

Surgical and functional outcomes after colectomy and ileorectal anastomosis in patients with familial adenomatous polyposis

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Introduction

Familial adenomatous polyposis (FAP) is a distressing genetic disease for both symptomatic and asymptomatic carriers. Thus, early management is paramount to reduce risk of malignancy. Surgical management options for FAP become more variable; but each option comes with a cost. While restorative proctocolectomy with ileal pouch anal anastomosis is considered the gold standard, further surgeries surpass its advantages after critical selection of candidates. Total colectomy with ileorectal anastomosis (IRA) can be the future gold standard when candidates are wisely selected. This study evaluates the surgical, functional, and oncological outcomes of IRA.

Patients and methods

This study is a prospective cohort study conducted between June 2013 and June 2018 with a minimum follow-up of 12 months and included 33 patients with FAP. All patients underwent total colectomy followed by IRA. Then they were followed up for a mean period of 28.4 months to evaluate the postoperative surgical and functional outcomes as a primary endpoint with evaluation of long-term risk of rectal carcinoma as a secondary endpoint.

Results

Thirty-three patients had total colectomy followed by IRA. Ten patients developed early postoperative complications and seven had late complications. Bowel function was well preserved in 94% of patients and only 30% of the patients continued to use antidiarrheal medications beyond 6 months after the operation. One month postoperatively, the mean of bowel motions was 4.8 a day, which decreased to 2.6 times after 1 year of follow-up. Recurrence of polyposis was detected in 30% of patients, and only one patient had developed rectal malignancy.

Conclusions

With appropriate patient selection, IRA provides better surgical, functional, and oncological outcomes. Patient characteristics and disease features should be considered in surgical decision making.

Keywords:

familial adenomatous polyposis, familial polyposis, ileorectal anastomosis

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Introduction

Familial adenomatous polyposis (FAP) is one of the most common adenomatous syndromes all over the world and along with its variants like Gardner's syndrome, Turcot's syndrome, and attenuated form (AFAP) account for less than 1% of all colorectal carcinomas. It is an inherited autosomal dominant syndrome attributed to germline mutations of the tumor suppressor adenomatous polyposis coli gene located on chromosome 5q21-q22 [1,2].

There are two types of FAP: classic and attenuated (AFAP). The former type is mostly hereditary, only 30% of it arises from new mutation. The attenuated type is more benign in nature, and this attributed to the location of the mutation within the adenomatous polyposis coli gene that is responsible for the severity of colonic polyposis, measure of cancer risk, age of

malignant transformation, survival, and the presence of extracolonic manifestations [3–5].

The complexity of FAP originates from its predisposition to colorectal cancer (CRC), the third most common cancer worldwide [6]. Untreated cases with FAP have about 100% risk of developing CRC, expectedly by the age of 40 years [7].

In the era of radiological and surgical advances, early screening for CRC and management of high-risk groups is a major concern. Consequently, prophylactic surgeries are considered an important

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step in the management plan for FAP. Various surgical options are available including proctocolectomy with terminal ileostomy (PCI), total colectomy with ileorectal anastomosis (IRA), and restorative proctocolectomy with ileal pouch anal anastomosis (RPC-IPAA) with ileal pouch anal anastomosis [7].

Each procedure should only be considered on a case-by-case basis depending on patient and disease characteristics. These characteristics involve the patients' age, their compliance and tolerability to postoperative plans, and also the site, size, and number of polyps [8].

For instance, PCI is usually associated with patients' dissatisfaction with their self-image, which is attributed to the evolving sexual dysfunction, and the presence of the stoma. Therefore, it is reserved for patients with high risk of rectal cancer. Regarding RPC-IPAA, it was thought to be the gold standard surgery for FAP; however, progression of adenoma inside the ileal pouch and malignant transformation are two significant drawbacks whereas, IRA offers better outcomes especially for the bowel functions; albeit, it has been linked to higher risk of metachronous rectal cancer, which necessitates continuous follow-up [9].

In light of the above, this paper aims at assessment of surgical and functional outcomes of IRA as a primary endpoint with evaluation of long-term risk of CRC as a secondary endpoint.

