

Different surgical strategies in the treatment of familial adenomatous polyposis : what's the role of the ileal pouch-anal anastomosis ?

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Abstract

Background and study aims : Restorative colectectomy (RCP) with ileal pouch-anal anastomosis (IPAA), is one of the surgical responses to the crucial question of prophylactic treatment in familial adenomatous polyposis (FAP). No consensus has been reached, until now, to choose between IPAA and ileo-rectal anastomosis (IRA), the rectal sparing prophylactic colectomy. This paper aims to review the latest issues related to IPAA and highlights its specificities compared to IRA.

Methods : PubMed database was searched using the following search items : familial adenomatous polyposis, surgery, ileal pouch-anal anastomosis, ileo-rectal anastomosis. Papers published between 1978 and 2010 were selected.

Results : Absence of mortality, acceptable morbidity and good functional results combined to high quality of life have promoted the IPAA technique. New technical issues such as the double stapled technique, mesenteric lengthening, omission of temporary protective stoma can be addressed almost systematically for these patients. A laparoscopic approach, lessening the body image impact, has proven to be as effective and safe as the open approach to perform IPAA. Further advantages of laparoscopic IPAA rely on the lower adhesion formation resulting in less small bowel occlusion. Sexuality, fertility and childbirth are important functional issues often cited as threatened by the pelvic manoeuvres of the IPAA technique which can be prevented by close rectal wall dissection and a laparoscopic approach.

Conclusion : IPAA offers the best available prophylaxis in FAP patients. Technical enhancements in IPAA will most probably decrease the functional risks. Thus IPAA remains the alternative to IRA for the prophylactic treatment of FAP. Nevertheless, based on the latest evidence, the choice between both procedures is still matter of debate. (*Acta gastroenterol. belg.*, 2011, 74, 427-434).

Introduction

The question of adequate prophylactic surgical treatment for familial adenomatous polyposis (FAP) patients remains unsettled. In this autosomal dominant syndrome due to a germeline mutation of the adenomatous polyposis coli (*APC*) gene, hundreds to thousands of polyps develop on the colorectal mucosa, inevitably leading to colorectal cancer in the early decades of life. Thus, prophylactic removal of the potentially cancer-baring colon is considered as standard management of these patients, but the exact extent of the resection is still questioned (1-5).

Either total colectomy and ileo-rectal anastomosis (IRA) can be proposed or restorative proctocolectomy (RPC) with ileal pouch-anal anastomosis (IPAA). In the

opposite of IRA, the latter strategy addresses the rectal cancer risk, and thus appears as a more radical solution. Nevertheless, specific questions have to be considered in the decision making strategy for IPAA : technical issues, potential residual transitional zone mucosa in double stapled anastomotic techniques, loop ileostomy omission, laparoscopic approach, morbidity and mortality, functional outcome and quality of life, pouchitis, pouch adenomas and adenocarcinoma, sexuality and fertility.

The aim of this article is to review literature discussing surgical prophylactic treatment of FAP with particular emphasis on IPAA specificity and to discuss the best surgical strategy between IPAA and IRA.

Original surgical technique of IPAA

The standard technique, introduced by A. Parks and R.J. Nicholls in 1976 (6) includes several important steps

- total colectomy,
- proctectomy,
- endoanal mucosectomy,
- ileal pouch-anal anastomosis,
- diverting ileostomy (1,7-14).

Except for the patients whose polyposis is complicated by colonic or rectal cancer or severe dysplasia, surgical dissection can be performed close to the serosa of the colon and rectal dissection away from the sacral promontory and sacral fascia to avoid damage of pelvic autonomic nerves. After removal of the colorectal specimen, mucosal stripping from the anorectal stump begins at the dentate line by a perineal approach. While exposing for this mucosal dissection, every effort is made to avoid excessive stretch or injury of the anal sphincter muscles. The mucosectomy is carried out circumferentially to the top of the anal canal above the levator ani.

