Laparoscopic Restorative Total Proctocolectomy With Ileal Pouch Anal Anastomosis for Familial Adenomatous Polyposis

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ABSTRACT

Background: Familial adenomatous polyposis is a hereditary disease characterized by the presence of thousands of colonic adenomas, which, if untreated, invariably undergo malignant transformation. Because this disease manifests at a young age, the laparoscopic approach to perform surgery would be desirable due to its cosmetic benefits. We describe our experience with this procedure and review the literature on the topic.

Methods: This is a case series of 15 patients who underwent restorative proctocolectomy with ileo-anal pouch anastomosis for familial adenomatous polyposis between 2000 and 2007. The salient operative steps are described.

Results: There were 9 males and 6 females, 32 to 52 years of age, with an average age of 44.8 years. The median body mass index was 21.5 (range, 17 to 28). Rectal cancer was already present in 4 patients at the time of diagnosis. The median operating time was 225 minutes. Mean blood loss was 60mL, with none of the patients requiring perioperative blood transfusion. None of the surgeries required conversion to the open approach. Bowel function resumed on the second postoperative day in 12 patients and on the third postoperative day in 3 patients. The median hospital stay was 8 days. Postoperatively, there was no mortality and no serious morbidity.

Conclusion: Laparoscopic restorative proctocolectomy with ileal pouch anal anastomosis is a feasible surgery for familial adenomatous polyposis, and considering its cosmetic benefit, is a desirable option for this group of predominantly young patients.

Key Words: Total proctocolectomy, Ileal pouch-anal anastomosis, Familial adenomatous polyposis.

INTRODUCTION

Familial adenomatous polyposis (FAP) is an autosomal dominant disease characterized by the presence of numerous adenomatous polyps in the colon. It affects between 1 in 8,300 to 1 in 14,025 live births, affecting both the sexes equally.[1] The polyps inevitably develop into cancer in 10 years to15 years after their appearance, and the only treatment available at present to prevent the cancer is total proctocolectomy.[2,3] The characteristic of this disease is the young age at presentation with the average age at diagnosis of colorectal cancer being 39 years.[4] In patients diagnosed with FAP, prophylactic colectomy is recommended by the late teens.[5] The young age of the patient makes cosmesis a significant consideration when choosing the mode of therapy. Hence, laparoscopic surgery is a desirable choice for these patients.

The aim of this study was to demonstrate the feasibility of laparoscopic restorative proctocolectomy with ileal pouch anal anastomosis (RPC-IPAA) for FAP and to verify its outcome.

METHODS

Between 2000 and 2007, 15 patients diagnosed with FAP underwent RPC-IPAA. A single operating surgeon (CP) assisted by a surgical team performed all the surgeries. Before embarking on this study, we had developed sufficient experience in laparoscopic colorectal surgery, which has been published elsewhere.[6] Preoperative bowel preparation consists of cleansing with polyethylene glycol solution. All patients are administered intravenous antibiotics in the form of ciprofloxacin 500mg IV, ornidazole 500mg IV one hour before surgery, and antithrombotic prophylaxis is administered with low-molecular-weight heparin and elastic stockings. A nasogastric tube and a urinary catheter are inserted before the start of the procedure.

Basically, the surgery consists of a combination of various phases of mobilization and ligation of pedicles involved in the performance of left, right, and transverse colectomy. The patient is placed in a semilithotomy position with arms outstretched. The thighs are flexed minimally so as to not to impede the movement of laparoscopic instru-

