Spontaneous Resolution of Rectal Polyps in Patients with Familial Polyposis Following Abdominal Colectomy and Ileorectal Anastomosis

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One hundred sixteen patients were reviewed after abdominal colectomy and ileorectal anastomosis (IRA) for familial polyposis to determine the rate of postoperative spontaneous regression of rectal polyps. The failure of the IRA procedure was correlated with the preoperative number of rectal polyps and the degree of rectal polyp regression. Spontaneous resolution of rectal polyps occurred in 64 percent of the patients (complete 38 percent, partial 26 percent). In those patients initially having complete resolution, 55 percent redeveloped polyps during follow-up. With a mean follow-up of 9.3 years, seven patients have developed rectal cancer. Rectal cancer development was more common in those patients who had innumerable rectal polyps prior to IRA. Initial polyp regression did not preclude later development of rectal cancer. There were 11 deaths during the follow-up period, but only one of these was from rectal cancer. Abdominal colectomy and IRA is an effective treatment for familial polyposis. Spontaneous regression of polyps occurred in 64 percent of patients, but continuous and complete follow-up is necessary to fulgurate recurrent polyps and to screen for the development of cancer. [Key words: Familial polyposis; Polyp regression; Ileorectal anastomosis]

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FAMILIAL POLYPOSIS IS a rare autosomal dominant disorder characterized by the development of multiple adenomas of the colon which, if untreated, will uniformly degenerate into colonic carcinomas that are often multiple.[1] Cripps (1882) first recorded the existence of a familial disorder that involved multiple colonic adenomas. Handford, in 1890, recognized that these multiple colonic adenomas were associated with a risk of carcinoma.[1]

The St. Mark's series of familial polyposis (FPC) confirmed that patients left untreated developed colonic carcinoma at a mean age of 39 years.[1] It has become standard practice to screen familial patients at risk for FPC and then to perform prophylactic surgery to prevent the inevitable development of colonic carcinoma.

A variety of surgical options have been suggested to treat FPC. Bess[2] and co-authors from the Mayo Clinic support total proctocolectomy and ileostomy because, in their series, there was a high incidence (up to 32 percent) of rectal cancer in the rectal remnant after ileorectal anastomosis (IRA). Watne and co-workers[3] also believe that the long-term risk of cancer developing in the rectal remnant after IRA makes removal of the colon and rectum the preferred option for these patients. Other groups, particularly at the Cleveland Clinic[4-6] and St. Mark's Hospital[1] have shown that if the ileorectal (IRA) is done with a short rectal remnant of 12 to 15 cm, and careful follow-up is done with fulguration of remaining rectal polyps, the risk of rectal cancer developing is quite small and controllable.

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Spontaneous regression of rectal polyps following colectomy and IRA has been described.[3,7-9] The Cleveland Clinic experience with IRA in patients with FPC was reviewed to determine the rate of spontaneous rectal polyp regression, the long-term status of the remaining rectum, and whether the failures of the IRA procedure could be predicted from the preoperative number of rectal polyps or from the degree of rectal polyp regression.

Materials and Methods

The case records of 99 familial polyposis patients who had total abdominal colectomy with ileorectal anastomosis performed at the Cleveland Clinic were reviewed. In addition, 17 patients who underwent surgery at outside institutions, but with ongoing surveillance of their rectal remnant performed at the Cleveland Clinic, were included in the analysis. A previous series[6] from this institution included patients with familial polyposis who were referred to the Cleveland Clinic specifically for the management of established cancer after IRA or desmoid tumors. These patients would have biased this series for the purpose of determining the efficacy of IRA in treating FPC and are excluded.

These 116 patients underwent surgery from August 1947 until July 1985. There were 56 men and 60 women with a median age of 23.8 years (range, nine to 57 years) at the time of IRA. Records were reviewed for the presence and number of colon and rectal polyps present preoperatively and at follow-up. The operation performed involved removal of the intra-abdominal colon with ileorectal-rectal anastomosis at the 12 to 15 cm level. The mean level of anastomosis was 14.0 cm as measured postoperatively by the rigid proctoscope.