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The mostly used type of ileal reservoir is J-shaped, as originally described by Utsunomiya (15). The J-pouch configuration is favoured for its simplicity, speed of formation, its excellent fit into the concavity of the sacrum, excellent emptying, its reservoir capacity (usually nearing 400 ml) and its paucity of long-term complications (7,8). No advantages in terms of functional outcome or complications could be achieved with other pouch types in particular with W-pouch (16).

IPAA itself is a hand-sewn anastomosis performed at the dentate line level through a perineal approach. A temporary diverting loop ileostomy was originally systematically performed and closed 2-3 months later after a pouchogram had confirmed the integrity of the pouch and the ileoanal anastomosis (7,9,14).

Since the first report of this original technique, constant improvements have been proposed to render this rather tedious procedure easier in order to improve morbidity and yet to offer cure to the patients. Quality of life and body image are central concerns of the surgeons regarding these patients operated in a prophylactic setting. Mesenteric lengthening techniques, double stapled anastomosis, omission of loop ileostomy, and laparoscopic approach are some of the topics referring to those requisites.

Mesenteric lengthening technique

The aim of all mesenteric lengthening techniques is to achieve a tension-free IPAA in order to avoid postoperative anastomotic dehiscence and thus the need for a temporary diverting ileostomy. Division of the ileocaecal artery has often been presented as the safest and most effective method for obtaining maximum length. Marginal arcade of the right colon, preserved with its blood supply from the middle colic artery, allows both the ileocolic artery and even distal superior mesenteric artery to be safely divided, gaining significant extra mobilisation of the apex of the pouch permitting tension-free IPAA in all cases with omission of a loop ileostomy even after endoanal mucosectomy and anastomosis down to the dentate line. Right colic arcade preservation is of course precluded in FAP patients with severe dysplasia or cancer of the colon (1,12).

Double stapled anastomosis technique

In this procedure anal mucosectomy is omitted and the very distal low rectum is transected with a linear stapler. The ileal pouch-anal anastomosis is performed by a circular stapler. Compared to the more difficult and time-consuming handsewn pouch-anal anastomosis, the stapled technique is faster and straightforward. Thus the stapled technique has become the most favoured by surgeons. Beside its simplicity, it might also reduce anastomotic tension therefore lessening the need for mesenteric lengthening and the need for loop ileostomy. Another argument in favour of the stapled technique is the rela-

tively better outcome if septic complications occur (17). Nevertheless, the choice between both anastomosis techniques is one of the major points of controversy that still persists today, especially regarding the fate of retained at-risk mucosa (18-21). Incidence of polyps of the residual rectal mucosa following pouch surgery was higher after stapled anastomosis (28% vs. 14%) as shown by Remzi *et al.* though not reaching statistical significance (22). In a review by Chambers and Mortensen, the benefit in terms of disease control in the mucosectomy group compared to the double stapled technique did not reach statistical significance (23). Functional outcome on the other hand was not as good when mucosectomy was performed. The authors concluded that mucosectomy should be indicated only for sub-groups of patients, especially those with severe polyposis and important rectal adenomatosis. This conclusion was mostly based on the meta-analysis by Lovegrove *et al.* based amongst others on six RCT's (24). The study analysed results for FAP and ulcerative colitis (UC). The author admits that both groups should be analysed separately and that there is a lack of power and follow-up to ascertain the results in terms of disease control. Schluender *et al.*, on the other hand, found no functional difference in the two groups (25). If necessary, because of polyps developing on the retained rectal cuff, secondary completion mucosectomy has been described and is feasible by a perineal approach without compromising the pouch (21). No strong conclusions can be drawn concerning the choice between the two techniques, but it can be emphasized that mucosectomy, if performed, has to be conducted with caution as functional impairment is linked to manoeuvres in the anal canal or inadvertent internal sphincter injury.

Omission of loop ileostomy

To protect the ileal pouch-anal anastomosis, a temporary diverting loop ileostomy was part of the original procedure. Nowadays, ileostomy is often omitted since many reports have been demonstrating the absence of additional adverse events especially no increase in the incidence of pelvic sepsis linked to anastomotic or pouch suture line fistula. Metcalf *et al.* were the first to demonstrate no difference in morbidity without diverting stoma (26). Other groups confirmed the feasibility and the safety of this approach. Thus the legitimacy of this temporary stoma is questionable, even more taking into account the specific complications of such a stoma like high ileostomy output in 20-33% of cases, stomal retraction, parastomal hernia, prolapse, fistula, and abscess and skin irritation. Ileostomy also exposes to higher risks of small bowel occlusion and entails the necessity of a second procedure to close the stoma with its specific morbidity (1).

