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Restorative proctocolectomy and ileal pouch-anal anastomosis for familial adenomatous polyposis revisited

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Abstract

Since restorative proctocolectomy (RPC) with ileal-pouch anal anastomosis (IPAA) removes the entire diseased mucosa, it has become firmly established as the standard operative procedure of choice for familial adenomatous polyposis (FAP). Many technical controversies still persist, such as mesenteric lengthening techniques, close rectal wall proctectomy, endoanal mucosectomy vs. double stapled anastomosis, loop ileostomy omission and a laparoscopic approach. Despite the complexity of the operation, IPAA is safe (mortality: 0.5–1%), it carries an acceptable risk of non-life-threatening complications (10–25%), and it achieves good long-term functional outcome with excellent patient satisfaction (over 95%). In contrast to the high incidence in patients operated for ulcerative colitis (UC) (15–20%), the occurrence of pouchitis after IPAA seems to be rare in FAP patients (0–11%). Even after IPAA, FAP patients are still at risk of developing adenomas (and occasional adenocarcinomas), either in the anal canal (10–31%) or in the ileal pouch itself (8–62%), thus requiring lifelong endoscopic monitoring. IPAA operation does not jeopardise pregnancy and childbirth, but it does impair female fecundity and has a low risk of impairment of erection and ejaculation in young males. The latter can almost completely be avoided by a careful ''close rectal wall'' proctectomy technique. Some argue that low risk patients (e.g. <5 rectal polyps) can be identified where ileorectal anastomosis (IRA) might be reasonable. We feel that the risk of rectal cancer after IRA means that IPAA should be recommended for the vast majority of FAP patients. We accept that in some very selected cases, based on clinical and genetics data (and perhaps influenced by patient choice regarding female fecundity), a stepwise surgical strategy with a primary IPA followed at a later age by a secondary proctectomy with IPAA could be proposed.

Introduction

FAP is an inherited, autosomal dominant syndrome associated with a germline mutation of the *APC* gene with complete penetrance [1–5], characterised by the development of from 100 to several thousand colorectal adenomas at a young age, inevitably resulting in colorectal cancer [1–5].

We argue that since every single epithelial cell of the colon and rectum carries the APC mutation, potentially leading to the adenoma–carcinoma sequence, any prophylactic surgery should ideally remove the entire diseased mucosa from the ileocaecal junction up to the dentate line at the anal verge [4].

A pouch with mucosectomy and endoanal anastomosis (IPAA: Figure 1) achieves this goal and for many has become firmly established as the standard operative procedure and the method of choice for classic FAP [4–9].

Since its introduction by A. Parks and R.J. Nicholls in 1976 [10], more than 1000 papers have been published about IPAA and there have been numerous attempts to reach a consensus, yet many controversies still persist. We have reviewed these current 'hot topics' for debate.

Standard IPAA procedure

A standard technique has been described and widely used for many years by many surgeons. It includes several important steps:

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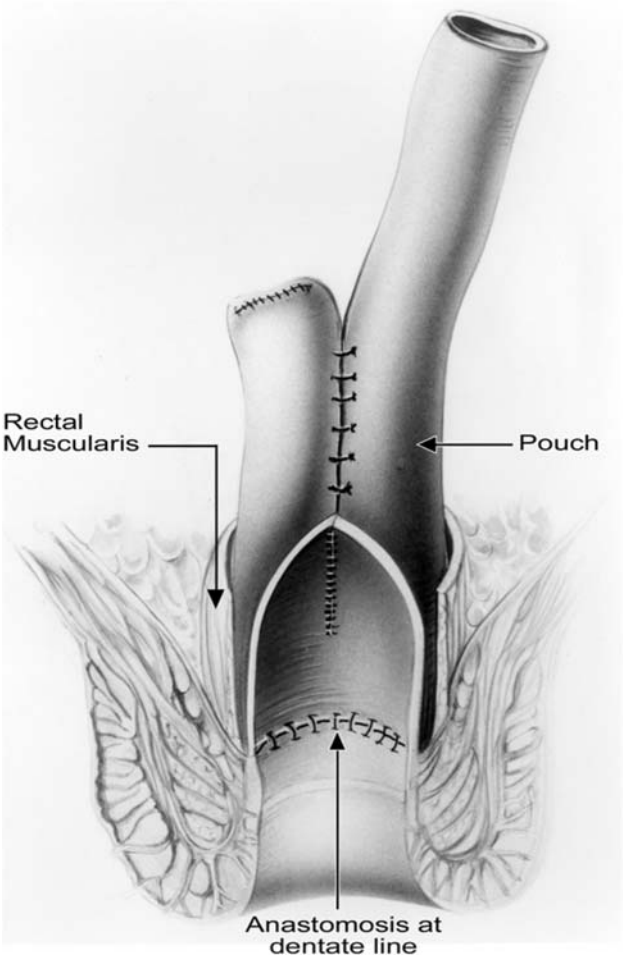