The number of rectal polyps preoperatively and postoperatively were divided into four groups (Table 1) as determined by the number of polyps recorded by the examining physician at rigid proctoscopy using the standard 25-cm proctoscope. The patient was considered to have complete resolution of rectal polyps if the patient had a total absence of rectal polyps at the first postoperative proctoscopy (median time, six months from date of ileorectal anastomosis). The patient was considered to have partial regression of rectal polyps if the number of rectal polyps seen at the first postoperative proctoscopy fell in the next lower grouping (Table 1). The number of polyps in the large bowel (excluding the rectum) were determined by reviewing colonoscopy or pathology reports.

TABLE 1. *Scoring of Number of Rectal Polyps*

Group	Number of Rectal Polyps Seen at Proctoscopy
1	-
2	<20
3	20-100
4	Innumerable

Patients were seen at intervals from 6 to 12 months postoperatively. Polyps present at follow-up examination were generally destroyed with electrocautery as an office procedure. If the polyps were exceedingly numerous or the patient desired, the patient was admitted and had electrocoagulation of the polyps under a regional or general anesthetic. A registry of familial polyposis patients exists at the Cleveland Clinic and functions to optimize follow-up of patients following ileorectal anastomosis and screening of patients at risk for the disease.

The patients have been followed for a mean of 9.3 years (range, four months to 38 years). Seven patients (6 percent) have been lost to follow-up.

Cross-tabulation data were analyzed with a chi-square or Fischer's exact test depending upon expected cell frequencies. *T* tests were used to compare continuous response variables. The cumulative probability of maintaining the IRA and remaining cancer-free curves generated for patients with innumerable rectal polyps or a countable number at presentation were compared by the generalized Wilcoxon test. SAS was used to provide descriptive statistics, make statistical comparisons, and for management of the data base.[10]

Results

The reasons for presenting for IRA are documented in Fig. 1. Those patients without a family history were included if they had sufficient polyps (>100) in the colon to justify a diagnosis of familial polyposis.

There were 102 patients who had an IRA performed as an initial procedure for familial polyposis. There were 14 patients who had a previous segmental resection of the colon prior to IRA. These patients who had a previous

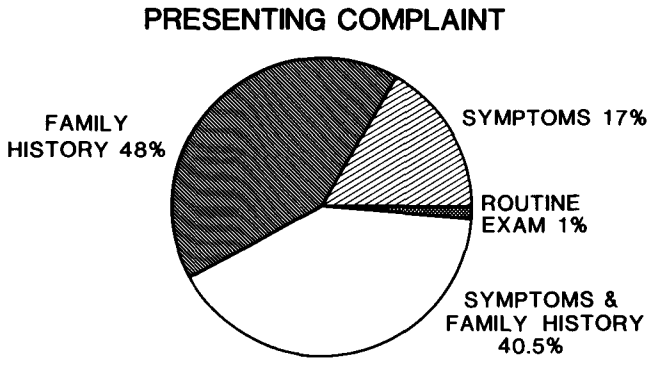

FIG. 1. Indications for proctoscopy at time of diagnosis of familial polyposis.

TABLE 2. *Complications of IRA*

Anastomotic leak	1
Pneumonia	1
UTI	1
Wound disruption	2
Small bowel obstruction	11 (5 requiring surgery)
Ureteric fistula	1

segmental resection presented for IRA at a later age (mean, 40 years; S.D., ten years) as compared to patients who had IRA done as an initial procedure (mean, 24 years; S.D., nine years; $P < .001$). The median frequency of bowel action was 4 bowel movements per day (range, one to 15). There were 98 patients (85 percent), who underwent IRA without complications. Complications are tabulated in Table 2.

Preoperative Number of Rectal and Colonic Polyps: The number of polyps seen in the various parts of the large bowel are illustrated in Fig. 2. A few patients had a predominance of polyps in one area of the colon but, in general, approximately one third of the patients had <20 polyps, one third had more than 20 to 100 polyps, and one third had innumerable polyps in each of the segments of large bowel.

Only two patients had no rectal polyps within reach of the rigid proctoscope. One of these patients (age 57) had a previous left hemicolectomy, which could have accounted for regression of his rectal polyps. The other patient (age 31) had polyps on flexible sigmoidoscopy, but none were seen on rigid proctoscopy.

