Functional Outcomes and Complications after Restorative Proctocolectomy and Ileal Pouch Anal Anastomosis in the Pediatric Population

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BACKGROUND: Data regarding the long-term outcomes of restorative proctocolectomy and ileal pouch anal anastomosis including pouch function and quality of life in the pediatric population are limited in pediatric patients.

STUDY DESIGN: Indications for surgery, complications, long-term function, and quality of life were evaluated in pediatric patients undergoing ileal pouch anal anastomosis. Assessment of quality of life was performed using the Cleveland Global Quality of Life score.

RESULTS: There were 433 patients with a mean age of 18.04 ± 2.9 years. Final pathologic diagnoses were ulcerative colitis or indeterminate colitis (78.3%), familial adenomatous polyposis (15.7%), Crohn's disease (5.1%), and others (0.9%). There were 237 patients (54.7%) who underwent total proctocolectomy and ileal pouch anal anastomosis; 196 (45.3%) underwent initial subtotal colectomy followed by completion proctectomy with ileal pouch anal anastomosis. Anastomosis was stapled in 352 patients (81.3%) and hand-sewn in 81 (18.7%) patients. Mean follow-up was 108.5 ± 78.4 months. At the most recent follow-up, mean Cleveland Global Quality of Life score was 0.8 ± 0.2 and numbers of daytime and night-time bowel movements were 5.3 ± 3.1 and 1.6 ± 1.3, respectively. The majority of the patients (86.8%) were fully continent or only complained of rare incontinence. Most patients had no seepage (day, 84.3%; night, 72.4%) and did not wear any pads (day, 89.3%; night, 84.3%). Most denied dietary (71.3%), social (84.8%), work (85.7%), or sexual restrictions (87.6%) at the time of last follow-up. There were 92.7% of patients who said they would undergo ileal pouch anal anastomosis again and 95.2% would recommend surgery to others.

CONCLUSIONS: Restorative proctocolectomy with ileal pouch anal anastomosis can be performed in pediatric patients with acceptable morbidity and is associated with good long-term results in terms of gastrointestinal function, quality of life, and patient satisfaction. (J Am Coll Surg 2014;218: 328–335. © 2014 by the American College of Surgeons)

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Ileal pouch–anal anastomosis (IPAA) has become the standard restorative surgical treatment for patients with ulcerative colitis (UC) and familial adenomatous polyposis (FAP), after removal of the colon and rectum.[1]

Ulcerative colitis is an inflammatory process limited to the colorectal mucosa. It is characterized by alterations in bowel function and symptoms of intestinal inflammation. Although UC is typically seen in adults with a bimodal age distribution of greatest frequency in the third and eighth decades,[2] more than 20% of patients diagnosed with UC present during childhood.[3] Children may have a more aggressive disease course than adults because total colonic involvement is more common in children. Indications for surgery include medical intractability, severe bleeding, cancer risk, obstruction, perforation, and toxic

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Abbreviations and Acronyms

CD = Crohn's disease
CGQL = Cleveland Global Quality of Life
FAP = familial adenomatous polyposis
IPAA = ileal pouch anal anastomosis
QOL = quality of life
RP = restorative proctocolectomy
UC = ulcerative colitis

megacolon.[4] Familial adenomatous polyposis is an inherited, autosomal-dominant disease caused by a germline mutation of the *adenomatous polyposis coli (APC)* gene.[5] If left untreated, the condition inevitably leads to colorectal cancer.[6] Total removal of the colorectal mucosa is indicated to prevent life-long colorectal cancer development.[7] Although UC and FAP are the most common indications for a restorative proctocolectomy (RP), indeterminate colitis and a highly select subset of patients with Crohn's disease can also be candidates for the procedure.[1]

Currently, IPAA provides acceptable functional results and good postoperative quality of life in all age groups.[8] Restorative proctocolectomy with IPAA has been proven safe and feasible for maintaining intestinal continuity. However, data regarding the role of RP for pediatric patients, especially in terms of the durability of the ileo-anal pouch and complications associated with the procedure over the long-term, are limited.[9,10] The aim of this study was therefore to evaluate long-term outcomes of RP-IPAA, including pouch function and quality of life (QOL), in the pediatric population.

METHODS

According to the American Academy of Pediatrics, the upper age limit of pediatric practice is 21 years old.[11] Based on this standard, pediatric patients ($\leq$21 years old) who underwent IPAA at the Department of Colorectal Surgery, Digestive Disease Institute at the Cleveland Clinic between January 1983 and August 2010 were included in this study. Data were obtained from an institutional review board approved, prospectively maintained ileal pouch database. Demographics, indications for surgery, type of operation (laparoscopic, open/1-, 2-, or 3-stage), pouch configuration, type of ileo-anal anastomosis, whether or not a diverting ileostomy was created, final pathologic diagnosis, and operative results were recorded in this database.

Complications were evaluated and data regarding anastomotic stricture, obstruction, pelvic sepsis, pouchitis, postoperative hemorrhage, wound infection, pouch failure, anastomosis leak, and fistula formation were recorded. Follow-up time was defined as short-term, (first 30 days after IPAA creation) and long-term (more than 30 days after IPAA creation). Pouch failure was defined as excision of the ileo-anal pouch, permanent diversion with a proximal loop ileostomy, or having an unreversed diverting ileostomy.[12] Pelvic sepsis was defined as development of an abdominal, pelvic, or perianal infectious process detected by clinical, radiologic, or operative means.[13]

Patients in our database receive annual questionnaires that have been designed to evaluate functional outcomes and QOL. If patients were followed-up in another hospital, this information was obtained by mail or telephone contact. In this questionnaire, frequency of bowel movements, fecal incontinence, urgency, daytime/night-time seepage, pad use, and restrictions were assessed. Fecal incontinence and urgency were scored from 0 to 4 (0, never; 1, rare; 2, sometimes; 3, usually; 4, always) to evaluate symptom severity; anal seepage, pad usage, and restrictions were simply recorded as being present or not (0, no; 1, yes). Quality of life assessment was performed using the Cleveland Global Quality of Life (CGQL) score, which includes 3 items: current QOL, current health, and current level of energy, each on a scale of 0 to 10 (0, worst; 10, best). The scores are added and the final CGQL utility score is obtained by dividing the resulting number by 30 (range, 0 to 1; 0, worst; 1, best).[14]

Definitions

We defined the perioperative period as the time between hospital admission and discharge, and anastomotic leak as a break in the integrity of the anastomosis, as documented by a combination of clinical, endoscopic, radiologic, and operative findings. Bowel obstruction was defined as the presence of at least 3 of the following 5 symptoms: nausea, abdominal pain, vomiting, abdominal distension, absence of flatus and/or stool within the last 24 hours, findings indicating obstruction on plain x-ray or contrast studies, or a diagnosis of intestinal obstruction as confirmed by surgery.[1] Anastomotic stricture was diagnosed with digital examination in the outpatient clinic or operating room. Clinically significant anastomotic stricture was defined as one requiring an intervention due to outlet obstruction symptoms. Pouch-related fistula was defined as an abnormal passage or sinus from the pouch to another surface or organ. Pelvic sepsis was defined as development of an abdominal, pelvic, or perianal infectious process detected by clinical, radiologic, or operative means, and which occurred either within 3 months of loop ileostomy closure or within 3 months of restorative proctocolectomy, when stoma diversion was not performed.[13] Patients who complained about their pouch