are also associated with colonic extension.[11,25] A recent meta-analysis reported that 10% to 20% of pediatric patients experienced EIMs and that PSC was observed in 5% to 12% of patients,[22] similar to the rates of EIMs (16.3%) and PSC (5.8%) noted in our study. EIMs have been identified as a marker of poor prognosis because they are associated with increased immunogenicity and severity and extension of UC.[26,27] The reported prevalence of PSC among adult Korean UC patients is 1.1%,[28] lower than the 5.8% reported in our study on pediatric UC patients. Seven of the 12 PSC patients had PSC at initial diagnosis. Asan Medical Center is the leading hospital for liver transplantation in South Korea; thus, many patients with liver disease are transferred to this institution, which may have increased the frequency of PSC in pediatric UC patients.

The cumulative probability of colectomy in our study was lower than reported in previous studies that estimated that approximately 20% of patients undergo operations after 10 years of follow-up.[22] Higher PUCAI scores at diagnosis and male sex are associated with an increased risk of colectomy.[9,29] The colectomy rate is also associated with the presence of EIMs at diagnosis and is higher among patients with disease extension.[11] Our study revealed a similar prevalence of EIMs and disease extent; however, the cumulative probability of colectomy was lower than that reported in Western studies, suggesting that Korean UC children tend to experience a more benign clinical course than children in the Western populations.

Lee *et al.*[30] reported that the proportions of E1, E2, and E3 disease extent at diagnosis were 44.8%, 25.9%, and 29.3%, respectively, among adult Korean UC patients, and the overall colectomy rate was 7.5%. The cumulative probabilities of colectomy at 10 and 20 years after diagnosis were 7.8% and 14.2%, respectively, in adult-onset UC, which are lower than 8.9% and 15.6% reported in our study. Our patients had a lower rate of proctitis and a high proportion of extensive disease at diagnosis than those reported previously in patients with adult-onset UC; however, they had similar cumulative colectomy rates at 20 years. The administration rates for corticosteroids, thiopurines, and anti-TNF agents were higher in our pediatric cohort than those reported for Korean adults. The median interval from the diagnosis of UC to the start of medication in

children was shorter than that in adults. These results suggest that the disease course is more aggressive in pediatric UC patients than in adult UC patients in Korea.

The cumulative probability of medication uses in our three subcohorts revealed the frequent early administration of immunomodulators and anti-TNF agents in the most recent subcohort; however, the risks of colectomy and colorectal cancer did not differ significantly among the subcohorts. Moreover, it is difficult to confirm the effects of these medications on the decreasing rates of surgery or colorectal cancer because immunomodulators and anti-TNF agents are used in patients with severe disease activity.

During follow-up, three patients were diagnosed with colorectal cancer and two patients died from disease-related causes. The crude cancer rate was 1.4%, while the disease-related mortality was 0.96%, similar to rates reported previously. A population-based study reported a mortality rate of 0.84% among pediatric UC patients, which did not differ from that in the reference population. The reported crude cancer was 1.3%, with a significant three-fold increased risk of cancer, and the cumulative probability of cancer was 3.7% after 17 years of follow-up.[25] In a study of adults conducted at our center, 18 patients developed colorectal cancer among 2,798 UC patients during 22,272 person-years of follow-up.[31] In a Swiss IBD cohort study, approximately 4% of UC patients had PSC, which was associated with considerable long-term morbidity and mortality. Significantly more patients with UC and PSC developed colorectal cancer than UC patients without PSC. Extensive disease and pancolitis increased the risk of PSC development.[32] Our results indicate that PSC is a strong risk factor for the development of colorectal cancer.

This study has several limitations. First, it was performed in a single tertiary center; therefore, the findings may not reflect the general prevalence and prognosis of all Korean pediatric UC patients. Second, we did not use the revised Porto criteria[15] to diagnose UC; therefore, we could not distinguish between UC, atypical UC, and IBD-unclassified.[16] Further studies applying the Porto criteria are needed to describe pediatric UC in our context. Finally, this study included results from an era before the introduction of biologic therapy as the first-line treatment. Thus, we could not evaluate the effect of medications on the long-term outcomes of pediatric UC. Further well-designed prospective studies are warranted to confirm the effects of drugs on the natural history of pediatric UC.

In conclusion, pediatric UC at our center was characterized by extensive disease at diagnosis and a high rate of disease extension at follow-up. The cumulative colectomy rate was lower in our Korean cohort than in Western children. Disease extent, severe PUCAI, and systemic steroid treatment were associated with a higher probability of colectomy. PSC was determined as a significant risk factor for the development of colorectal cancer.

CONFLICTS OF INTEREST

No potential conflict of interest relevant to this article was reported.

AUTHOR CONTRIBUTIONS

Study design: J.J., K.M.K., S.H.L., I.S.J., J.C., H.J.K., S.H.O. Data analysis: J.J., K.M.K., S.H.L., I.S.J., J.C., H.J.K., S.H.O., D.Y.K., H.S.L., S.H.P., B.D.Y., S.K.Y. Writing - original draft: J.J., K.M.K. Writing - review & editing: S.K.Y.

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SUPPLEMENTARY MATERIALS

Supplementary materials can be accessed at https://doi.org/10.5009/gnl20337.

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