Resolution of Rectal Polyps: There were 88 patients who had sufficient documentation of the numbers of rectal polyps preoperatively and postoperatively in the first postoperative year. Of these 88 patients, 56 patients (64 percent) showed some spontaneous resolution of their rectal polyps as defined by a downstaging by at least one category of the number of rectal polyps (Table 1). There were 33 patients (38 percent) who had complete spontaneous polyp resolution and 23 (26 percent) with partial polyp resolution. Figure 3 correlates the occurrence of polyp resolution with the number of rectal polyps present preoperatively. Even in those patients with innumerable polyps 17 of 27 (63 percent) had some degree of spontaneous resolution.

Of the 33 patients who initially had complete spontaneous resolution, 15 patients have never redeveloped polyps (mean follow-up, 2.1 years; S.D., 1.08). The other 18 patients have redeveloped polyps (mean follow-up, 6.8 years; S.D., 4.85; $P < .001$). This would suggest that with further follow-up more of these patients will redevelop polyps.

Fulguration of Rectal Polyps at the Time of Surgery: Fulguration of polyps in the rectum was done before or concurrent with abdominal colectomy and IRA in 33 patients (28 percent). In recent years, with the knowledge that spontaneous resolution may occur, fewer patients have had polyp fulguration prior to IRA. The median date of operation for those patients undergoing preoperative rectal polyp fulguration was July 1972 and the median date of operation for those patients not undergoing fulguration was April 1978.

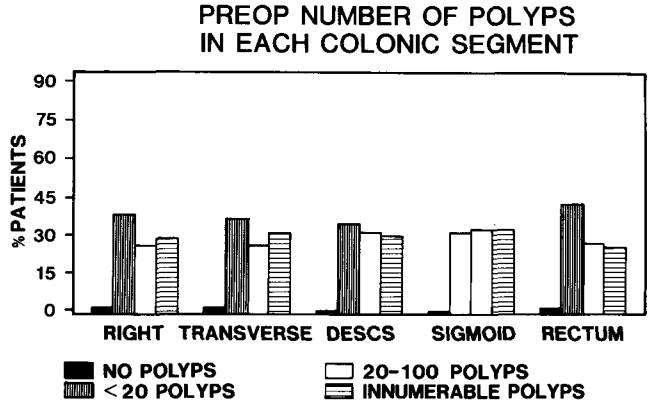

FIG. 2. Number of polyps present in each segment of the colon prior to colectomy and ileorectal anastomosis.

Of the 33 patients undergoing preoperative polyp fulguration, only seven patients were fulgurated to the point where there were no polyps left in the rectum at the time of IRA. Of these seven patients, only one patient remains free of polyps (follow-up, eight years). Four patients had no polyps at their one year examination, but they were reformed at later follow-up and the remaining two patients had reformed polyps by the time of their initial postoperative check.

Relationship of Extracolonic Manifestations to Rectal Polyp Resolution: Extraintestinal manifestations of FPC

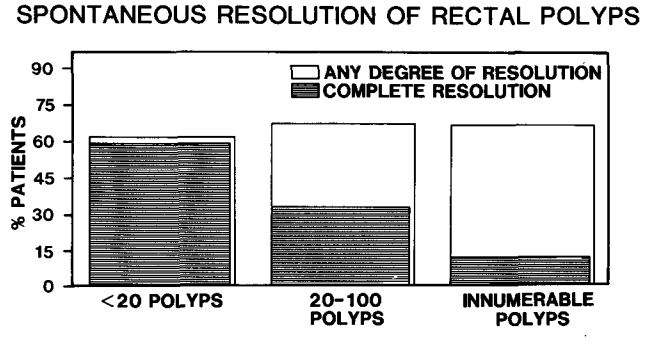

FIG. 3. Correlation of the frequency of patients having spontaneous resolution of rectal polyps with the number of rectal polyps present before colectomy.