Figure 1. The colon and the rectum have been completely removed, and the distal rectal mucosa has been excised to the dentate line in the anal canal, leaving all the pelvic floor muscles intact and allowing preservation of normal anorectal function. An ileal J-shaped reservoir has been constructed. The apex of this ileal J-pouch is anastomosed down to the dentate line at the anal verge.

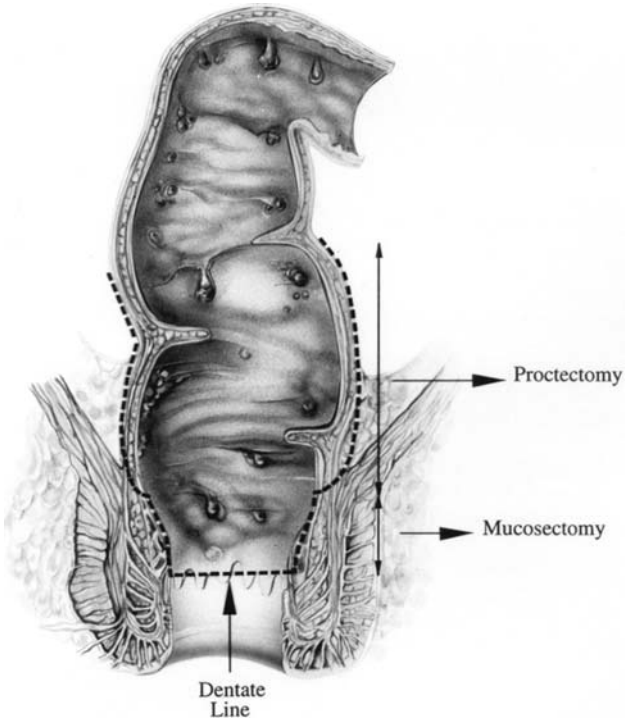

Figure 2. With respect to rectal excision, the aim is to provide a bloodless dissection and at the same time avoid injury to other pelvic structures, particularly autonomic nerves. The perimuscular dissection of the rectum is indicated by dashed lines. The level of division of the gut tube is at the anorectal junction, i.e., the upper border of the levator ani muscles. Further down, the dashed lines represent the plane for endorectal mucosectomy to the dentate line with strict preservation of the internal sphincter. This last step ensures complete removal of all at-risk or diseased epithelium.

– total colectomy,
– proctectomy,
– endoanal mucosectomy,
– ileal pouch-anal anastomosis,
– diverting ileostomy [4, 6, 11–16].

Except for the patients whose polyposis is complicated by colonic or rectal cancer, colonic 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 (Figure 2). Obviously, for rectal cancer, complete removal of the mesorectum to the level of the levator ani has to be performed [17].

After removal of the surgical 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 (Figure 3b).

The type of ileal reservoir mostly used is J-shaped, as originally described by Utsunomiya [18]. 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 [4, 6].

