Chapter 8
Crohn's Colitis and Ileal Pouch Anal Anastomosis

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Introduction

Traditionally, the ileal pouch anal anastomosis (IPAA) operation has not been offered to patients with Crohn's disease (CD). Patients with Crohn's colitis are often excluded from undergoing IPAA related to a number of key concerns: the risk of developing recurrent disease in the pouch necessitating pouch excision with possible ensuing short bowel syndrome, coupled with the risks of significant pouch dysfunction and the need for long-term medical therapy. However, surgical dogma is being challenged in more recent times with authors now reporting encouraging outcomes following IPAA in patients with either a preoperative or postoperative diagnosis of Crohn's colitis.

Patients with Crohn's colitis for whom an end ileostomy is not an acceptable option at that time have three potential reconstructive options to restore bowel continuity: ileorectal anastomosis, ileal pouch rectal anastomosis (IPRA) or ileal pouch anal anastomosis (IPAA), also known as restorative proctocolectomy. The first two restorative operations require either complete rectal sparing or sparing of the distal rectum whereas the latter is the only option to restore intestinal continuity in patients requiring proctectomy as a result of Crohn's proctitis. This chapter focuses specifically on these patients i.e., patients with documented CD of the colon and rectum requiring either a proctocolectomy or completion proctectomy after initial subtotal colectomy for disease management.

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Methods

A search of all English language PubMed articles from 1990 to 2015 was performed using the following terms: Crohn's disease, Crohn's colitis, ileal pouch anal anastomosis, restorative proctectomy, restorative proctocolectomy, completion proctectomy, proctocolectomy and ileostomy. These terms were in keeping with the PICO table below on which this chapter is based. All relevant articles were reviewed and appropriate references interrogated.

Patient Population	Intervention	Comparator	Outcomes studied
Crohn's colitis	Ileal pouch anal anastomosis (IPAA)	Proctocolectomy/completion proctectomy with end ileostomy	Pouch morbidity; pouch excision; quality of life

Results

There is a clear division in the literature regarding the outcomes of IPAA in CD in terms of the time of diagnosis of the primary disease. Studies divide the timing of the CD diagnosis as preoperative (resulting in an 'intentional' IPAA formation), perioperative (IPAA formation with 'incidental' or 'accidental' CD diagnosis on analysis of the surgical specimen) or at a later date following IPAA creation (so called 'delayed' diagnosis). A comparison of the data for these three distinct groups has been reported in prior studies. However, the ensuing recommendations and debate are based solely on those studies pertaining to patients with a documented diagnosis or high clinical suspicion of CD prior to undergoing IPAA, the aforementioned 'intentional' IPAA cohort.

The first published paper of 'intentional' IPAA formation in CD (patients in whom there was a high clinical suspicion based on the findings described below) was from Hyman and colleagues from the Cleveland Clinic in 1991 [1]. They reported on 25 patients with a postoperative pathologic diagnosis of CD out of 362 consecutive patients undergoing IPAA for a preoperative diagnosis of ulcerative colitis (UC). Of these 25 patients, 9 had preoperative features suggestive of CD: 5 with perianal disease (fistula, fissure or stricture), 2 with abnormal distribution of colonic disease, 1 with a cecal stricture and possible terminal ileal disease and 1 with a rectovaginal fistula. Although none of these 9 patients had a definitive preoperative diagnosis of CD, the above pathology would frequently be cited as a reason not to perform IPAA in cases with indeterminate pathology. At a mean follow-up of 34.8 months, only 1 of the 9 patients had a functioning pouch. Of the remainder, 1 died, 1 remained diverted and 6 had their pouch excised at a mean of 17.6 months postoperatively. The authors concluded that patients who manifest clinically as CD and have confirmatory pathology do very poorly following IPAA with short disease-free intervals and a high pouch failure rate.

Following this, Panis and colleagues published their initial results [2]. From 1985 onwards, they considered IPAA in selected CD patients in whom a proctectomy was required for either proctitis or rectal stenosis. Strict inclusion criteria were employed to ensure the disease was confined solely to the colorectum: all patients underwent an examination under anesthesia prior to IPAA to exclude anoperineal disease and also had a small bowel contrast study to exclude concurrent enteric disease. Eighteen patients were recruited over an initial 7-year period. These 18 patients were combined with a further 13 patients with a pre-IPAA diagnosis of indeterminate colitis (IC) which was subsequently shown to be CD in the postoperative specimen. This group then totaled 31 patients and reported outcomes were for the group as a whole (i.e., n = 31) and were not subdivided into the specific diagnostic timeframes of pre-operative (n = 18) or post-operative (n = 13) CD diagnosis. The results were encouraging: 6 patients had a CD-related complication with 2 of these ultimately requiring pouch excision and the remaining 4 patients reporting acceptable pouch function. Overall, 90 % of the cohort had a functional pouch at 5-year follow up. When compared with a corresponding ulcerative colitis (UC) cohort (n = 71) over the same time period, there was no demonstrable difference in terms of stool frequency, continence, gas/stool discrimination, leak or need for protective pads and sexual activity.

