



Ileal pouch anal anastomosis in pediatric familial adenomatous polyposis: A 24-year review of operative technique and patient outcomes



Raelene D. Kennedy ^{a,*}, Abdalla E. Zarroug ^a, Christopher R. Moir ^a, Shennen A. Mao ^a, Mounif El-Youssef ^b, D. Dean Potter ^c

^a Division of Pediatric Surgery, Department of Surgery, Mayo Clinic, Rochester, MN

^b Division of Gastroenterology and Hepatology, Department of Pediatrics, Mayo Clinic, Rochester, MN

^c Division of Pediatric Surgery, Department of Surgery, University of Iowa, Iowa City, IA

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ABSTRACT

Background: Total proctocolectomy with ileal pouch anal anastomosis (IPAA) is the operative procedure of choice for familial adenomatous polyposis (FAP) patients. We review 24 years of operative experience and outcomes in pediatric patients with FAP.

Methods: Patients with FAP, age < 20 years, presenting to a single institution between 1987 and 2011 were included. Operative technique and outcomes were reviewed retrospectively. Primary outcomes included postoperative complications (30 days), long-term bowel function, and polyp recurrence at the anal anastomosis.

Results: 95 patients with FAP underwent IPAA. Mean age at IPAA was 15.5 years with a mean follow-up of 7.6 years. 29 patients underwent 1-stage IPAA, 65 patients had a two-stage IPAA, and 1 patient underwent a 3-stage procedure. 67 patients had an open procedure, 25 underwent a laparoscopic approach, and more recently 3 patients underwent single incision laparoscopic IPAA. Patients with 1-stage IPAA demonstrate better long term bowel control vs. 2-stage IPAA patients (10.7% vs. 36.0% occasional incontinence, $p = 0.018$). However, 1-stage IPAA patients suffered increased short-term complications, such as anastomotic leak (17.2% vs. 0%, $p = 0.002$) and reoperation (20.7% vs. 4.6%, $p = 0.02$) compared to 2-stage IPAA. Anal anastomosis polyp recurrence occurred in 22.7% of 1-stage patients and 10.0% of 2-stage patients. Short-term complications, polyp recurrence, or long-term continence were equivalent between open and laparoscopic cases.

Conclusion: Single-stage IPAA in children with FAP is associated with better bowel control but increased anastomotic leak, reoperative rate, and polyp recurrence. In experienced hands, laparoscopic IPAA is equivocal to open IPAA.

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Familial adenomatous polyposis (FAP) is an autosomal dominant syndrome characterized by mutation in the Adenomatous Polyposis Coli (APC) gene on chromosome 5 [1]. FAP leads to formation of numerous adenomatous polyps in the colorectum, beginning in the first and second decades of life. Progression to colorectal cancer (CRC) occurs in almost 100% of cases by 40 to 50 years of age [2]. Approximately 80% of patients have a documented family history of FAP, while 20% appear to be de novo mutations [1,2].

Prevention of colorectal cancer in FAP patients has long been managed by colectomy. Total proctocolectomy with ileal pouch anal anastomosis (IPAA) is a restorative procedure which effectively minimizes the risk of CRC by maximal removal of at-risk mucosa

while maintaining bowel continence [3]. Abdominal colectomy with ileorectal anastomosis has been used for FAP patients with rectal sparing, however 30% of these patients will develop rectal cancer by age 60 years [4]. Therefore, our preferred approach has been and remains IPAA.

IPAA has been widely used since the early 1980s and can be performed with many technical variations. As with any surgical procedure over the past two decades, IPAA is now commonly performed laparoscopically in addition to the traditional open technique. It has also recently been described with a single site laparoscopic technique [5]. Furthermore, the procedure has been performed with and without diverting ileostomy [6]. Finally, the anal anastomosis may be performed with a circular stapler or involve a rectal mucosectomy with hand sewn anastomosis.

The purpose of this study was to review our experience with ileal pouch anal anastomosis in pediatric patients with familial

* Corresponding author at: Department of General Surgery, Mayo Clinic Rochester, 200 First Street SW, Rochester, MN 55905. Tel.: +1 507 538 1330; fax: +1 507 284 5196.

E-mail address: kennedy.raelene@mayo.edu (R.D. Kennedy).

adenomatous polyposis at a single institution, with emphasis on technical surgical variations and patient outcomes.

1. Materials and methods

Our institution is a tertiary care center which serves the local population of southeast Minnesota and a large referral population. This study was approved by the institutional review board. All patients with a diagnosis of FAP who underwent total proctocolectomy with ileal pouch anal anastomosis from 1987 to 2011 and were ≤ 20 years of age at the time of surgery were included. Patient medical records were retrospectively reviewed for diagnosis, operative technique, postoperative outcomes and complications.

Diagnosis of FAP was confirmed by either a positive genetic test in recent patients, or clinical presentation with pathologic confirmation 9 (colonoscopy with biopsy) in older cases. Pathology reports were reviewed to confirm presence of adenomatous polyps. Final surgical pathology reports were reviewed to confirm extent of polyposis, degree of dysplasia and presence/absence of invasive cancer.