IPAA itself is a hand-sewn anastomosis performed at the dentate line level through a perineal approach (Figures 1 and 3a).

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 [4, 5, 11, 16].

Technically, RCP with IPAA is a rather complex and demanding procedure requiring a steep learning curve [4, 5, 16, 19, 20]. Possible strategies for reduction in the steepness of the learning curve include formal training courses in IPAA surgery, close intra-operative supervision and monitoring by expert practitioners, and assistance from other well-trained staff [19, 20]. Continued technical advances and greater surgeon experience can only further improve function, outcome and patient satisfaction [16].

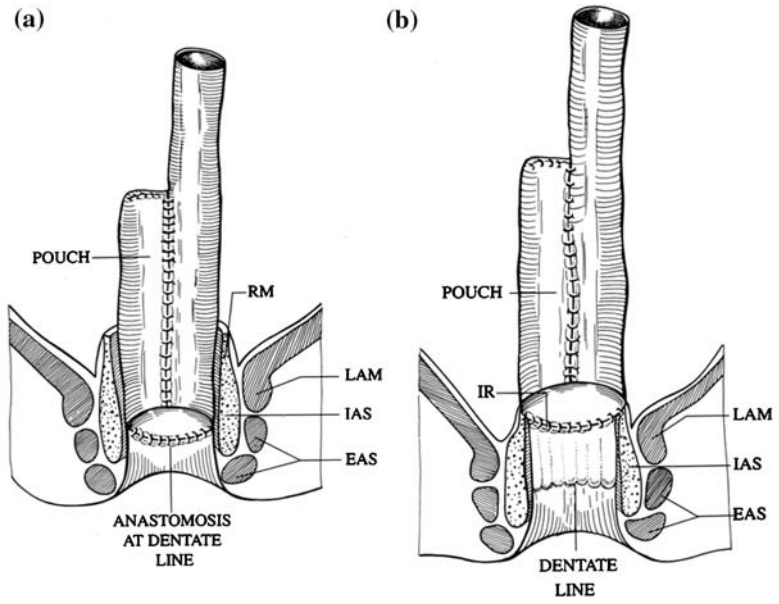

Figure 3. (a) Removal of the epithelium of the transitional zone by mucosectomy. An ileal pouch-anal anastomosis is performed at the level of the dentate line. EAS = external anal sphincter, IAS = internal anal sphincter. (b) Preservation of the anal transitional zone mucosa. The anastomosis performed is, in fact, a very low ileo-rectostomy. LAM = levator ani muscle, RM = rectal muscle, IR = ileorectal anastomosis.

Technical improvements, challenges and controversies

Since its original description about 30 years ago [10], the IPAA procedure has been modified in an attempt to obtain more technical simplicity together with better functional results, lower morbidity and mortality, and yet provide cure of the disease [20–24]. We believe that the current 'hot topics' still for debate are: optimising techniques of mesenteric lengthening; pelvic nerve preservation by close rectal dissection; transanal mucosectomy with hand-sewn anastomosis vs. the double-stapled technique; the use or omission of a diverting ileostomy; and the emerging role of laparoscopy.

Optimizing mesenteric lengthening

The aim of all mesenteric lengthening techniques is to achieve a tension-free IPAA in order to avoid postoperative anastomotic problems and thus the need for a temporary diverting ileostomy. In fact, by means of both univariate and multivariate analysis, Heuschen et al. [25] have identified anastomotic tension as a significant risk factor for pouch-related septic complications in patients with FAP.

Many lengthening techniques have been described [26, 27]. Division of the ileocaecal artery has often been presented as the safest and most effective method for obtaining maximum length and has been used for many years in our unit [5, 12, 14, 28, 29]. More recently, we have systematically preserved the marginal arcade of the right colon with its blood supply from the middle colic artery as described by Goes et al. [30] (Figure 4) [14]. With this manoeuvre, both the ileocolic artery and even distal superior mesenteric artery may safely be divided under the control of mesenteric transillumination, allowing for significant extra mobilisation of the apex of the pouch and for a tension-free IPAA in all cases with omission of a loop ileostomy. Since none of these patients experienced an anastomotic leak, we tend to use this procedure routinely [14].