Patients and methods

A prospective cohort study on 33 patients was conducted during the period from June 2013 to June 2018 at two settings; the surgical department at Suez Canal University Hospital in Egypt and the Surgical Department at Kuwait Cancer Control Center in Kuwait. After approval of Ethics Committee in both centers, and after informed consent was taken, patients who met the following criteria were included: classic or attenuated FAP diagnosed by means of colonoscopy and genetic testing and their colonoscopy showed rectal polyposis of less than 20 polyps. In addition, both symptomatic and asymptomatic (detected via screening) groups were included. Patients with heavy rectal involvement, high-grade dysplasia, and colonic or rectal cancer were excluded.

First, patients were exposed to complete history taking, physical examination, routine preoperative investigations, and assessment of fitness for

anesthesia. Second, a colonoscopy was performed to assess distribution and severity of colonic and rectal polyps. Moreover, multiple biopsies were obtained. After that, patients underwent total colectomy followed by IRA either end-to-end or side-to-end using hand sewing or stapling technique. The used surgical procedure was performed through an open abdominal exploration via midline incision with careful examination of abdominal and pelvic cavities.

Postoperatively, a pathological investigation was done of the resected colon for assessment of any underlying malignancy. All patients resumed enteral feeding as early as possible and discharged after a short period postoperatively. On discharge, they were started on antidiarrheal medications such as loperamide hydrochloride (Imodium; Johnson & Johnson Middle East, UAE).

All patients after discharge attended the follow-up proctoscopy for the first time after 3 months, then after another 3 months for the second time, then 6 months later, thereafter every year.

Primary outcome measures included incidence of postoperative complications, functional outcomes (patient satisfaction, stool frequency, and need for antidiarrheal medications beyond 6 months postoperatively), while occurrence of malignancy on follow-up was considered a secondary outcome.

Results

The study included 33 participants who had FAP. There were 20 men and 13 women whose mean age was 32.5 years. The majority of these participants were symptomatic (70%) on diagnosis. Most commonly reported feature was bleeding per rectum followed by altered bowel habits, and then mucous discharge. Only two patients had abdominal desmoid. Only 27% of the study participants had comorbid diseases including abdominal desmoid (9%), hypertension (9%), diabetes mellitus (6%), and lymphoma (3%) (Table 1).

Classic FAP was the predominant type estimated at 85%, and the rest showed 10–100 polyps (AFAP). Around half of the patients had 5 to less than 20 rectal polyps. Similarly, nearly half of the patients had rectal polyps of more than 5 mm in size (Table 2, Figs 1–4).

Concerning the surgical technique, side-to-end anastomosis was the most commonly used type

Table 1 Characteristics of patients

Characteristics	n (%)
Age	
Mean (SD)	32.5 (12)
Minimum	18
Maximum	64
Sex	
Male : female ratio	1.5 : 1
Male	20 (61)
Female	13 (39)
Presentation	
Screening	10 (30)
Symptomatic	23 (70)
Bleeding per rectum	11 (33)
Altered bowel habits	6 (19)
Mucous discharge	4 (12)
Abdominal desmoid	2 (6)
Comorbid diseases	
Abdominal desmoid	3 (9)
Hypertension	3 (9)
Diabetes	2 (6)
Lymphoma	1 (3)

Table 2 Distribution and severity of colonic adenoma and rectal polyposis

Variables	n (%)
Colonic involvement	
Classic FAP	28 (85)
AFAP	5 (15)
Number of rectal polyps	
0	3 (9)
<5	12 (36)
5<20	18 (55)
Size of rectal polyps	
<5 mm	11 (36)
≥5 mm	19 (63)

AFAP, attenuated familial adenomatous polyposis.

(58%) along with the stapling technique (61%). Hospital stay ranged between 4 and 16 days with a mean of 7.3 days. The majority of patients were followed up to 28 months (Table 3).

Only 10 patients had early postoperative complications, while seven patients had late complications. Early complications included wound infection which was the most common one followed by ileus, while the least common early complications were anastomotic leak and deep vein thrombosis. On the other hand, only seven patients experienced late complications such as bleeding per rectum and adhesive intestinal obstruction which was the most common (Table 4, Fig. 5).