ments. The ports are placed as depicted in **Figure 1**. The importance of a remote-controlled operating table that can be tilted on any axis cannot be overemphasized. It is gravity that is most often used as a retractor to keep the bowels away from the operating area. The procedure starts with mobilization of the sigmoid and rectosigmoid, which is continued cranially towards the descending colon. This phase is done with the surgeon on the right side of the patient. The surgeon moves between the legs of the patients for mobilization of the splenic flexure and transverse colon. The ascending colon and hepatic flexure are mobilized from the left side of the patient, and the distal ileum is transected using laparoscopic staplers. The surgeon returns to the right side for rectal mobilization and ultra-low resection using staplers. During all the phases of the surgery, the dissection has been medial to lateral. We follow the planes of oncological clearance as these planes are avascular, and secondly, to provide adequate clearance even if the final histopathological examination of the
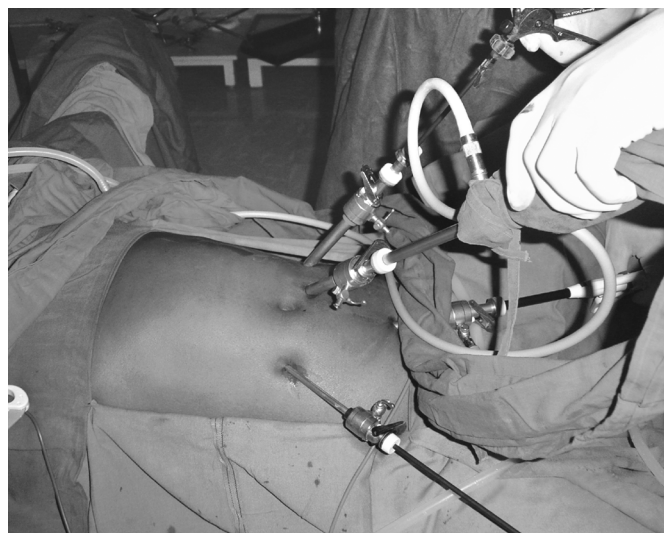

Figure 1. Port positions. **A.** 10 mm – Camera port for mobilization of rectum/sigmoid/hepatic flexure and right half of transverse colon; Right-hand working port for mobilization of descending colon, caecum, and ascending colon; Left-hand working port for mobilization of splenic flexure and left half of the transverse colon. **B.** 10 mm or 5 mm – Right-hand working port for mobilization of transverse colon; Retraction for mobilization of rectum, sigmoid colon, and descending colon. **C.** 10 mm – Camera port for mobilization of descending colon, splenic flexure, left half of transverse colon, caecum, and ascending colon. **D.** 5 mm – Left-hand working port for mobilization of rectum, sigmoid colon, descending colon, caecum, ascending colon, hepatic flexure, and right half of transverse colon; Retraction for mobilization of splenic flexure and left half of transverse colon.

specimen reveals malignancy that was not detected by preoperative sampling of the polyps. The inferior mesenteric pedicle is flush ligated during the rectosigmoid dissection. Thereafter, we deal with the left colic and middle colic pedicles in a similar fashion during the phases of dissection of the left and transverse colon, respectively. During the phase of transverse colon mobilization, the greater omentum is released from the transverse colon and placed in the left upper quadrant. Flush ligating the pedicles and use of Ultracision for dissection enables a relatively bloodless field and minimal perioperative blood loss. Through a minilaparotomy Pfannenstiel incision, the specimen is extracted after adequate measures are taken to protect the wound. The stapled end of the terminal ileum is delivered into the wound, and a J pouch is fashioned by using the linear stapler. The length of each limb of the J-pouch is 15cm. An enterotomy is made at the apex of the pouch, and the blades of the linear stapler (7.5cm) are introduced into each of the limbs of the pouch and sequentially fired twice to obtain the pouch. The anvil of the circular stapler is inserted into the apex of the pouch through the enterotomy and secured with purse-string sutures. The pouch is delivered into the abdomen, the Pfannenstiel incision is closed, and pneumoperitoneum reestablished. After confirming the correct orientation of the pouch, it is anastomosed to the anal canal via a transanally introduced circular stapler **(Figure 2)**. The magnification afforded by the laparoscope allows ultra-low dissection so that the rectal stump is divided at the dentate line itself. In case a proximal division occurs, rectal mucosectomy is carried out transanally prior to
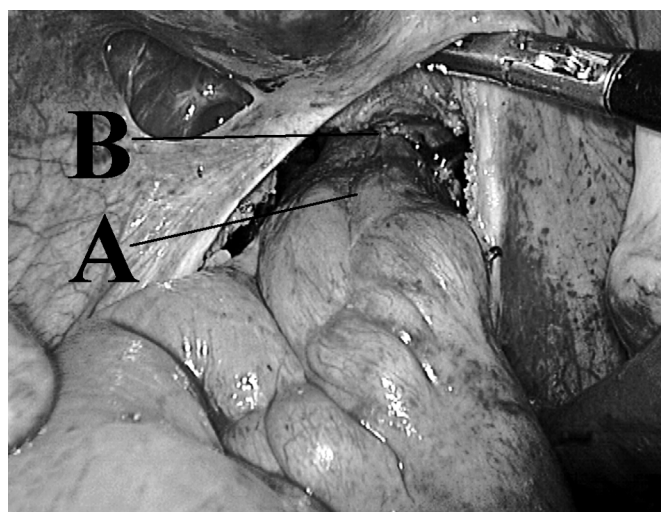