For the FAP population, i.e. young patients with good general status, one of the only reasons to perform a temporary loop ileostomy would be anastomotic tension as this independent factor has been shown by Heuschen *et*

al. to increase septic related pouch complications (27). As already discussed, anastomotic tension can be reduced by mesenteric lengthening techniques as well as the double stapling technique.

Laparoscopic approach

The majority of the patients undergoing IPAA for FAP are young patients in good general condition. Surgical prophylaxis should remain as less invasive as possible. A laparoscopic approach offers advantages in terms of, not only cosmetics, but also abdominal wall trauma. It is becoming more and more the approach of choice even for complex procedures such as IPAA. It has been shown that the procedure can be performed laparoscopically in all its details especially for the mesenteric lengthening techniques, loop ileostomy omission and pelvic nerve preservation. The feasibility is sustained by numerous studies (1,28-43). According to Larson *et al.* operative time tends to be longer in the laparoscopic approach which is compensated by a shorter median time to ileostomy output and to regular diet, length of stay and decreased IV narcotic use although postoperative morbidity was equivalent (28). These findings are confirmed by Michelassi *et al.*: laparoscopic patients had faster return of flatus ($p = 0.008$), faster assumption of a liquid diet ($p < 0.001$), and less blood loss ($p = 0.026$). While complications were similar, the incidence of incisional hernias was lower in the laparoscopic group ($p = 0.011$) (44). When focused on one-step laparoscopic procedure, except for a longer operative time, no difference in outcome are reported (45).

Functional outcome of laparoscopic IPAA remains comparable to the open approach as well as quality of life (33,36,42,46). More recently, Indar *et al.* confirmed the suspected advantage of laparoscopic IPAA, namely less adhesion development which could lead to less occlusive accidents and might preserve female fertility (47).

Antolovic *et al.* have published the study protocol of an upcoming RCT comparing the open with the laparoscopic approach. This two armed, single centre, expertise based, preoperatively randomized, patient blinded study is designed as a two-group parallel superiority study. The primary objective is to investigate intra-operative blood loss and the need for blood transfusions. Additionally a set of surgical and nonsurgical parameters related to the operation will be analysed as secondary objectives. These will include operative time, complications, postoperative pain, lung function, postoperative length of hospital stay, a cosmetic score and pre-and postoperative quality of life (48). Results are still awaited. Finally, new technical advances that should reduce even more the surgical trauma become available. The first case of single port laparoscopic (SILS) total colectomy with IPAA has been described by the Cleveland clinic team showing that this complex surgery is feasible with a virtual scarless abdomen (49).

Morbidity – Mortality

IPAA has a low operative mortality ranging from 0% to 1%. (1). On the other hand, complications occur between 10% and 25%. The meta-analysis by Lovegrove *et al.*, compared outcome of IPAA in FAP and UC in more than five thousand patients, 782 out of those were FAP patients (24). The anastomotic leakage rate was 1,3% and anastomotic stricture occurred in 9,7% of the cases. Heuschen *et al.* reported that the cumulative risk of developing septic complications is less than 10% (27). More recently, the group from the Cleveland Clinic reported a septic complication rate of 6,2%. The vast majority of their cohort ($n = 3233$ patients) included UC patients and only 5,8% of FAP patients. Nevertheless, on multivariate analysis, body mass index greater than 30, final pathologic diagnosis of ulcerative/indeterminate colitis or Crohn's disease, intraoperative and postoperative transfusions, and surgeon were all found to be independent factors associated with septic complications after restorative proctocolectomy (50). Small bowel occlusion was reported in 17,7% of the patients. Cumulative risk of SBO is estimated at 31,4% at 10 years after IPAA although most of the patients in this series were operated for ulcerative colitis (51). SBO risk is increased by the construction of an ileostomy and might be lowered by a laparoscopic approach as some have shown less pelvic and peri-stomal adhesions after laparoscopic IPAA (1,47). Pouch failure, defined by the need to remove the pouch, usually results of pelvic sepsis, poor function or pouchitis. The incidence of pouch failure is evaluated between 0-15% (2). Surgical revision after failure of an IPAA is possible in most patients and yields an acceptable level of bowel function in two-thirds of the patients (52,53). Perineal procedures should be attempted first, if appropriate, although in some cases abdominal procedures will be the best initial reoperative choice. Multiple attempts can be justified and do not necessarily lead to pouch excision (54). Abdominal salvage surgery is associated with a failure rate of 15% to 21,4%, especially when the procedure is carried out for septic than for non-septic indications (53).