The main inconvenience is that right colic arcade preservation is time-consuming and might potentially lead to more ileal pouch intussusception or prolapse, otherwise already described after IPAA [31, 32]. Another drawback is an increased risk of pouch ischemia [14, 31]. Right colic arcade preservation is of course precluded in FAP patients with severe dysplasia or cancer of the colon. Further investigation and clinical evaluation are therefore required to settle this issue of mesenteric lengthening.

Proctectomy

With respect to the technique of proctectomy, the aim is to provide a bloodless dissection and at the same time avoid injury to important pelvic structures. Since FAP patients are usually young, healthy and asymptomatic, urinary or sexual complications, such as urinary retention, impotence or retrograde ejaculation, are not acceptable. Particular attention should therefore be given to avoid injury to presacral nerves (sympathetic plexus) and to the nervi erigentes (parasympathetic plexus).

For rectal dissection, the most widely used, easiest and fastest technique is to perform a total mesorectal excision (TME) or 'TME-like' operation with, of course, pelvic nerve preservation [15, 16]. Posteriorly, the

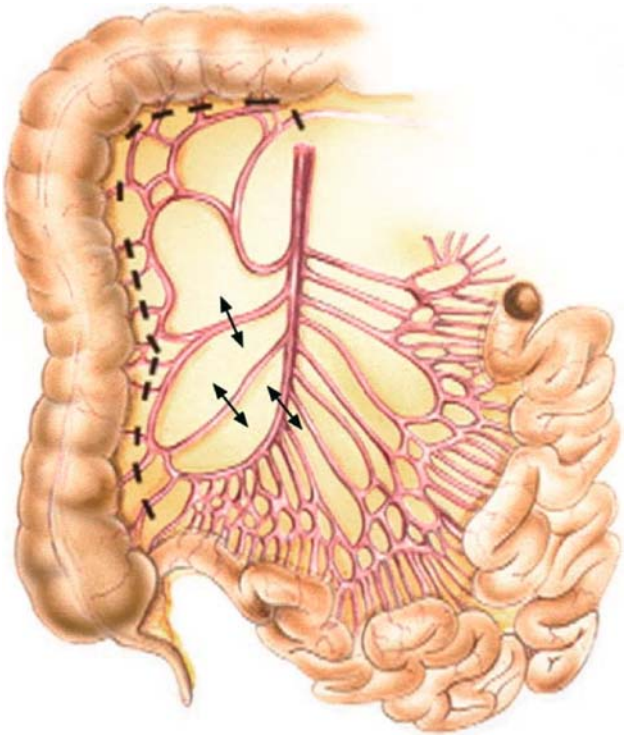

Figure 4. While performing the right colectomy, the dissection is carried out close to the colon serosa (dotted line) in order to preserve its marginal arcade with its blood supply from the middle colic artery. Further division of the right colic, the ileocolic and the distal superior mesenteric arteries (↔) will allow an optimal mesenteric lengthening.

dissection is carried out to the pelvic floor in the loose areolar tissue in the presacral space. The lateral rectal ligaments are divided as close as possible to the rectal wall. Anteriorly, the dissection is undertaken posterior to Denonvilliers' fascia to the inferior border of the prostate in male patients, and in the rectovaginal space to the perineal body in female patients.

But even with this careful 'nerve-sparing technique', male as well as female sexual dysfunction is still reported [33]. Therefore, from the beginning of the IPAA experience, some centres have chosen to routinely dissect the rectum in the perimuscular plane as described by Lee and Dowling [34]. This allows better and almost absolute safeguarding of autonomic nerves to pelvic organs [6, 12, 35–37].