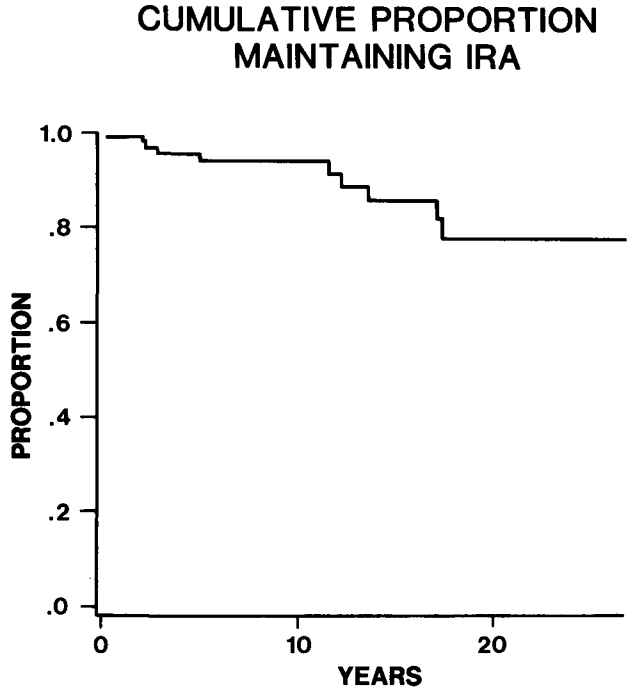

FIG. 4. Life table graph showing that at 25 years 79 percent of the patients still had a functioning rectum after IRA.

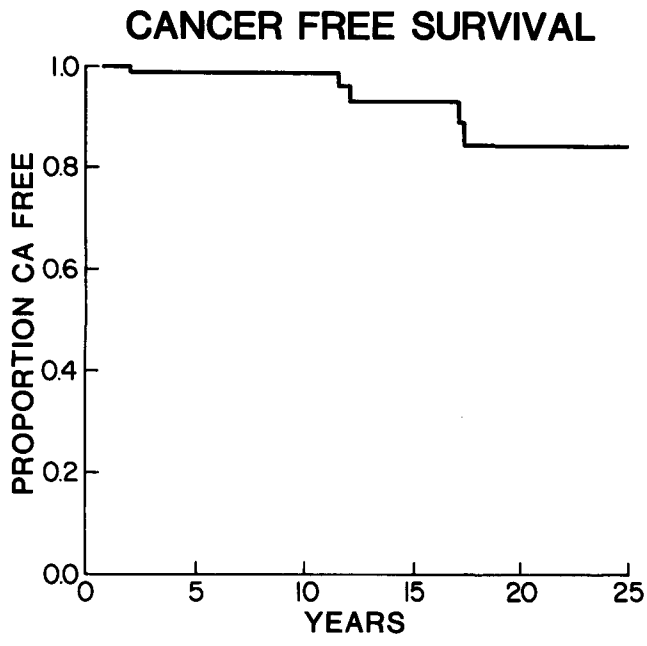

FIG. 5. Life table graph showing 85 percent of the patients to be free of rectal cancer 25 years after IRA.

include osteomas, epidermoid cysts, desmoid tumors, upper gastrointestinal polyps, adrenal tumors and medulloblastomas, and other malignancies. The frequency of extracolonic manifestations of FPC was 71 of 116 (61 percent). In the patients (56) with polyp resolution, the incidence of all extraintestinal manifestations was 38 of 56 (68 percent, $P = .48$).

Relationship of Patient Age to Rectal Polyp Resolution: In patients younger than age 30 years, 45 of 61 (74 percent) had some degree of spontaneous resolution of rectal polyps. In patients older than age 30 years, 9 of 27 (33 percent) had spontaneous resolution of rectal polyps ($P < .001$). This difference was independent of the number of polyps present initially.

Status of Patients: Twelve patients (10 percent) underwent conversion of IRA to proctectomy and ileostomy (10) or J-pouch (2). These patients failed the IRA procedure because of cancer developing in the rectal stump (7), recurrent cancer developing at the IRA anastomotic line (1), uncontrollable polyposis of the rectal remnant (3), and poor functional result (1). Figure 4 is a life table graph showing the cumulative proportion of patients maintaining a functioning rectum after IRA. At 25 years, 79 percent of the patients still had their rectum after IRA.

Rectal cancer has developed after IRA in 7 of 116 (6 percent). Cancer developed an average of 14.7 years after IRA (range, 2 to 32 years). When life table analysis is used (Fig. 5), 85 percent of the patients are free of rectal cancer at 25 year follow-up after IRA.