Functional results

Functional outcome of IPAA have been widely described and are dramatically better in FAP patients compared to UC patients. Unfortunately, in most of the series functional results after IPAA in FAP and UC patients are mixed. In FAP, stool frequency ranges from 4 to 6 per 24 hours, with 0-1 night-time faecal elimination. A normal daytime continence can be expected in 80 to 95% of the patients, whereas faecal spotting during the night is reported in up to one third of the patients and night-time soiling in 1% of patients (55-57). Significantly more incontinence was found in the sub-group of patients older than 50 years at time of surgery (58-60). Furthermore, functional results of secondary proctectomy

and IPAA after ileo-rectal anastomosis remain comparable to those after primary IPAA (61).

Quality of life

Quality of life after IPAA for FAP is good, though life spend with a temporary ileostomy is experienced with poor quality of life (62). Again, quality of life is radically better after IPAA for FAP compared to UC. Large series report more than 98% of patients satisfied with minor impact on daily activities including social, home, travel and sports (1). No differences were found compared to a normative population by Hassan *et al.* (63). Compared to IRA, the same author found no differences except for health perception and energy/fatigue subscales favouring IPAA. The authors argue that fear of cancer in the remnant rectum could be the reason for this significant difference. When comparing open RCP with a laparoscopic approach, the most important finding is that LRP results in a superior body image and cosmesis, especially for women (36).

Pouchitis

In contrast to the high incidence of pouchitis (15-50%) in patients operated for UC, pouchitis after IPAA seems to be rare in patients with FAP (0-11%) (1,64,65). The reason for this very low incidence of pouchitis in FAP is still unknown (7). It has therefore been suggested that the likely aetiology is related somehow to that of UC, and whether pouchitis actually occurs after IPAA for FAP is debatable (1,7). No hard data is available until now to conclude (2,66).

Pouch adenomas and adenocarcinoma

After IPAA, FAP patients are exposed to the development of polyps in the pouch. Estimated cumulative incidence at 5, 10 and 15 years are around 7%, 35% and 75% respectively (67). Hand-sewn anastomosis with mucosectomy considerably lowers the risk of polyp development compared to double stapled anastomosis (2). Groves *et al.* reported a significant association between increasing age and the developing of pouch adenomas as well as the length of follow-up (68). On the other hand no relationship between the severity of the polyposis and the risk of pouch adenomas could be found. Another prospective study found that 22,8% of the patients were bearing pouch adenomas after 5 years of follow-up (69).

Most of the reports have shown no severe dysplasia of the detected pouch polyps (70). Implication of this life-long risk of developing adenomas in the pouch is a regular surveillance by pouchoscopy. The underlying risk of malignant transformation of pouch polyps is not precisely known. To date 13 cases of adenocarcinoma arising in the pouch have been reported in FAP patients (71-76). This severe complication remains anecdotal though some suspect that the prevalence might rise with increasing

follow-up. Preventing the adenoma-carcinoma transformation is the aim of Sulindac treatment. While chemoprevention of colorectal adenomas in FAP using Sulindac works, at least partially and for a limited time, its effectiveness in the ileal pouch has not been systematically studied (77). No formal guidelines have been published concerning the type and the frequency of surveillance, although most of the authors suggest a yearly basis.