The same group subsequently reported on their experience with 41 patients, 26 of whom had a preoperative CD diagnosis [3]. Once again, the results in terms of CD-related complications are reported for the whole group and not reported in subgroup analysis for the intentional IPAA patients and incidentally diagnosed CD patients following IPAA. Twenty patients were followed for 10 years or more with a CD-related complication rate of 35 % and an impressive pouch excision rate of only 10 %.

The Cleveland Clinic adopted the intentional IPAA in CD patients in the late 1990's and subsequently reported its initial experience [4]. The analysis included 20 patients who underwent an intentional IPAA out of the study cohort of 204 patients (additional 97 patients with incidental diagnosis and 87 patients with delayed diagnosis). These 20 patients had a median time of 6.6 years from CD diagnosis to IPAA with a median follow up of 5 years and were more likely to be female. The 10- year pouch retention rate in the 20 patient strong intentional group was 85 % and thus closely mirrored the long-term follow up reported by Regimbeau and colleagues of 90 % pouch retention at 10 years as described above. For those patients with retained IPAA, 72 % reported near-perfect or perfect continence, 68 % reported rare or no fecal urgency and the median number of daily bowel movements was 7 (range 2 – 20). Interestingly, these patients also reported their quality of life and quality of health as 9/10 and 9/10 respectively and happiness with the IPAA procedure as 10/10.

The Mount Sinai Medical Center, New York, reported their experience with 13 patients who received an IPAA, 4 of whom were definitively diagnosed preoperatively with CD [5]. None of these patients had perianal disease and all had disease solely limited to the colon. Two of these 4 patients (50 %) subsequently developed perianal disease, 2 (50 %) developed postoperative complications and 1 patient

(25 %) required a pouch excision. Of note, the outcomes for all 13 CD patients were compared with a matched cohort of patients undergoing IPAA for chronic UC; the CD patients had fewer bowel movements per 24 h, a lower incidence of incontinence and a lower incidence of pouchitis.

The most recent series on the intentional use of IPAA in CD patients reported on 17 patients [6]. Seven of 17 patients (41 %) developed recurrent CD following IPAA and this compared with a corresponding postoperative incidence of 11 % in a UC cohort undergoing IPAA during the same time period. The pouch excision rate over an average follow up of 60 months in the 17 preoperatively diagnosed CD patients was an impressive 6 %. This study is also notable in that 9 of the 17 patients had a preoperative diagnosis of CD outside of the colorectum: 5 patients had previously undergone small bowel resections with no evidence of active small bowel disease and 4 patients had perianal disease (3 perianal fistulae, 1 anal stenosis), where the fistulae were managed by insertion of draining setons with subsequent evaluation demonstrating no evidence of active perianal sepsis.

The most current study on this topic is a United States multi-institutional study examining the cost-effectiveness of two surgical options in patients with Crohn's colitis [7]. They compared what is referred to as 'colectomy with permanent ileostomy' with IPAA. It should be noted that some of the evidence for the former group involves patients described in a prior study who underwent either total abdominal colectomy with end ileostomy or panproctocolectomy with end ileostomy [8] and the reader cannot determine whether it was only the panproctocolectomy patients who were included in the cost analysis by Taleban and colleagues. Additionally, Taleban and colleagues assumed that patients undergoing J-pouch formation would have 'complete mucosectomy', yet this is clearly not the operative approach employed by all. Nonetheless, colectomy with permanent end ileostomy was shown to be more cost-effective unless the associated surgical cost exceeded $20,167 at which point IPAA was the more effective option. They also reported that IPAA was the more effective strategy with an incremental cost-effectiveness ratio of $70,715 per QALY gained.