Surgical reports were reviewed for operative technique. Variables recorded included use of laparoscopy, number of surgical stages employed (1-, 2-, and 3-stage procedures), and anal anastomosis technique (stapled vs. mucosectomy). One-stage procedures included total proctocolectomy with creation of IPAA with no diverting stoma. Two-stage procedures included total proctocolectomy with ileal pouch anal anastomosis and diverting ileostomy, where the diverting ileostomy was closed during the second-stage procedure. Three-stage procedures included total abdominal colectomy with end ileostomy, followed by a second stage with completion proctectomy, IPAA creation and diverting loop ileostomy, and then a final third stage with closure of the diverting ileostomy. Operative technique was determined by surgeon preference and local conditions at the time of surgery.

All postoperative complications were recorded with particular attention to anastomotic leak, abscess, wound infection, small bowel obstruction, and pouchitis. Anastomotic leak and bowel obstruction were confirmed with imaging (CT and/or X-ray). Wound infection and pouchitis diagnoses were made clinically. Short term complications were those occurring within 30 days after the final stage of the IPAA procedure, whereas long term complications were those encountered after 30 days. Reoperation for these complications was also tracked.

Bowel function was recorded as noted in last clinical follow-up documentation from the medical record. Polyp recurrence was monitored with annual or biannual pouchoscopy. Polyp recurrence was recorded as documented in follow-up pouchoscopy reports.

Statistical analysis was performed using T-tests for continuous variables. Chi square analysis and Fisher exact tests were employed for comparison of categorical variables. Logistic regression analysis was also performed where appropriate. Statistical significance was defined as $p \leq 0.05$ for all of the above.

2. Results

Ninety-five patients ≤ 20 years with FAP underwent IPAA at our institution between 1987 and 2011. Average age at FAP diagnosis was 13.9 years (range 4–20 years). Mean age at surgery was 15.4 years (range 4–20 years) (Fig. 1). Mean follow-up was 7.6 years (range 0–24 years). Gender was equally distributed with 43 males (45%) and 52 females (55%). The indication for surgery was primary cancer prevention in 92 patients and treatment of active bleeding in the remaining 3 patients. All patients had greater than 50 colorectal polyps demonstrated by endoscopy preoperatively.

Median and mean postoperative length of stay was 9 days (range 3 to 48 days). Twenty-one short-term (30-day) postoperative complications occurred in 19 of 95 patients, where 9 (9.4%) patients required reoperation (Table 1). Fifty-two long term complications were documented in 35 of the 95 patients, with reoperative rate of 19.8% for these complications (19 patients) (Table 2).

Overall functional outcomes were positive, with 58 (72.5%) patients reporting no problems with incontinence. Nighttime soiling was occasional in 8 patients (10%) and occasional daytime leakage was reported by 13 (16%) patients. No patients reported complete incontinence. Fifteen patients had no documentation of bowel continence.

Ninety-four (98.9%) patients had a functional pouch at last follow-up, and only one has required pouch excision (secondary to early obstructive complications). Nine (9.4%) patients had documented recurrence of adenomatous polyps at the anal anastomosis, all of which were successfully managed with local transanal or endoscopic resection, and no patients had adenocarcinoma in these specimens.

Twenty-nine patients underwent 1-stage IPAA, 65 patients underwent 2-stage IPAA, and one patient had a 3-stage procedure.

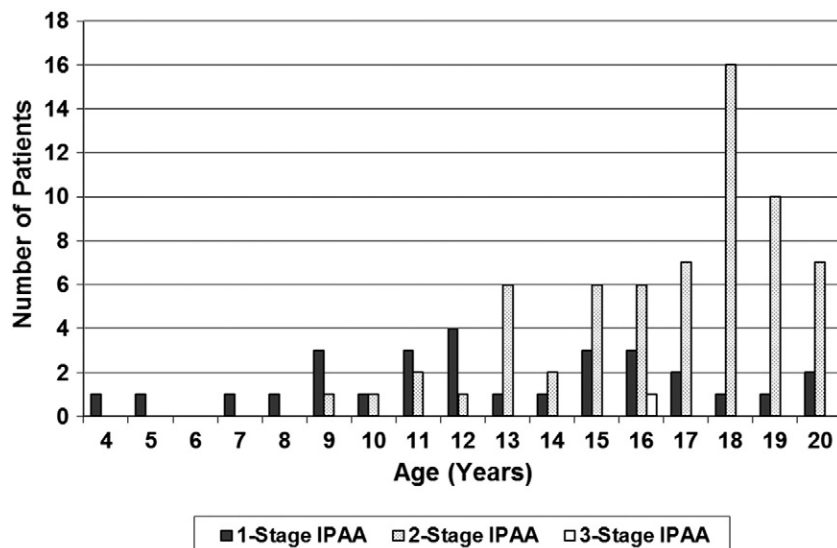


Fig. 1. Age Distribution by IPAA Stages.

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