Sexuality, Fertility, Pregnancy and Childbirth

Recent reports did not find significant impairment on sexual function after IPAA except slight worsening of sexual function in women compared to a normal population (33,78). Fertility in women is of more concern after IPAA. A dramatic decrease in fertility has been reported in UC patients with loss of fertility of about 80%. Fortunately in FAP, Olsen *et al.* have published less bad, but still worrying results (79). To date, the mechanisms of this fecundity drop are largely accepted to rely on pelvic adhesions. Spanos *et al.* reviewed the literature dealing with female fecundity and colorectal cancer surgery and came to the conclusion that pelvic surgery in itself is a factor affecting female fertility (80). It seems plausible that it is the extent of dissection and the location right down to the pelvic floor of the IPAA surgery that causes such a severe reduction in fertility by partial or complete occlusion of the Fallopian tubes, altering the normal tubo-ovarian relationship necessary for ovum capture and transport (1). Regarding this argument, the close rectal wall dissection down to the pelvic floor, initially recommended for pelvic nerve preservation, allows also an almost complete peritoneal surface preservation especially in both "tubo-ovarian para-rectal fossa". On the other hand, the hypothesis claiming that laparoscopic surgery might positively affect this outcome, by lessening adhesion development, has been suggested by Indar *et al.* (47). In their study, the abdominal cavity of FAP patients, having undergone IPAA with protective ileostomy, was inspected for adhesions to the anterior abdominal wall and to the gynaecologic organs, during the loop ileostomy closure, through the stomal orifice. They came to the conclusion that the laparoscopic approach leads to significantly less adhesions which could imply better results on fertility. Nevertheless, young female patients should always be informed of these risks on fertility before surgery keeping in mind that the above described technical preventive measures could lessen fertility impairment.

IPAA operation does not jeopardise pregnancy and childbirth. Caesarian section does certainly not have to be recommended systematically but should follow the classical obstetrical criteria (1).

Decision-making strategy

Since the introduction of the IPAA operation for FAP in the late 1970s, the choice between IPAA and IRA with

FAP still remains controversial. Therefore, some attempts have been made to design the best surgical strategy for each FAP patient, taking multiple parameters into account, as listed in Table 1. The role of ileorectal anastomosis is discussed by A.M. Wolthuis *et al.* in the same issue. The decision relies not only on the rectal cancer-risk, but also on the potential ability to perform a secondary proctectomy after an IRA when rectal malignancy is diagnosed. We have shown that conversion of IRA to IPAA was technically impossible in 3 out of 29 FAP patients (10%) with IRA because of unexpected pelvic desmoid tumours (DT) (82,83).

It could be very attractive to use the APC molecular genetic testing as an aid in decision-making with respect to the type of surgical procedure, i.e. total colectomy with IRA vs. total proctocolectomy with IPAA. This strategy has been proposed for at least three subsets of FAP phenotypes :

- Patients with severe polyposis.
- Patients with attenuated polyposis.
- Patients with DT.

1. Severe polyposis phenotype

From the beginning of the search for genotype phenotype correlations, it has been found that APC germline mutations between codons 1250 and 1465 are associated with a profuse phenotype in which > 1000 colorectal polyps develop (1,7). There is a particular ‘hot spot’ mutation at codon 1309 that always causes severe disease, usually with thousands of polyps. Vasen *et al.* (84)

first suggested that the results of DNA testing in relation to the phenotypic expression in the patient and family could be helpful in surgical decision-making. They found that these severely affected patients have such a high risk of rectal cancer after IRA that subsequent proctectomy is almost routine and initial IPAA is to be preferred (Table 1) (1). However, it has been demonstrated both inter- and intrafamilial variations of polyp density in patients with mutations in codon 1309. In fact, a wide phenotypic variability has been observed, not only within different kindred carrying the same APC mutation but also within kindred. Furthermore, increased risk of early colorectal cancer associated with two areas before codon 1250 : 514-713 and 976-1067 have also been described (1). This means that one should act with caution before setting strict surgical guidelines based on mutational analysis.