Six of the seven patients developing rectal cancer in the rectal remnant presented with innumerable rectal polyps. This subgroup of patients with innumerable rectal polyps at presentation has a significantly increased risk of rectal cancer development as compared with patients presenting with fewer rectal polyps ($P < .003$, Fig. 6).

Thirteen patients had cancers in their colectomy specimens at the time of initial IRA. None of these patients has developed a cancer in their rectal remnant after IRA.

Two of the seven patients ultimately developing rectal cancer initially had spontaneous resolution of their rectal polyps. Five of the seven patients developing rectal cancer after IRA had occasional periods when they were free of macroscopic rectal adenomas and three patients had no benign polyps at the time of cancer diagnosis. In one of these patients, microscopic adenomatous changes were described by the pathologist.

The seven patients developing rectal cancer have been followed a mean 5.1 years after cancer diagnosis (range, 2 to 10 years). Only one of these patients has died from rectal cancer. Of the surviving patients all are at least four years following cancer excision with no recurrence.

Of the seven patients developing rectal cancer, three were not compliant with annual proctoscopic examination after their IRA and developed rectal cancers that were diagnosed at advanced stages (1 Dukes' B, 2 Dukes' C). In the four patients who were compliant with annual

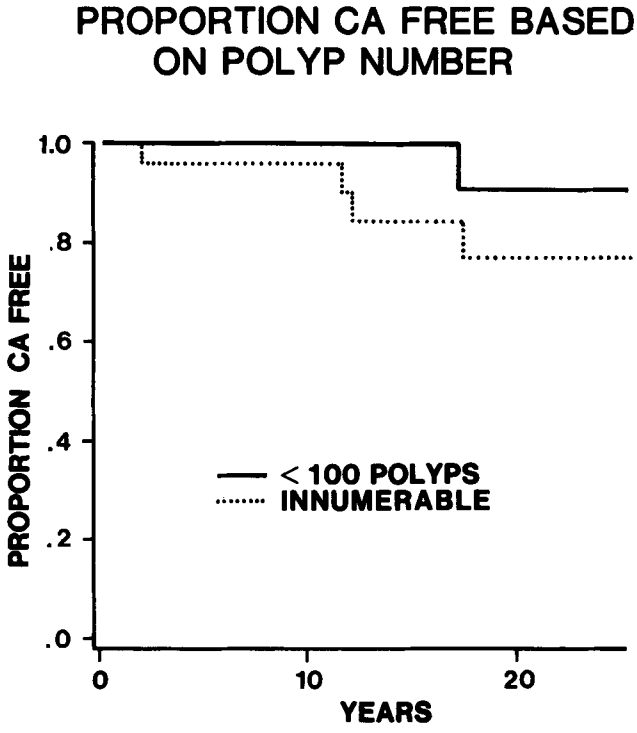

FIG. 6. Life table graph showing that patients with < 100 rectal polyps before IRA had a greater rectal cancer-free survival when compared with patients with innumerable rectal polyps ($P < .003$).

checkups after IRA, all rectal cancers were Dukes' stage A with apparent cure (mean follow-up, 5.6 years.).

One patient presented at an outside hospital one month following IRA with an incomplete small-bowel obstruction and diarrhea. The outside surgeon performed an abdominoperineal resection (APR) for these complaints at one month following initial IRA.

Three patients at a mean of 6.7 years following IRA underwent removal of the rectum (2 J-pouch, 1 end ileostomy) for uncontrollable polyp formation despite numerous electrocoagulations.

One patient underwent APR because of an anastomotic recurrence of a rectosigmoid cancer that had been present at the time of IRA two years previously.

In addition to the one death from rectal remnant cancer described above, there have been ten other deaths in the series of 116 patients having colectomy and IRA. Table 3 shows a breakdown of the causes of death in these 11 patients.

TABLE 3. *Patient Deaths*

From colon cancer present at time of initial colectomy	5
From cancer developing in rectal remnant	1
Pancreatic cancer	1
Adrenal cancer	1
Death from extensive desmoid tumor	1
Death following unrelated surgery	1
COPD	1
TOTAL	11

Discussion

Familial polyposis predictably leads to carcinoma of the colon if it is left untreated.[1] This knowledge has led to the development of registries to identify family members afflicted by this disease so that they may be treated before the development of malignant degeneration of the polyps.