Figure 2. Ileal J-pouch anal anastomosis: **A.** Ileal J-pouch. **B.** Staple line. **C.** Pelvic diaphragm. **D**. Anal canal.

pouch anal anastomosis. We perform a protective ileostomy in all patients, which is closed after 6 weeks. We prefer a Brooke's type of everting ileostomy placed in the right iliac fossa.

RESULTS

Fifteen FAP patients underwent restorative total proctocolectomy with ileal pouch anal anastomosis (RPC-IPAA). The patients consisted of 9 males and 6 females. The average age was 44.8 years (range, 32 to 52). BMI ranged from 17 to 28, with the median being 21.5. All the patients had a family history of multiple colonic polyposis. None of the patients had extracolonic manifestations. Four of the patients had rectal malignancy. In these 4 patients, preoperative colonoscopy and biopsy had revealed adenocarcinoma in rectal polyps. The American Joint Committee on Cancer (AJCC) staging of 2 patients was T2N0M0, and the other 2 patients had T3N0M0 stage malignancy. The tumors were located 8 cm, 12 cm, 13 cm, and 16 cm from the anal verge in these patients.

The operating time ranged for 190 minutes to 365 minutes with a median of 225 minutes. The mean blood loss was 60 mL with a range of 30 mL to 180 mL. None of the patients were given a perioperative blood transfusion. No conversions were necessary.

The average incision length of the mini-Pfannenstiel incision was 5 cm (range, 4 to 6.5). Twelve patients resumed a liquid diet on the second postoperative day, while 3 patients did so on the third postoperative day. The malignancy and staging was confirmed in the 4 patients with proven rectal adenocarcinoma. In one patient with preoperative staging of T3N0M0, the final staging was pT3N1M0. In all the patients, the margins were free from tumors, and the minimum distal margin was 4cm.

The hospital stay ranged from 6 days to 12 days (median, 8). Stoma closure was undertaken in all patients after 6 weeks. All patients were continent with stools and flatus. The pouch frequency varied between 3 and 6, with the median being 4 times. Though initially, patients complained of liquid stools, by one month postoperatively, they were passing semisolid stools and did not suffer from urgency.

No intraoperative surgical complications or deaths occurred. No postoperative mortalities occurred. One patient developed wound infection that was treated by a change in antibiotic, according to the culture sensitivity report. One patient had postoperative urinary retention, which was managed conservatively with catheterization.

This patient was an elderly male of 52 years and had an enlarged prostate (42g) on ultrasonography preoperatively. He was started on Tablet Tamsulosin 0.4mg once a day at night, and the catheter was removed on the seventh day. He did not have any problems thereafter. One patient had subacute obstruction after 6 months, which was treated conservatively. She was kept nil orally with Ryle's tube decompression of the stomach and repeated enemas to clear the pouch. A large amount of inspissated feces were evacuated from the pouch. The patient recovered and was started on oral feedings by the fifth day of treatment and was discharged on the seventh day. The patient was taught self-administration of enemas and pouch irrigation. With this, the patient has not experienced any problems after a mean follow-up of 3 years and 10 months. None of the male patients had problems with potency, orgasm sensation, ejaculation, or micturition. In response to a questionnaire, all the patients declared themselves to be highly satisfied with the outcome of the surgery and the cosmesis and would recommend the procedure to other patients. All the patients were called for follow-up after 7 days, 1 month, 6 months, and yearly thereafter, except for the patients with malignancy who were called monthly after the first month and up to 1 year and then yearly thereafter. At every visit, the patient is examined. At the 3-month visit, endoscopic pouch examination with a flexible sigmoidoscope and upper GI endoscopy is performed, which is repeated after 6 months and yearly thereafter. In case patients miss their scheduled visit, they are reminded via telephone. All patients have maintained follow-up, and the average follow-up has been 3 years and 10 months. There has been no recurrence of malignancy or polyps in any of the patients. Moreover, the upper GI endoscopy has revealed the absence of polyps in the duodenum. The patient with T3N1MO malignancy was referred to an oncologist and received adjuvant chemotherapy consisting of irinotecan, 5-FU, and folinic acid. He has tolerated the chemotherapy well and is disease-free after 2 years and 4 months of follow-up. Though all the patients' first-degree relatives have been counseled to undergo screening for familial adenomatous polyposis, only 13 such relatives belonging to 5 patients have come for a colonoscopy. Of these, FAP was diagnosed in 3 patients, and they are awaiting surgery.