At the level of the upper part of the mesorectum, the superior haemorrhoidal vessels are preserved, and dissection is carried out up to the rectal wall. The muscular fibres of the rectal wall are progressively and circumferentially freed of all their vascular and fatty attachments straight down to the pelvic floor, leaving all the perirectal fat behind in the pelvis. This particular step of the RPC can be very tedious, as well as time-consuming, especially in males with a narrow pelvis and in obese patients. However, in St-Antoine's experience [6] and in our own [14], the use of this rigorous technique of rectal dissection has decreased the risk of urinary and sexual dysfunction to virtually nil.

In obese patients, the perirectal fat left behind could adversely affect the IPAA functional results by pouch compliance restriction. To date we have not encountered this potential problem but keep an open mind as to the possibility.

In the case of FAP patients with severe rectal dysplasia or cancer, such a 'close-rectal wall' technique is contraindicated and classic TME should be performed [17, 38].

Endoanal mucosectomy vs. double-stapled anastomosis

Some surgeons have abandoned the step of endoanal mucosectomy in order to simplify the procedure and improve functional outcome [23, 39–45], preferring instead a technique of double stapling of the anastomosis, transection with a linear stapler of the very distal rectum or proximal anal canal, thereby avoiding a mucosectomy, and anastomosing the pouch by means of a circular end-to-end stapler (Figure 3b) [23].

This stapling technique has permitted relatively effortless and faster pouch construction and anastomosis [23, 34, 46–48], because of which it is preferred by the overwhelming majority of surgeons [39, 49].

On the other hand, endoanal mucosectomy and hand-sewn pouch-anal anastomosis at the level of the dentate line (Figure 3a), are more tedious, time-consuming and technically demanding.

The complexity of hand-sewn anastomosis was reflected in a lack of a true learning curve among senior surgeons, whereas a shorter learning curve was evident for stapled IPAA [20, 50].

The choice between both anastomosis techniques is one of the major points of controversy that still persists today, especially regarding technical issues, morbidity, functional outcome and the fate of retained at-risk mucosa [40, 42, 51, 52].

Technical issues

- Double-stapling is obviously simpler and faster as it avoids the tedious and critical step of endoanal mucosectomy, which is technically more challenging and requires great surgical skill and experience [4, 46, 51].
- Tension at the ileoanal anastomosis is usually decreased after stapled rather than after sutured anastomosis. This could theoretically make the stapled anastomosis safer, avoiding the need for mesenteric lengthening techniques and protective ileostomy [53, 54]. But since some patients have a short mesentery, and since the right bordering arch is not always available for mesenteric lengthening, it follows, therefore, that the surgeon should be competent at performing either technique of anastomosis [54].
- A very likely pitfall of the double-stapled IPAA is the difficulty of achieving an anastomosis at the anorectal junction in fat, male patients with a narrow pelvis, and thus the prospect of leaving a rectal

stump longer than desired [22, 42, 54, 55]. Moreover, Thompson-Fawcett et al. [56] have demonstrated that the anal transitional zone (ATZ) is shorter than most people have recognised, and that after double-stapling restorative proctocolectomy there remains anyway a 1.5–2.0 cm cuff of diseased columnar epithelium [39, 57].

- It can be anticipated that with laparoscopic surgery it will be even more difficult to reach the anorectal junction for an accurate transection with the linear endo-stapler, thus potentially leaving even more rectal stump. This will of course have to be evaluated in the future, but at the same time, we recommend systematic use of mucosectomy with hand-sewn anastomosis when using a laparoscopic approach [14].

Morbidity

Some authors have suggested increased complication rates after mucosectomy [44, 46, 50, 58]. But none of the three randomized controlled trials comparing stapled vs. manual anastomosis showed a significant difference in complications related to the IPAA technique [22, 53, 59], and it has to be underlined that the complication rate is very much related to the surgeon's experience [50]. In fact, low rates of pelvic sepsis, anastomotic leak and remote anastomotic stricture can also be achieved with careful mucosectomy, and minimal anal stretching [4, 6, 12, 30].