At the present time there are three basic surgical approaches to this problem: 1) total proctocolectomy with permanent end ileostomy; 2) restorative proctocolectomy with ileal reservoir and ileoanal anastomosis; 3) abdominal colectomy and ileorectal anastomosis. Each procedure has its own benefits and drawbacks.

Total proctocolectomy eliminates the problem of malignant degeneration of colorectal polyps but involves a pelvic dissection with attendant risks of surgical morbidity and autonomic nerve dysfunction and necessitates a permanent intestinal stoma.

The restorative proctocolectomy with ileal reservoir and ileoanal anastomosis eliminates all large bowel polyps but carries a significant risk of pelvic sepsis and often has a less than optimal functional result. In a recent report[11] a large series of 188 patients (177 with ulcerative colitis), underwent mucosal proctectomy and ileoanal anastomosis with ileal pouch. In these patients there was an 11 percent incidence of pelvic sepsis and 14 percent of patients developed an anastomotic stricture. These same patients had a 2 percent rate of daytime incontinence and a 5 percent incidence of nocturnal incontinence. Similar rates of operative complications and suboptimal functional results have been reported from other major centers.[12,13]

At the Cleveland Clinic abdominal colectomy with ileorectal anastomosis at the 12 to 15 cm level has been the preferred surgical treatment for familial polyposis.[4-6] The current series confirms the low incidence of significant complications and quite acceptable functional results (median frequency of bowel motions, 4; range, one to 15 per day). The major disadvantage of abdominal colectomy and IRA is the absolute necessity to continue annual surveillance examinations of the remaining rectum. During a mean follow-up of 9.3 years (range, four months to 38 years), 12 patients underwent excision of the rectum because of rectal cancer (7), recurrent rectal cancer (1), uncontrollable polyposis (2) and poor functional result (1). This crude rate of 10 percent (12 of 116) of surgical excision of the rectum after IRA is corrected to 21 percent at 25-year follow-up by life-table analysis (Fig. 4).

The long-term risk of malignant degeneration is emphasized by the mean time to develop rectal cancer after IRA (14.6 years). In those patients who were compliant with annual proctoscopic examination, only four patients developed rectal cancer and all were curable Dukes' A lesions. Despite the development of seven rectal cancers, there has been only one death from rectal cancer. By contrast there have been 10 deaths from other causes in these patients (Table 3). This includes deaths from desmoids (1), adrenal cancer (1), and periampullary cancer (1). Familial polyposis coli is a generalized disorder and those patients in our series after IRA were at greater risk of death from extracolonic malignancy rather than rectal cancer developing in the rectal stump.

Of the seven patients who developed rectal cancer after IRA, six presented with innumerable rectal polyps prior to colectomy. This group of patients has a statistically significant increased risk of developing rectal cancer when compared with patients with fewer numbers of rectal polyps prior to surgery. All patients with FPC need careful follow-up after IRA, but this group, in particular, needs careful screening for the development of rectal cancer. In the present series there were 27 patients who had virtual carpeting of the rectum with polyps preoperatively, so that even in this unfavorable subgroup, the majority of patients can be managed with rectal preservation and IRA.

The absence of polyps in the rectal remnant over many years of follow-up does not eliminate the risk of rectal cancer. In patients developing rectal cancer after IRA, five of seven went for periods of years without any rectal polyps. In three of seven patients developing cancer, there were no macroscopic adenomatous polyps in the rectum at the time of proctectomy. In one of these patients there were microscopic adenomas and in the other two the remaining mucosa was normal.

Initial spontaneous resolution of rectal polyps does not preclude the later development of rectal cancer after IRA. Two of the seven patients developing rectal cancer after IRA, initially had spontaneous resolution of rectal polyps.