DISCUSSION

Familial adenomatous polyposis (FAP) is a rare autosomal dominant disease caused by a defect in the apc gene of the 5q21 chromosome. All affected untreated patients will

die of colorectal adenocarcinomas in the fourth or fifth decades, which was the fate of most patients in the first half of the 20th century.[2] However, thereafter, several national registries were established, and screening programs were started.[7–9]

Early detection and treatment of this disease has resulted in a reduced incidence of death from colorectal cancer. It has been demonstrated that establishment of a national registry improves the colectomy rate, reduces the prevalence of colorectal cancer, and improves survival.[9] Today, the most frequent causes of death in screening detected patients are duodenal cancer and desmoid tumor.[10,11] Similarly, in a study based on the Hong Kong registry, the median age of diagnosis was significantly lower in patients detected by screening than those diagnosed by symptomatic presentation (29 years vs. 34 years, respectively). At the time of diagnosis, 9.7% of the screened patients had malignancy compared with 61% of the unscreened patients.[12]

However, India does not have a national registry for FAP, nor does it have a screening program. As a result, most of our patients presented late (mean age being 44.4 years), and 4 of the 9 patients had already developed malignancy. Of the 9 patients, 5 were diagnosed with FAP while being investigated by us for abdominal symptoms, including the 4 patients with malignancy. The remaining patients had siblings or parents suffering from the disease and were referred to us for management of their disease. Thus, establishment of national or regional registries is the need of the hour to help early detection and treatment of these patients.

Though primary chemoprevention of FAP has been attempted with sulindac and cyclo-oxygenase inhibitors, it has not proven completely successful.[13,14] Currently, surgery is the only effective therapy to prevent progression to colorectal cancer. The surgical options available include total proctocolectomy with ileostomy (TPC) and restorative proctocolectomy with ileal pouch anal anastomosis (RPC-IPAA), which ensures elimination of all rectal and colonic mucosa. However, TPC mandates a permanent ileostomy, which is not a desirable option in these predominantly young patients. Thus, RPC-IPAA is the procedure of choice. The laparoscopic approach would seem a logical choice in these patients due to the improved cosmetic benefit. However, laparoscopic total colectomy is a most complex and demanding procedure in laparoscopic colorectal surgery, requiring significant experience in advanced laparoscopic surgery. Surgeons have to operate in all the quadrants of the abdomen and should have a thorough knowledge of anatomy of these regions before performing this procedure. Though the initial reports indicated that laparoscopic colectomy took a significantly prolonged time to accomplish compared with open colectomy,[15,16] with increasing experience, this difference has been reduced.[17] Various studies have shown that laparoscopic surgery is associated with decreased analgesic requirement, earlier return of bowel function, reduced length of hospital stay, earlier return to routine work and improved cosmesis compared with open surgery.[18–20]

Restorative proctocolectomy with ileal pouch anal anastomosis (RPC – IPAA) was first described in 1978.[21] Since then, this procedure has become the procedure of choice in the surgical management of patients with ulcerative colitis (UC) and familial adenomatous polyposis (FAP). The laparoscopic approach was used for this procedure for the first time in 1992.[22] Laparoscopic pouch formation was described in the same year.[23] This is the preferred approach, especially in this subset of patients who are usually young and hence appreciate the cosmetic advantage and have a better body image.[24] Experience gained in performing partial and segmental colectomies helps in mastering the techniques of total colectomy.[25] It is difficult to judge the impact of laparoscopic surgery in treatment of FAP compared with conventional techniques due to a lack of adequate data in the literature. A few controlled studies have compared laparoscopic RPC-IPAA with open procedures for patients with ulcerative colitis.[24,,26–28] Some of these series have also included patients with FAP. Most of the series have shown prolonged operative times with the laparoscopic approach compared with the open approach, except for a series by Araki et al.[24,26–28]

The latter showed similar operative times in both the arms probably because only the colonic mobilization was being done laparoscopically, while the vessel transection and rectal mobilization was being done by a mini-laparotomy incision.