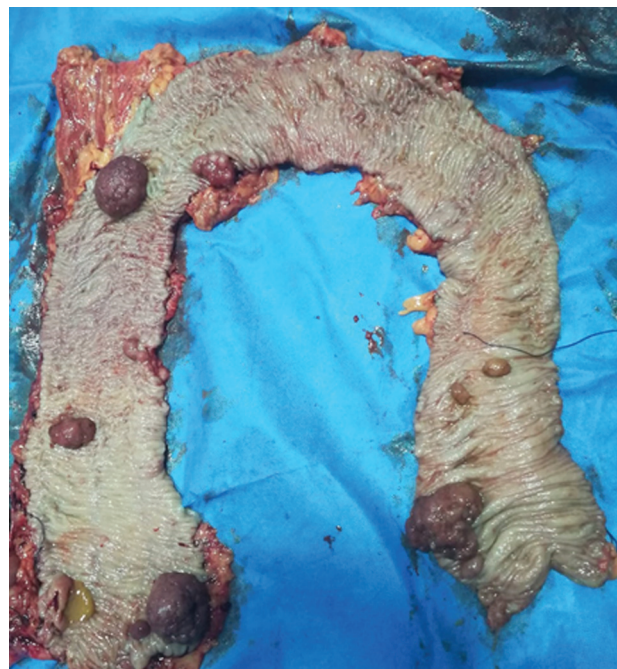
Regarding functional outcomes, about 94% of patients rated their satisfaction with the operation as fair to

Figure 1



Part of a resected colon showing polyposis.

