

Ileal Pouch-anal Anastomosis for Ulcerative Colitis – The Rhode Island Experience

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INDICATIONS AND RATIONALE FOR SURGERY IN ULCERATIVE COLITIS

Despite significant progress in the medical management of ulcerative colitis (UC), over 20% of patients affected by this chronic and medically incurable disease still require surgical intervention. There are three main groups of indications for an operation in UC: acute life-threatening complications, medical intractability or side effects, and cancer risk.¹

Severe complications include massive hemorrhage, obstructing stricture, toxic colitis or megacolon, and perforation with sepsis. UC patients often require emergency surgical care while debilitated by anemia, malnutrition and immunocompromised from their chronic disease or from medications (e.g. azathioprine, 6-mercaptopurine, corticosteroids, infliximab, etc.). Timing of surgery in patients hospitalized for severe UC is best determined by a coordinated effort between gastroenterology and colorectal surgery staff.

Patients may become unresponsive to medications, or dependent on them, or develop severe side-effects from chronic use. Particular care and precautions have to be considered in these patients who are at higher risk of surgical infections, poor wound healing and venous thrombo-embolic events from their hypercoagulable state. Growth failure in children is also a recognized indication for colectomy.²

In patients with early onset and long-standing (> 10 years), extensive disease (pancolitis), there is also a progressive increase in the dysplasia-carcinoma sequence over time. The cumulative cancer risk has been estimated at 2% at 10 years, 8% at 20 years, and 18% after 25 years of colitis.³ Such risk is quadrupled in patients with primary sclerosing cholangitis as one of the extra-intestinal manifestations associated with UC.

Since UC is limited to the large intestine, removal of the target organ will achieve definitive cure. The colon and rectum are not essential for nutrition and/or survival and their removal does not

require any nutritional supplementation. Small bowel adaptation over several months allows it to assume some colon-like absorptive functions, a startling example of the adaptability of our gastrointestinal tract to anatomical and environmental changes (e.g. stasis, pH, bacterial flora).

HISTORY OF SURGICAL OPTIONS FOR ULCERATIVE COLITIS

Until about thirty years ago, surgery for UC patients essentially meant accepting the need for a permanent ileostomy, after removal of the whole colon, the rectum as well as the anus. Understandably, the prospect of a permanent ileostomy bag remains a powerful deterrent to patients who may be surgical candidates.

IPAA...is now the most commonly performed procedure for the definitive treatment of UC.

Ideally an operation for ulcerative colitis should satisfy four major criteria: 1) ability to cure by completely eliminating the disease; 2) safety in its acceptable complication rate; 3) applicability to most affected patients; 4) effectiveness in restoring quality of life and preserving enteric continence.⁴ The operation that comes closest to this "ideal" is the **ileal pouch-anal anastomosis (IPAA)** or restorative proctocolectomy. It involves complete removal of the colon and rectum, but it preserves the anal mucosa and sphincteric complex, to which an ileal reservoir is attached, thereby maintaining continence. Sir Alan Parks et al in 1980 in England first reported experience with ileal pouch-anal anastomosis in humans, and the operation bore his name for several years thereafter as "Parks' procedure."⁵ However, as is often the case, such accomplishment was the culmination of decades of efforts on the part of multiple scientists and investigators.

Nils Kock in Sweden in 1969 had described an ileal reservoir that was attached to the skin of the lower abdomen after complete removal of anus, colon and rectum.⁶ This "continent ileostomy" still did not obviate the need for a stoma, required significant patient compliance and education in emptying it several times a day, and was associated with a high complication and revision rate.⁷ A more recent modification, the **Barnett continent ileal reservoir (BCIR)**, larger than the "Kock pouch", has a different valve mechanism; presently this operation is rarely indicated and performed.