2. Attenuated polyposis phenotype

APC germline mutations occurring in the 5' end of the gene (particularly exons 3 and 4) are associated with far fewer polyps and a delayed onset of cancer (1,7). This relatively mild form of FAP, characterised by an extremely wide intrafamilial variability, has been designated as ‘attenuated’ adenomatous polyposis coli (AAPC) or ‘attenuated’ familial adenomatous polyposis (AFAP). Evidence indicating a much lower rate of CRC in AFAP families than in classic FAP families has been reported in recent years. In all AFAP kindred, a predominance of right-sided colorectal adenomas and rectal polyps sparing was observed. Accordingly, if surgery for

Table 1. – Prophylactic surgery for FAP patients in our practice (1)

Factor	IRA	IPAA
Indication for surgery	< 20 rectal polyps AFAP	Most of FAP patients > 1000 colonic polyps Cancer anywhere in the large bowel > 20 rectal polyps Severe dysplastic rectal adenoma Large(> 3 cm) rectal adenoma Resectable rectal cancer Desmoid family history
Sex	Female before procreation	Female after procreation
Laparoscopic surgery	Yes	Yes
Age at surgery	Within 2 years of phenotypic expression and molecular diagnosis	Within 2 years of phenotypic expression and molecular diagnosis
Mortality Rate	Very low	Very low
Morbidity Rate	Low	High
Functional outcome		
Early	Good	Average
Late	Good	Good
QOL	Good	Good
Follow-up	Rectal endoscopy : 2x/year	Pouch endoscopy : 1x/year, 2x/year if polyps

AFAP attenuated familial adenomatous polyposis, FAP familial adenomatous polyposis, IPAA restorative proctocolectomy with ileal pouch-anal anastomosis and mucosectomy, IRA total colectomy with ileo-rectal anastomosis, QOL quality of life.

otherwise intractable polyps is indicated, total colectomy with IRA is recommended (Table 1) (1,7,81). Because the natural history of AFAP is not well documented yet, and therefore the exact risk of colorectal cancer remains unknown, caution should again be advocated when choosing IRA, and lifelong rectal monitoring should be mandatory (1,7,81). Even the most recent and expert group report remains cautious in their conclusion and states that “Genetic information never will substitute for major parameters such as the number of colonic adenomas and the rectal phenotype in determining the surgical choice, but it may add useful information” (85). The same authors distinguished an intermediate genotype risk group that has a 40% risk of reoperation after IRA and therefore recommend to base surgical decision on a combination of clinical, genetic data as well as patient’s preferences.

3. Desmoid tumours (DT)

Specific 3’-APC germline mutations (distal to codon 1399), associated with a high risk of DT are frequently linked to a lower density of colonic polyposis and have a later and reduced cancer risk. Moreover, there is evidence that surgical trauma can precipitate the formation of DT, although the underlying mechanism is not clear. Therefore, it has been advised for such patients – i.e. mutation after codon 1400 and a strong family history of desmoids – to postpone elective colectomy and to manage the colon by close monitoring and chemoprophylaxis until surgery is required (1). However, once colectomy is required, we advocate avoiding repeated surgery and therefore performing the more definitive operation, namely IPAA, directly without a preliminary stage of IRA (Table 1) (1,7,8,88). More than a decade after the discovery of the APC gene and identification of its mutations, it appears that the genotype-phenotype correlations are far more complex than expected. The legitimate hope that molecular genetic analysis would guide our surgical practice has to be tempered, and more relevant clinical data should be provided to support it. A recent report could not detect such clinical risk factors (86). Whereas clinical inferences from APC mutational analysis seem to be justified, these have, at least in our practice, not yet been completely integrated into standard management guidelines for decision-making between IRA and IPAA (7).

Conclusion

IPAA offers the best available prophylaxis in FAP patients. Technically demanding, the operation carries almost no mortality and acceptable morbidity. Long term results in terms of function and quality of life are comparable to those of the major alternative, namely IRA. Nevertheless, the debate between IRA and IPAA is still open. When proposing IPAA, young female patients should be informed of the potential harm on fertility and

all patients should be aware about the necessity of long-term follow-up of the pouch.

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