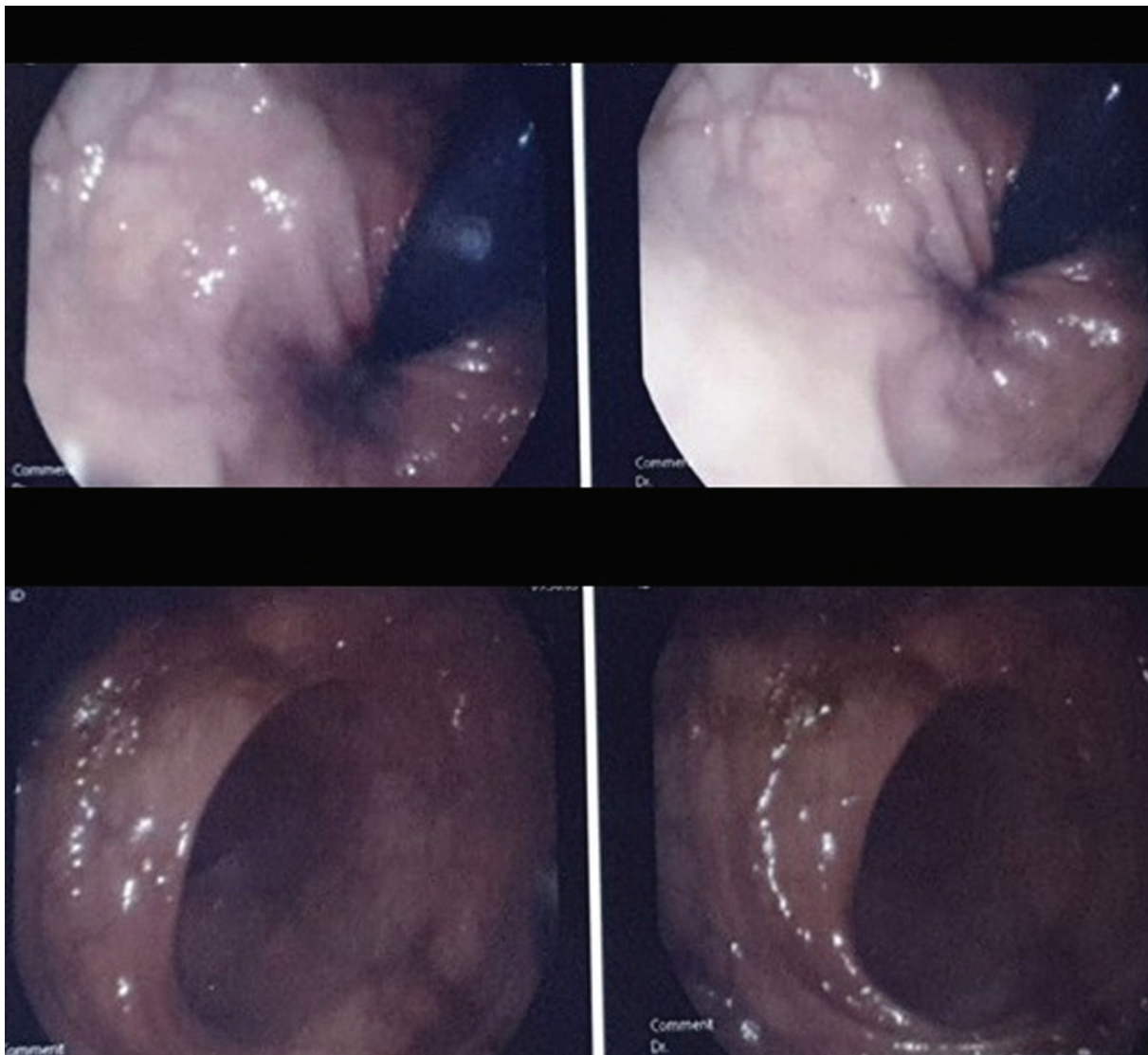
Figure 2



Part of a resected colon showing attenuated familial adenomatous polyposis.

excellent, with only two reporting poor outcome. Only 30% of patients needed antidiarrheal medications beyond 6 months postoperatively. The mean number of daily bowel motions was 4.8 times by the first month of follow-up and continued to decline reaching 2.6 times by the end of the year (Table 5).

Figure 3



View of the rectum using flexible sigmoidoscope at the follow-up session.

After IRA, about one-third of patients had secondary and tertiary recurrence of polyposis and needed excision or cauterization. There were three patients who had secondary recurrence by the sixth month, one of them had another event by the 12th month. At the 12th month, another three secondary relapses were detected; one of them had a tertiary event by the 18th month, and another one by the 24th month. At the 24th month, only one secondary event was detected (Table 6).

Discussion

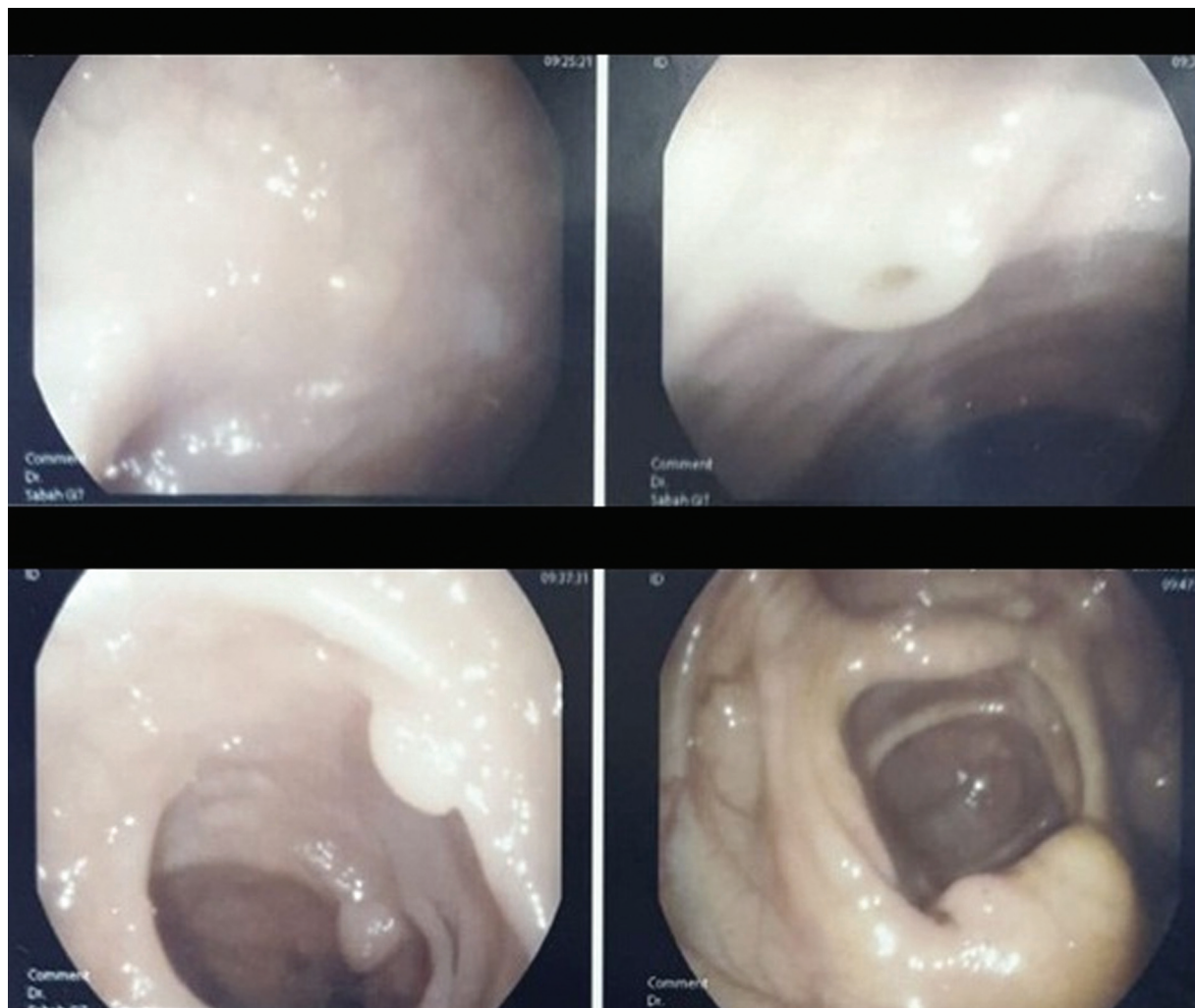
The study proved that IRA offers invaluable surgical, functional, and oncological outcomes that outweigh its disadvantages; moreover, it surpasses other modalities. The authors believe that wise selection of the included patients favored better outcomes found in this study.