The idea to reestablish enteric continuity by attaching the ileum to the anus was first suggested in 1933 and later the United States in the late 1940s.⁸ Therefore, the ileal pouch-anal anastomosis can be seen as a merger of the two concepts of anal preservation and fashioning of a reservoir, in order to maintain continence and improve quality of life after proctocolectomy for UC and other conditions. IPAA is generally contraindicated in patients with Crohn's disease, given the high risk of recurrent disease and complications leading to pouch failure.

In acute life-threatening emergency situations, the preferred surgical procedure is a **total or sub-total colectomy** with temporary ileostomy. At a later date, a completion proctectomy can be associated with an IPAA.

In patients with very poor anal sphincter function, anal preservation is not warranted, in view of expected poor functional outcome. Such patients are best served by a **proctocolectomy with permanent ileostomy**, when surgery is necessary.

In most patients undergoing IPAA in the elective or urgent setting (not emergency), an IPAA is generally accompanied by a temporary ileostomy that can be reversed eight to twelve weeks later. In selected patients who come to surgery in very good overall health, a temporary ileostomy may be omitted and the whole procedure may be performed in one stage.

IPAA has gained progressive acceptance over the past two decades and is now

the most commonly performed procedure for the definitive treatment of UC.

PATIENT OUTCOME AFTER RESTORATIVE PROCTOCOLECTOMY

Proctocolectomy with IPAA is safe, durable and applicable to most patients with UC.⁹ Although its overall complication rate can be in the 20-30% range, its mortality is generally below 1% in large series from very experienced surgeons.^{10, 11} Complications of IPAA can occur in the immediate postoperative period or months and years later.

Early surgical complications include bleeding, small bowel obstruction, pelvic sepsis and anastomotic leak. In some situations, reoperation may be required to preserve pouch function, but often percutaneous interventional techniques may suffice.

Late complications may include incisional hernia, anastomotic stricture, pouch-cutaneous or pouch-vaginal fistulas, and pouchitis. Some fistulas may be managed medically, especially if Crohn's disease is suspected, but most eventually require surgical correction. Pouchitis is the most common long-term complication and its incidence increases with time. However, in most cases this pouch mucosal inflammation is transient; it responds to antibiotic therapy and may be controlled with probiotics as well. Pouch failure requiring excision and conversion to a permanent ileostomy is rare and generally occurs in less than 3% of patients. Pelvic cysts may occur as a result of adhesions and fertility problems in women are more frequent after IPAA than in the general population.¹² (See "Reproductive Issues in Inflammatory Bowel Disease" by Sumona Saha, MD, and Silvia Delgi Esposti, MD in April 2009 *Medicine & Health/Rhode Island*.) The safest delivery method for a pregnant woman after IPAA remains controversial. Cesarean section prevents anal sphincteric and pudendal nerve injury from forceps delivery, vaginal tears or episiotomies. Vaginal delivery has been reported as safe in the short term follow-up, but may lead to higher rates of incontinence with advancing age. Although sexual dysfunction in men after proctectomy from injury to the hypogastric plexuses and pelvic autonomic nerves may occur, most men describe an improvement in sexual function after IPAA, likely as a result of their improved overall health.

The functional results have yielded quality of life standards that approach those of the healthy population and demonstrate consistently improvement over pre-surgical performance status for patients that are not doing well with medical management.¹³ The average number of daily bowel evacuations is higher than normal controls at 2-8 (average 4-5) during daytime and 0-1 at nighttime. Nonetheless, in terms of quality of life, a more important element is the ability to defer defecation, which is greatly improved to 30-60 minutes with a healthy ileal reservoir and a good anal sphincter. In fact, some of the most aggravating symptoms in UC patients are the urgency and tenesmus secondary to the inflammation and lack of distensibility of the rectum, with consequent loss of its reservoir volume and compliance. Over a period of ten to twelve months, the pelvic ileal pouch undergoes progressive dilatation and mucosal adaptation to colonic metaplasia, flattening of its villi and increase in goblet cells. Such adaptive changes, likely secondary to stasis and increased bacterial anaerobic counts in the pouch, enhance its storage capacity, absorptive function, ability to thicken stool and decrease frequency. It is important to follow patients with dietary advice and stool retardant medications as needed, to optimize functional outcome. Medications used to treat UC before surgery can be discontinued postoperatively and corticosteroids can be tapered off gradually.

