

Chapter 30

Deciding on an IRA vs. IPAA for FAP

James Church

Setting the Stage

Familial adenomatous polyposis (FAP) is a dominantly inherited form of cancer predisposition due to a germline mutation in the colorectal cancer gateway gene, *APC*. The syndrome usually presents as colorectal adenomatous polyposis of varying severity, which, if untreated, will lead to colorectal cancer at a young age. While other organs are also affected by the cancer predisposition, by far the most serious threat to life and lifestyle comes from the large bowel. This is therefore the initial focus of treatment.

Patients with FAP are usually diagnosed on screening because dominant inheritance combined with 100% penetrance makes the family history compelling. Genetic testing identifies affected family members, who begin colonoscopic surveillance at puberty. If genetic testing is not done or is uninformative, colonoscopic surveillance is the same, but is applied to every at risk relative. Patients diagnosed by screening are usually asymptomatic and the polyps are small. There is plenty of time to answer the next two important questions: what surgery and when?

About 25% of patients with FAP do not have a family history, do not suspect the syndrome they have and the risks they carry, and ultimately present with symptoms due to relatively advanced disease [1]. These patients have a high risk of having a colorectal cancer at diagnosis, and in general have more severe disease than those diagnosed by screening. The same two questions apply however: what surgery, and when?

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Aims of Surgery in Patients with FAP

Some studies addressing the issue of colorectal surgery in FAP seem to have lost sight of the true aims of the procedure [2, 3]. The focus tends to be exclusively on treating and preventing cancer (in particular, preventing death from cancer) while the secondary aim of lifestyle preservation is often disregarded. This leads to the preference of ileal pouch-anal anastomosis over ileorectal anastomosis for all or most patients with FAP, regardless of polyposis severity. However, when operating on young asymptomatic patients at a critical time in their social, sexual, academic, educational and psychological development, it is critical that prophylactic surgery does not cause harm. In fact, the two main surgical options are considerably different in their impact on lifestyle.

The Surgical Options

To absolutely prevent colorectal cancer, all of the colon and rectum must be removed. This leads to an end ileostomy, which is unacceptable to most patients, especially if they are young and asymptomatic. Before 1980, total colectomy and ileorectal anastomosis (IRA) was performed as a reasonable compromise, reducing the risk of cancer considerably but maintaining normal defecation. However, in patients with profuse polyposis, the risk of rectal cancer after IRA was high, as the surgery was too conservative [4]. The game changed in 1979 when the ileal pouch-anal anastomosis (IPAA) entered practice. It achieved near-complete removal of the colon and rectum while per anal defecation was preserved. Since then the IPAA has become an important option for the treatment of patients with FAP, and yet there is still debate over the indications for IRA and IPAA in patients with FAP. This chapter is devoted to a discussion of this choice and to providing guidance about making it.

How Are the Outcomes of Surgery to Be Judged?

Surgery is judged on the extent to which it achieves its aims. Prevention of cancer after surgery in patients with FAP is judged on the rate of metachronous cancer. However one of the advantages of an IRA is that the rectum can be removed at a second operation before cancer arises or before cancer has spread. In this circumstance, proctectomy can almost always be accomplished, and an IPAA can be constructed most of the time [5–7]. On the other hand patients with an IPAA are not free of cancer risk, either in the pouch-anal anastomosis, or the body of the pouch itself [8]. Anal transition zone (ATZ) cancer is more likely in patients with a stapled IPAA than a handsewn IPAA, arising from residual glandular epithelium [9, 10]. However

both types of anastomosis carry some type of risk [8, 9]. The literature is split in terms of the relative complication rates and function of stapled IPAA vs mucosectomy and hand-sewn IPAA [11–14]. However stapled IPAA is certainly easier to survey.

Quality of life, the second outcome to be considered, is difficult to measure or judge. There is often little correlation between bowel function and quality of life, as this measurement is always subjective and relative [15]. Under the best of circumstances, both IRA and IPAA can be followed by a nearly normal lifestyle. Under the worst of outcomes, life is miserable [16]. The quality of functional outcomes after IRA and IPAA depend largely on surgical skills and patient factors such as BMI, gender and compliance with follow-up. Adding to the complexity of evaluating these operations is the source of much of the information, specialty units, where there is broad experience and high skill. The relevance of these reports to the less experienced surgeon in usual practice can be debated.

Quality of Surgery

The “elephant in the room” when discussing surgery for patients with FAP is the quality of surgery, as this has a huge effect on quality of life. The stakes are high in this disease because many patients are young and are at critical developmental stages physically, emotionally, socially and academically. In addition, the majority are asymptomatic. To take a young, asymptomatic patient and leave them incontinent, impotent, or dealing with a permanent ileostomy may be considered a tragedy, especially when the operation that was so complicated was either unnecessary or too radical for the disease [16].