Of these characteristics are young age, mild FAP, and compliance to follow-up.

IRA is known to be associated with fewer complications than other modalities (RPC-IPAA and PCI) [8]. On the basis of Ribeiro's findings, RPC-IPAA has a higher morbidity rate: 38.9% of early and 27.8% of late complications [10]. Undoubtedly, one of the advantages of IRA is avoidance of complications, which result from pelvic dissection, associated with RPC-IPAA and PCI such as with urinary and sexual dysfunction, male impotence, and low fertility among women [11].

According to Renkonen-Sinisalo and colleagues, Campos and colleagues, and Roberto and colleagues, the incidence of early complications of IRA reaches 20%. However, in this study, the incidence of early

Figure 4



View of the colon of a patient with attenuated familial adenomatous polyposis using a colonoscope.

Table 3 Operative and postoperative data

Variables	<i>n</i> (%)
Type of anastomosis	
End to end	14 (42)
Side to end	19 (58)
Technique of anastomosis	
Hand sewn	4 (12)
Stapling	20 (61)
Hospital stay (days)	
Mean (SD)	7.3 (3)
Minimum	4
Maximum	16
Follow-up period (months)	
Mean (SD)	28 (14)
Minimum	3
Maximum	48

complications is considered high in comparison to similar studies [12–14]. It is worth mentioning that most of these studies were performed laparoscopically. This confirms the observation of relatively higher incidence of complications associated with open

Table 4 Postoperative complications

Complications	<i>n</i> (%)
Early (first 30 days)	10 (30)
Wound infection	5 (15)
Ileus	3 (9)
Anastomotic leak	1 (3)
DVT	1 (3)
Late (>1 month)	7 (21)
Bleeding per rectum	2 (6)
Adhesive IO	2 (6)
Rectovaginal fistula	1 (3)
Recurrent abdominal wall desmoid	1 (3)
Rectal malignancy	1 (3)

DVT, deep vein thrombosis; IO, intestinal obstruction.

techniques [11], similar to our study. Our results agreed with Claude and colleagues, who reported ileus as the second common early complication after IRA, but with a lesser percentage (2%) [15].

Regarding late complications, previous studies depicted lower rates of late postoperative

Figure 5



Distal loopogram with gastrografin injection via temporary ileostomy showing contrast in the rectum and vagina indicating rectovaginal fistula.

Table 5 Measures of functional outcome

Variables	n (%)
Patient satisfaction	
Poor	2 (6)
Fair	21 (64)
Good	8 (24)
Excellent	2 (6)
Use of antidiarrheal drugs beyond 6 months postoperatively	
Yes	10 (30)
No	19 (58)
NA	4 (12)
Mean of stool frequency per day [mean (SD)]	
1 month	4.8 (0.9)
3 months	4.4 (0.6)
6 months	3.6 (0.6)
1 year	2.6 (0.5)

NA indicates a case with missed follow-up.

complications than ours, ranging between 2 and 12% [12,13].

Albeit, Claude and colleagues reported a much more higher rate of late complications (29.7%) [15]. Comparable to our results, previous studies confirmed that intestinal obstruction is the most common late postoperative complication [10,12,16].