There have been previous studies[2,3] that have reported rates of 32 percent and 22 percent for development of rectal cancer after IRA. In one of these series[2] there were a number of patients who had an ileosigmoid rather than ileorectal anastomosis. In the other series[3] the authors coagulated only large polyps found at follow-up, rather than routinely destroying all polyps found on proctoscopic examination. These factors may account for the high rate of rectal cancer development described in these two series. Our results have been very similar to those reported from St. Mark's Hospital[1] where of 126 patients with ileorectal anastomoses, only three patients (2.3 percent) required removal of the rectum after a mean follow-up of 11.3 years. In the current series, 6 percent of the patients developed rectal cancer. This is adjusted to a 15 percent risk of rectal cancer development at 15 years when life table analysis is performed (Fig. 5).

Following abdominal colectomy and IRA, there is commonly a spontaneous decrease in the number of rectal polyps remaining in the rectal stump. In this study 64 percent (56 of 88) of patients showed definite evidence of decrease in the numbers of rectal polyps in the first year after IRA. There were 33 of 88 patients (38 percent) who showed complete spontaneous resolution of rectal polyps.

There have been several theories proposed to explain spontaneous regression of polyps after IRA. Initial suggestions[8] that the diminished blood supply to the rectum may play a part seem unlikely since there is enough circulation to the remnant rectum to support healing of the anastomosis. More likely would be theories that an abdominal colectomy and IRA change the fecal flora and influence the chemical environment of the rectum. Watne *et al.*[3] have described a patient who had an ileosigmoid anastomosis converted to an IRA at 14 cm. Postoperatively this man had a spontaneous decrease in the number of rectal polyps. Coincident with this, there was complete disappearance of coprostanol from his stool and the presence of only primary bile acids. Within two years coprostanol reappeared in his stool and only 35 percent of the bile acids were of the primary type; coincident with this reversion in the chemical composition of his feces, numerous rectal adenomas redeveloped. It is widely suggested that colorectal carcinoma is a neoplasm induced by an abnormal diet and resulting carcinogens in the stool. Further study of the change of fecal composition after IRA, correlated with rectal polyp growth, may provide some answers to the possible etiology and prevention of colorectal cancer.

There is generally no need to fulgurate rectal polyps before or concomitant with abdominal colectomy. As long as none of the rectal polyps appear malignant, the rectum should not be fulgurated at the time of initial IRA. Fully 64 percent of patients show some spontaneous regression of polyps and indeed 38 percent of patients will have complete regression of rectal polyps in the first postoperative year. Even in those patients having innumerable polyps at the time of presentation 17 of 27 (63 percent) had at least some degree of regression. At the six-month follow-up appointment, the patient should undergo proctoscopy, and complete fulguration of any remaining polyps is indicated. By waiting until six months, the need to fulgurate any polyps is eliminated in about one third of patients and, in a further third, there are far fewer polyps to coagulate as compared with initial presentation.

All patients, even those undergoing complete regres-

sion, should be followed rigorously for the rest of their lives with at least an annual proctoscopic examination. Of the 33 patients having complete polyp regression, 15 never redeveloped polyps (mean follow-up, 2.1 years) and 18 patients redeveloped polyps (mean follow-up, 6.8 years). This would suggest that the chance of redeveloping polyps increases as length of follow-up increases.

Conclusion

Abdominal colectomy with IRA is an effective surgical treatment for familial polyposis coli. Even in those patients who have innumerable rectal polyps preoperatively, spontaneous regression of rectal polyps is commonly seen. In 64 percent of patients there is at least partial regression (complete spontaneous regression in 38 percent). Because of this high rate of spontaneous regression, virtually all patients with FPC (with the exception of patients with rectal cancer at presentation) are candidates for IRA. Fulguration of polyps is not necessary preoperatively and patients should be rigorously followed at yearly intervals for coagulation of remaining rectal polyps. If rectal cancer or excessive polyp growth develops and is not easily treatable by coagulation, then the patient should be subjected to total proctocolectomy with either end ileostomy or restorative proctectomy with ileal reservoir and ileoanal anastomosis. Even if the patient is free of polyps for many years, there should be no change in the vigor of annual follow-up. Five of the patients developing rectal cancer had periods of several years when they formed no polyps, and three of these patients had no macroscopic polyps at the time of cancer diagnosis. Those patients who initially presented with innumerable rectal polyps appear to be at highest risk for later development of rectal cancer.

The documented spontaneous resolution of adenomatous polyps after IRA may serve as a useful research model for analysis of environmental and dietary factors that play a role in the promotion of adenoma formation and cancer development.

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