Both operations for FAP are technically challenging. An ileorectal anastomosis involves a difficult anastomosis between two ends of very different diameters. It is probably the most prone of intra-abdominal anastomoses to leak. An IPAA is also technically demanding, as there are multiple aspects of techniques that have to go well. There can be no tension on the small bowel mesentery. The bowel has to descend into the pelvis straight, without as much as a 90° twist to the side. The anastomosis should be at the level of the pelvic floor or below and the pelvic nerves and other organs must be protected. We have seen many poor outcomes due to sub-optimal technique and have reported on some of them, including a 360 twist in the small bowel around its mesentery, an ultra-long efferent limb of small bowel from an S pouch to the anus, an IPAA 7 or 8 cm from the dentate line, incorporation of the vagina in an anastomotic staple line, and construction of a tiny pouch that holds very little stool [16]. Functional problems include passing up to 20 stools per day, severe fecal incontinence, disabling anal pain, and impotence. Surgeons should be very familiar with the technique of whichever operation they choose, or refer the patient to a high volume center. Bad outcomes have effects beyond the patient when relatives fail to be screened or to follow through on surgery out of fear of having a similar outcome.

What Do the Data Say?

A Medline and Pubmed search using the terms FAP, Familial adenomatous polyposis, surgery, ileal pouch anal anastomosis, ileo-anal anastomosis was conducted and then extended by searching by the names of those this author knew had written about the topic, going from 2015 back to 1946.

There are no randomized, prospective studies upon which to base surgical decisions in patients with FAP. Most are retrospective reviews of experience from large clinics, comparing cohorts of patients [17–22]. There is also one decision analysis [23] and one reasonable meta-analysis [24]. The decision analysis is flawed due to the weight given to the incidence of rectal cancer after IRA, many of which date back to the “pre pouch” era. Many studies of IPAA function include patients with ulcerative colitis and FAP, and should be excluded from consideration, as the diseases are so different. In addition there are few recent studies, most dating back at least 10 years. During this time surgery has changed considerably with minimally invasive techniques now almost routine [25].

Perhaps the most sensible data on oncologic outcome of an IRA come from the Cleveland Clinic. They were the first to explain the high rates of rectal cancer after IRA as being due to the lack of surgical options prior to 1980, when IPAA entered practice [4]. When the only options are IRA or a permanent ileostomy, it is not surprising that most patients choose IRA, even those with severe polyposis. These are the patients who would go on to develop rectal cancer or advanced rectal polyposis. After 1980, patients with profuse polyposis had an IPAA and the incidence of rectal cancer after IRA dropped significantly.

The Cleveland group also set the criteria for either operation, based on rectal and colonic polyp counts at the preoperative colonoscopy [26]. Patients with <20 rectal polyps could safely have an IRA while those with >20 rectal polyps would be better served by an IPAA. These standards have stood the test of time and have resulted in an almost 50:50 ratio of IRA to IPAA in that institution [25].

While some institutions perform IPAA on every patient with FAP [2], most use criteria to select for IRA. Polyp count is the most powerful factor but others enter into the decision-making. Genotype has been suggested as a criterion for triaging patients according to the location of their mutation [27, 28]. However operating by genotype adds nothing to the use of polyp counts, as the correlation between profuse polyposis and genotype is close to absolute, and that between genotype and attenuated polyposis is also predictable. In young female patients an IRA may be selected to avoid the possibility of reduced fecundity after an IPAA. This sort of “staged” pouch (IRA first, knowing that proctectomy is likely to be needed later after childbirth) also avoids a stoma in the young, provides better bowel function during the key stages of a patient’s life, and may well reduce the risk of desmoid disease [29]. It is a strategy that has become increasingly popular, especially as there is often a spontaneous decrease in rectal polyps for several years after IRA [30], and rectal polyposis can often be controlled

by aggressive endoscopy. Of course a rectum that is carpeted with adenomas, usually in a symptomatic patient, has to be removed and some patients must have an IPAA.