Functional outcome

In several retrospective studies, it has been shown that patients with stapled IPAA had a better manometric and subjective functional outcome, especially at night-time, than patients who underwent mucosectomy [16, 23, 41, 44, 50, 60–70].

It is self-evident that the more the anal canal is manoeuvred and stretched, the more likely there will be an adverse impact on sphincter control. Furthermore, it has been speculated that continence could be improved by preserving ATZ, which is thought to be important because it contains nerve endings that differentiate solid and liquid stools from gas [51, 64, 71–75]. Care must also be taken for the extent of anorectal smooth muscle resected at the time of mucosal proctectomy in order to preserve postoperative bowel and anal sphincter function [4, 16, 64, 76, 77].

Against this, the three randomised trials already mentioned that compare IPAA with mucosectomy with ileal-pouch distal rectal anastomosis without mucosectomy have failed to demonstrate any functional advantage of the technique retaining transitional zone mucosa [22, 53, 59], although in one study Reilly et al. [59] found higher resting pressure and less night-time incontinence in the stapled group.

Finally, in a large retrospective review of 119 FAP patients, Remzi et al. [66] found similar quality of life and overall satisfaction in both groups.

Retained mucosa

This important question focuses on the completeness of excision of the disease itself. There is always a trade-off between neoplasia control and functional results [17, 39]. Since every epithelial cell carries a germline mutation of the *APC* gene, every island of mucosa left behind is at risk of dysplastic or neoplastic changes and we argue that this should therefore ideally be removed down to the dentate line [4, 40]. The risk of cancerous change of residual mucosa remains but is nearly impossible to predict and will be discussed later [4, 16, 39, 44, 78].

An obvious shortcoming of the double-stapled pouch is the retention of some anorectal mucosa, which harbours the potential to undergo carcinomatous changes [4, 40, 49, 79–81], confirmed by the demonstration of polyps or dysplasia in surgical doughnuts or biopsies taken just distal to the stapled IPAA for FAP [49, 82]. In a short series of 12 cases of FAP patients who underwent IPAA in our unit, examination of the mucosectomy specimen showed that micropolyps were already present in all cases with low-grade dysplasia [77]. Therefore, we believe that complete removal of the ATZ ensures maximal reduction of risk of future disease such as dysplasia or carcinoma [40].

But even with mucosectomy, it appears that islands of diseased epithelium can be left behind in up to 14% of cases and these can be obscured by the pelvic pouch, making adequate follow-up impossible [40, 52, 66, 79, 83–91]. This raises the question whether complete mucosectomy is even possible in practical terms [87]. Notwithstanding, endoanal mucosal stripping should be performed very carefully and as completely as possible, which requires high technical skill [4–6, 12].

Conclusions

Although stapling techniques simplify pouch construction, some (including ourselves) believe that stapled IPAA is unacceptable in FAP patients because the mucosa of the ATZ is left intact and a hand-sewn IPAA should therefore be obligatory [4, 6, 8, 12, 16, 40, 42, 92]. It is incumbent on surgeons who leave the ATZ to inform their patients of the lack of long-term knowledge of the natural history of these mucosal remains, therefore requiring lifelong monitoring.

Loop ileostomy omission

The original procedure of IPAA included a routinely performed diverting loop ileostomy to facilitate the pouch suture line and the anastomosis itself healing, and to minimise the risk of leakage and pelvic sepsis [4, 6, 12, 16, 51, 93, 94]. However, many reports have now revealed that there are complications related to the loop ileostomy itself [4, 44, 63, 94–119].

The disadvantages of a diverting stoma are well recognised and include high ileostomy output in 20–33% of cases, stomal retraction, parastomal hernia, prolapse, fistula, and abscess and skin irritation [44, 113,

119]. The construction of an ileostomy may also increase the risk of small bowel obstruction up to 15–21% [44, 106, 109, 113, 119–121]. Moreover, patients treated with IPAA as a two-stage procedure have to undergo another operation for reversal of the ileostomy, entailing an additional risk of perioperative complications, such as anastomotic leak from the ileostomy closure site in 2.5–7.5%, considerable time off work for the patient, and markedly higher overall costs for surgical therapy [25, 44, 74, 108, 110, 113, 119, 121, 122].