THE RHODE ISLAND EXPERIENCE

Since the author performed the first IPAA procedure in Rhode Island, at Rhode Island Hospital in April 1991, the collaboration and dedication of gastroenterologists and surgeons, combined with enterostomal therapists, nutrition and other support services has allowed the growth of a very successful and comprehensive program. The proximity of Women and Infants' Hospital on the same campus has also facilitated collaboration of care in pregnant women with exacerbation of inflammatory bowel disease and toxic colitis. Since the late 1990s, pediatric surgeons at Hasbro Children's Hospital have performed IPAA procedures in children. More recently, the contribution of additional colorectal surgeons in the

state has broadened the availability of these procedures to other hospitals.

Over the past eighteen years, the author's personal experience includes 312 patients,

166 males and 146 females, ranging in age from 9 to 75 years old (mean = 36 years old).

The indications for surgery were UC in the vast majority of cases (287), familial polyposis in 23 patients and colorectal inertia in 2 cases. Six of the UC patients had a carcinoma at the time of surgical resection. Although ileal pouches can be S-shaped, W-shaped, T-shaped or J-shaped, all patients in this series had the J-shape for its ease of construction and equivalent storage ability after adaptation. Another area of controversy involved the choice of transanal mucosal excision and hand-sewn anastomosis versus the double-stapled technique. We have used both options depending on patient's variables, after conducting a trial to individualize operative choices on the basis of age, anal sphincter baseline function, as well as cancer risk.¹⁴ To date, 275 pouches were double-stapled and 37 were hand-sewn.

Most patients (n = 222, 71%) underwent a two-stage procedure, while 26 (8%) required a three-stage IPAA as a result of life-threatening indications. The remaining 64 patients (21%) could be done in a single stage, by virtue of their overall good preoperative health. An increasing number of IPAA's have been done with the laparoscopic minimally invasive techniques in recent years. This technical advance has decreased postoperative convalescence time more than hospital length of stay for this particular operation in our experience. The use of bioresorbable membrane application to reduce peritoneal adhesion formation during surgery and other technical innovations have contributed to a low rate of small bowel obstruction, pelvic cyst formation and preservation of fertility in both female and male patients.

Major short-term complications in our series included two pouch anastomotic leaks (0.6%), one ileostomy closure leak (0.3%), one pouch necrosis (0.3%), five deep venous thromboses (1.6%) and one death secondary to pulmonary embolism (0.3%).

Major long-term complications have included pouch-vaginal fistula in 5 patients (3 with unsuspected Crohn's disease)

(1.6%), one anal cuff carcinoma twelve years post-IPAA (0.3%), and chronic idiopathic pouchitis requiring treatment in 34 patients with a diagnosis of UC (11%). Eventually, 11 patients (3.5%) had their diagnosis changed to Crohn's disease.

Overall, only 5 patients (1.6%) have required conversion to permanent ileostomy, one for anal cuff cancer, one for pouch necrosis, and three for intractable complicated pouch and perineal Crohn's disease.

Our patient support network allows patients considering having IPAA surgery to talk to individuals who have undergone the procedure. Given our large case series, we match patients by age and gender, so they are more comfortable in discussing issues of recovery, lifestyle, and quality of life.

In addition to our clinical efforts, our program has collaborated with basic scientists in the Division of Gastroenterology at Brown University. This fruitful interaction has led to several publications that have expanded knowledge and deepened understanding of UC, especially as it relates to alterations in smooth muscle contractility as a result of mucosal inflammation.¹⁵ Until we discover the etiology and can better elucidate the pathogenesis of this complex disease, surgery is likely to remain the only option able to provide definitive cure.

In summary, Rhode Island can offer state-of-the-art expertise in management of ulcerative colitis, including surgical expertise with a track record of outcome results that exceed national standards.

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The author has no financial interests to disclose.

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