However, with the use of advanced instrumentation like the LigaSure (Valleylab, Bolder, CO, USA) for vessel transection and Harmonic scalpel (Ethicon Endo-Surgery, Cincinnati, OH, USA) for dissection and tissue transection, the speed of surgery increases, which is borne out by the fact that our median operating time of 225 minutes was comparable to open resection times reported in the former series. The return of bowel function is also early. While the earlier study by Schmitt et al[26] reported a higher morbidity of 68% by the laparoscopic approach compared with 35% by the open approach, all of the later series showed lower morbidity by the laparoscopic approach. In

our series, we did not encounter any significant complication except for one patient who presented with subacute intestinal obstruction 6 months after the surgery and was managed conservatively. Length of hospitalization in our series averaged 8 days, which is similar to that reported by others and slightly less than for the open counterparts. However, this is not a true reflection of the need for hospitalization of these patients because our patients preferred to remain in the hospital until sutures were removed from the mini-Pfannenstiel incision, and the final histopathological examination report of the specimen was ready. Due to these peculiar geographical and socioeconomic factors, in spite of most (80%) of our patients having started an oral diet by the second postoperative day and all of them being on oral diet by the third day, the length of hospitalization was inordinately long.

In fact, cosmetic benefit alone has been shown to be of paramount importance to patients, especially as the young age group of patients is more concerned about body image.[29] Denker et al[24] have shown that satisfaction with the cosmetic result of the scar was significantly higher with the laparoscopic RPC-IPAA compared with the open group. We had a similar experience as the cosmetic benefit of a small Pfannenstiel incision (average length of 5cm) was appreciated by all our patients in the postoperative surveillance questionnaire, and all of them would recommend this procedure to other patients.

All the patients with malignancy underwent total mesorectal excision and after a mean follow-up of 2.1 years are free from recurrence. One of the controversies surrounding RPC-IPAA has been the development of pouch adenomas, the incidence of which has been reported to be between 8% and 60% on long-term follow-up.[30–33]

One of the risk factors for the development of pouch polyps is the retained rectal mucosa as a result of the stapled anastomosis. Van Duijvendijk et al[33] found that the cumulative risk of developing polyps after RPC and IPAA at the anastomotic site was 31% for the stapled group and 10% for the sutured group. In contrast, Polese et al[34] did not find any increase in risk of pouch polyp formation after stapled anastomosis. Though our follow-up period has not been long (range, 1 year to 8 years, median 3.4), we have not encountered pouch polyposis in any of our patients. However, we suggest a regular follow-up regime for our patients for routine screening, which consists of yearly endoscopy of the pouch with biopsy of any suspicious lesions. Finally, the contentious issue is the cost factor. Though no studies to date have objectively dealt with this issue, we feel that the initial higher instrument cost for laparoscopic surgery can be offset by having a dedicated laparoscopic surgical unit, use of reusable instruments, faster operative times due to availability of advanced instrumentation, and the shorter hospitalization. Moreover, the cosmetic benefit and patient satisfaction afforded by the laparoscopic approach have also to be factored in. Based on this, we are of the opinion that laparoscopic RPC-IPAA should be the approach of choice in the predominantly young patients afflicted by FAP.

CONCLUSION

We have shown that laparoscopic total proctocolectomy with ileal pouch anal anastomosis is a feasible surgery with minimal morbidity. While the laparoscopic approach offers definite cosmetic benefit as well as early return of bowel function, its real worth can only be established by comparative studies with the open approach, which, given the rarity of the condition, may be a difficult proposition. We feel that in the absence of the peculiar geographical and socioeconomic constraints that we face in our country, the length of hospitalization can also be reduced in these patients, further adding to its benefits. Meanwhile, the final verdict on laparoscopic RPC-IPAA is still awaited.

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