Author	Year	Number of patients	Postoperative morbidity	Pouch excision	Quality of evidence
Hyman	1991	9	8/9 (89 %)	6/9 (67 %)	Low
Panis	1996	31 (18 intentional; 13 incidental)	11/31 early (35 %) 6/31 CD related (19 %)	2/31 (6 %)	Low
Regimbeau	2001	41 (26 intentional; 15 incidental)	10/41 early (24 %) 11/41 CD related (27 %)	3/41 (7 %)	Low
Melton	2008	20	Not reported	2/20 (10 %)	Low
Grucela	2011	4	2/4 (50 %)	1/4 (25 %)	Low
Le	2013	17	4/17 early (24 %) 7/17 CD related (41 %)	1/17 (6 %)	Low

Recommendations Based on the Data

Since the introduction of IPAA as part of our surgical armamentarium, there have only been 67 patients reported with a preoperative diagnosis of CD and thus an intentional IPAA. This number can be increased to 76 when the 9 patients with a high preoperative suspicion of CD reported in the initial study from the Cleveland Clinic are included. Based on this, the evidence for intentional IPAA in Crohn's colitis is low and the recommendation for IPAA formation in patients with Crohn's colitis is weak.

A Personal View of the Data

The top priority is providing a personalized and tailored plan of care for each patient. We believe that some of the most critical and complex parts of working with a patient with Crohn's disease occur outside of the operating room. Not only is it imperative that detailed medical and surgical histories are obtained, but it is also essential to develop a relationship with the patient at the first encounter and to gain an understanding of the patient's goals in terms of the potential for surgery and possible outcomes. The patient and their family/caregivers should be approached on a personal level, understanding their own goals and work for open, honest dialogue whilst forming a specific individual surgical strategy. Having done this, together the patient and colorectal surgeon embark on a lifelong relationship. In our experience, this specific group of patients are very well informed on the potential surgical options and present to us with the intention of undergoing IPAA.

The formation of an IPAA for Crohn's colitis is considered provided there are no gross manifestations of small bowel disease (unless it is backwash ileitis) or perianal CD; a single, limited perianal fistula can be acceptable but a rectovaginal fistula is not. CT enterography is the preoperative imaging modality of choice to examine the small bowel and a thorough bedside perianal examination is performed and if there are questionable findings, patients proceed to a formal examination under anesthesia. Risk factors, especially a personal history of smoking and a family history of CD, are always sought as these patients are at increased risk for subsequent development of CD of the ileal pouch. Patients referred from other institutions may undergo repeat colonoscopy with biopsies and all previous outside pathology slides are reviewed again by a dedicated inflammatory bowel disease histopathology team. All patients have their nutritional status optimized preoperatively. Preoperative counseling regarding the potential for complications is extensive, with particular emphasis on the risk for significant small bowel loss if there is a requirement for pouch excision and that a re-do pouch may not be an option. Similarly, patients are advised that even if preoperative imaging is reassuring, there is always the potential that small bowel CD may be discovered perioperatively.

The technical approach to IPAA relies on careful and meticulous handling of the bowel and dissection in natural, anatomic tissue planes. When presented with a new patient with isolated Crohn's colitis, a 3-stage procedure is recommended and patients should ideally be steroid and biologic free prior to the second stage (i.e., pouch formation). We do not recommend a one-stage procedure and will perform a 2-stage procedure in select cases. Regardless of the operative approach (open or laparoscopic), the small bowel must be examined in its entirety from the ligament of Treitz to the ileocecal valve and if there is a suspicious area, this should be interrogated and may require an enterotomy to ensure there is no luminal evidence of disease. We strongly favor the total mesorectal excision technique when performing proctectomy. Residual distal tissue may lead to pouch emptying issues, which may significantly affect pouch function and quality of life. The J-configuration is the pouch subtype of choice and the double-stapled pouch-anal anastomosis technique is favored. In highly motivated patients who wish to avoid a permanent ostomy in whom there is limited perianal disease as previously referred to, a mucosectomy and hand-sewn anastomosis can be utilized when necessary. We have previously reported on the learning curve for IPAA formation which is estimated to be 23 cases when performing the stapling technique [9]. All new IPAAs are defunctioned after their creation for a minimum of 3 months and interrogated with a radiological contrast enema prior to ileostomy closure.

In the unfortunate case when a Crohn's patient with an IPAA develops anoperineal sepsis or anastomotic issues, the algorithm is to begin by checking one's own 'footsteps': it is critical to distinguish symptoms due to sequelae of Crohn's disease from a technical complication (which are more likely to develop within 3 months of surgery). These have very different solutions and management approaches to say the least.

IPAA surgery in patients with Crohn's disease is technically and emotionally challenging, but is also rewarding in that it offers a life-changing avenue for the patient and surgeon alike.

Patients with Crohn's colitis and no evidence of small bowel or perianal disease are candidates for IPAA following thorough preoperative counseling and discussion regarding potential postoperative outcomes (evidence low; weak recommendation).

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