Another important outcome to consider is the oncological safety. Only one (3%) case developed rectal cancer, which is low in comparison to other populations. In Kimura *et al.* [17], 50% of patients who underwent IRA had primary rectal cancer. In the latter example, a longer follow-up period (3–32 years)

Table 6 Recurrence of polyp excision or cauterly

Recurrence date	n (%)
No	22 (67)
NA	1 (3)
6th month	
Secondary	3 (9)
Tertiary	0
12th month	
Secondary	3 (9)
Tertiary	1 (3)
18th month	
Secondary	0
Tertiary	1 (3)
24th month	
Secondary	1 (3)
Tertiary	1 (3)

NA indicates a case with missed follow-up.

was an advantage for detection of more cases. Yet, with shorter follow-up periods (10 years) incidence of rectal cancer following IRA reaches 18.18% [12].

Noticeably, these high figures come from retrospective studies where there were no data about preoperative endoscopy for detection of cancer. Moreover, they built their assumptions based on pathological examination of resected lesions. Thus, preoperative endoscopy facilitates early detection of cancer lesions which, if found early, would have been treated differently with another modality saving the patient from another future surgery. This justifies the low cancer risk observed in this study.

While RPC is supposed to reduce cancer risk radically, still there is an 8–74% risk of adenoma progression within either the pouch or the transition zone [8]. Recently, Tajika *et al.* [18] found a positive correlation between the pouch age and maximum size of pouch adenoma, which proves that patients undergoing RPC will not be cancer immunized.

IRA has been well known for its better functional outcome than other modalities [8]. Unlike most studies, we used both subjective and objective measures to evaluate functional outcomes. Our results agree with of the study of Sun *et al.* [19] in which 95.8% of their patients rated satisfaction as fair to excellent. As per Schneider's conclusions, IRA received higher satisfactory rate than other modalities such as RPC-IPAA [20].

Interestingly, when the frequency of defecation was assessed during regular follow-up to 1 year, it markedly declined to a mean of 2.6 times/day. This outweighs the results of other studies where the mean number of

daily motions after IRA reached four [15]. In comparison to IPAA, IRA has less stool frequency. It was found that 70% of patients have increased stool frequency per day after IPAA [21].

Also, it is well established that the need for antidiarrheal drugs is lesser with IRA than with RPC-IPAA. In a Brazilian study by Ribeiro *et al.* [10] there was a need for antidiarrheal medications in 11% of cases, while in a comparative Iranian study, the need for antidiarrheal drugs was higher among the IRA group (50 vs 26.3% among the IPAA group) [21]. It is worth noting that many studies report the patients' need for antidiarrheal medications immediately after the operation and not after a period of follow-up. Furthermore, it is believed that such variability among different studies is due to various surgical approaches used within each study group, such as the type of anastomosis (end-to-end vs end-to-side), technique of anastomosis (hand sewn vs stapling), and the type of surgery (open vs laparoscopic).

Additionally, patient characteristics may affect the functional outcome too including age, chronic illnesses, associated gastrointestinal disease, and the type of FAP.

One of the most common inevitable postoperative complications is reappearance of polyps in all surgical modalities. That is why regular endoscopy is paramount. Kimura *et al.* [17] reported a recurrence rate comparable to ours after a long-term follow-up (25%). In contrast, a retrospective analysis of post-IRA outcomes by Campos *et al.* [13] reported a 72.2% polyp recurrence. It is believed that recurrence depends on multiple factors including the patients' age, follow-up period, rectal length, and type of genetic mutation [13]. Tajika *et al.* [18] found that polyp recurrence post-IPAA is more than with IRA. Furthermore, a larger size of polyps was prevalent among the former group.

Conclusions

Total colectomy with IRA for FAP provides good surgical, functional, and oncological outcomes. IRA is a less complex procedure than other modalities. Wise selection of candidates highly influences the success rate. Young age, mild FAP, and compliance to treatment constitute major determinants of success of IRA.

The study was characterized by multiple strengths points. First, critical selection of cases based on

patients' features (age and sex) and endoscopic findings (severity of the disease and risk of cancer). Second, performance of preoperative and periodic postoperative colonoscopy for early detection of cancers, and allocation of patients to a more appropriate modality.

Third, it is a prospective study where the study variables were controllable and measurable. Additionally, data collection and follow-up was feasible and accurate. Although the study provided promising results, undeniably, the sample size was rather small. Besides, there was no comparative group.

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Conflicts of interest

There are no conflicts of interest.

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