Studies measuring functional outcomes and quality of life after IRA and IPAA generally report similar themes: that bowel function is better after IRA than IPAA with less lifestyle restrictions and is stable over time [31–35]. IPAA function is very variable over a range of stool frequencies and continence scores. However, quality of life seems high. In many reports there is an important difference between a stapled IPAA and a handsewn IPAA with a mucosectomy. A stapled IPAA generally has better function with fewer complications than a handsewn IPAA, and is definitely easier to survey. It has twice the incidence of anastomotic and ATZ neoplasia however [9, 36]. This ATZ neoplasia can be difficult to deal with if the residual ATZ/rectal stump is over 2 cm long. A handsewn IPAA does not guarantee a neoplasia-free zone, and is trickier to survey during unседated pouchoscopy. Some studies report good functional results with low complication rates after handsewn IPAA [2, 11–14]. If the technical ability of the surgeon can produce such results then a handsewn IPAA is a good choice. Some surgeons have better outcomes after a stapled IPAA, and this option offers the chance of an undiverted pouch. We would recommend that residual ATZ be less than 2 cm in length for easier management of neoplasia [36].

The role of surgical choice in stimulating desmoid disease is controversial. Data from the Cleveland Clinic suggest that IPAA doubles the risk of desmoid disease, and that laparoscopic IPAA is particularly desmoidogenic [29]. Others disagree and confirmatory data has not been reported to date [37, 38]. However there have been no other similar studies. It is plausible that the stretching of the small bowel mesentery that is part of an IPAA is the key factor in producing desmoid disease in the small bowel mesentery. When this is done in young women with a family history of desmoid disease, the perfect storm for desmoid formation occurs. Such patients should have an IRA.

Recommendation

Patients with <20 rectal and <1000 colonic adenomas are candidates for IRA. Patients with >20 rectal and >1000 colonic adenomas, or a curable rectal cancer on presentation, are better served with an IPAA. The IPAA can be stapled as long as the ATZ is free of adenomas and the length of the residual ATZ is minimized (<2 cm). Patients at high risk of desmoid disease should have an IRA.

Regardless of the procedure chosen, every patient should be surveyed endoscopically at least once a year.

Table 30.1 shows the advantages and disadvantages of IRA and IPAA, and the indications and contraindications for these procedures. Table 30.2 show the indications and contraindications for each operation.

Table 30.1 Indications and contraindications, advantages and disadvantages of ileorectal anastomosis and ileal pouch-anal anastomosis, in patients with familial adenomatous polyposis

	Ileorectal anastomosis (IRA)	Ileal pouch-anal anastomosis (IPAA), mucosectomy and handsewn anastomosis	Ileal pouch-anal anastomosis (IPAA), stapled anastomosis
Indications	<20 rectal adenomas Young patient High desmoid risk Woman	>20 rectal adenomas Older patient Low desmoid risk Adenomas in ATZ	>20 rectal adenomas Older patient Low desmoid risk ATZ clear of adenomas
Contraindication	Rectal cancer Uncontrollable rectal polyposis	Fecal incontinence Obese High desmoid risk	Fecal incontinence Obese High desmoid risk ATZ adenomas
Advantages	No ileostomy Low complications Good bowel function Low risk of desmoid disease	Minimal risk of rectal cancer Per anal defecation No urgency Low risk of ATZ neoplasia	Minimal risk of rectal cancer Per anal defecation No urgency Minimal seepage/incontinence
Disadvantages	Risk of rectal cancer	Temporary stoma Higher complication rate, both early and late Abnormal bowel function with seepage and incontinence High risk of desmoid disease Risk of ATZ polyps and cancer	High risk of ATZ neoplasia

Table 30.2 The outcomes of ileorectal anastomosis and ileal pouch-anal anastomosis: a literature review

Study	Variable	IRA	IPAA	P
Campos et al. (2009) N=88 1977–2006	Complications Cancer	19.0% 16.6%	48.1% 3.8%	0.03
Gunter et al. (2003) N=151 1970–2000				
Soravia et al. (1999) n=131 1980–1997	Anastomotic leak Bowel obstruction Function	3% 15% Less nighttime stooling, better continence, less skin irritation	12% 24%	0.21 0.58
Bjork et al. (2001) n=131 1984–1996	Complications Function	26% Less night time stooling, better continence, less skin irritation	40%	<0.05
Vasen et al. (2001)	Cancer	Risk of death from cancer 12.5% by age 65	Increase in life expectancy by 1.8 years	
Koskenvuo et al. (2014)	Secondary proctectomy Anus preservation rate during secondary proctectomy Cancer rate	39/140 49% 24% at 30 years		
Niewenhuis et al. (2009)	Secondary proctectomy by genotype Attenuated Intermediate Severe	10% 39% 61%		
Ko et al.	Bowel movements per day Leakage Pads usage Perianal skin problems Food avoidance Inability to distinguish gas	5.2 0 0 7% 43% 7%	7.5 43% 17% 33% 80% 37%	<0.05 0.01 <0.01 <0.01 <0.01 <0.01
Wuthrich	Soiling >6 bowel movements/24 h		25% 67%	

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