In 1986, Metcalf et al. [109] from the Mayo Clinic Group first challenged the need for a diverting loop ileostomy in carefully selected patients operated by surgeons experienced with IPAA. Since then, several other studies have reported that septic complications and functional results were the same after one- or two-stage procedures [14, 25, 63, 95, 98, 103, 110, 113, 119, 122–126]. Moreover, there are also fewer episodes of intestinal obstruction, fewer instances of re-exploration and fewer total days in the hospital [95, 97, 102, 106, 109, 113, 119].

One possible inconvenience advocated after one-stage IPAA is that there may be transient incontinence due to stretching of the anorectal sphincters while performing the IPAA, and that an ileostomy could allow this to settle down [110]. The immediate postoperative course may also be more difficult for one-stage patients because the pouch must adapt to storing liquid stool [74]. This could account for the early postoperative results reported [96]. However, we have to admit that until now only one prospective randomised study has been published [97].

The generally proposed criteria that enter the decision process of whether to perform IPAA with or without ileostomy include general health status, adequate nutrition, advanced age, co-morbidity, elective procedure, a motivated patient, experienced surgeon, lack of intraoperative complications, good blood supply to the pouch, satisfactory anastomosis performance, and absolute lack of tension on the anastomosis [14, 25, 44, 74, 78, 95, 98, 109, 122].

In a large series of 212 IPAA for FAP, Heuscher et al. [25] have shown by multivariate analysis a significantly greater and independent risk factor for pouch-related septic complications in patients in whom the anastomosis was done under tension.

In our experience, the lack of tension on the anastomosis itself is also the most important key for protective ileostomy omission [14]. Every effort should be made to achieve pouch mobility by mesenteric lengthening, as already described. From this point of view, the double-stapled anastomosis technique could offer some advantages over mucosectomy and hand-sewn anastomosis by decreasing tension at the anastomosis performed at the top of the anal canal instead of the dentate line. This could make the stapled anastomosis safer than suture anastomosis, at least in theory [44, 63, 94, 101, 102]. However, since we recommend a mucosectomy with complete removal of all at-risk mucosa in FAP patients, we propose routinely using the preservation technique of the marginal arcade of the right colon in order to achieve a tension-free anastomosis at the dentate line with loop ileostomy omission in every case, without increased risk of anastomotic leak or pelvic sepsis [14].

As far as the above selection criteria are concerned, it is generally admitted that FAP patients are best suited for one-stage IPAA, since they are usually young, in good physical condition, symptom-free and their tissues have not been modified by inflammation or previous medication [113]. Therefore, with IPAA for FAP, the anastomosis is usually safe enough to allow routine consideration of the option of avoiding a temporary ileostomy [21].

Role of laparoscopy

For FAP, operations are prophylactic and this is appealing for a minimally invasive laparoscopic approach in often young and asymptomatic patients at a critical period in their social, academic and professional development [21, 127].

To date, there are about 50 reports on laparoscopic IPAA, including mainly UC rather than FAP patients. Of all these studies, only one is a prospective randomised comparative study related to hand-assisted laparoscopic pouch surgery [128].

Almost all papers concluded that the laparoscopic approach for RPC is feasible and safe, with the main advantage being a better cosmetic result [111, 119, 128–141]. The main drawbacks are increased operating time and a technical challenge requiring a steep learning curve [111, 119, 127, 130, 132, 137, 142–148]. The technique is still evolving and more time and experience are required to refine the procedure [119, 133, 149].

Due to the current lack of experience, the theoretic benefits compared with conventional operations, such as less pain, shortened postoperative ileus, reduced hospital stay and more rapid return to normal activities, are clearly not yet emerging for laparoscopic IPAA [111, 119, 128, 132, 142, 145, 146]. Moreover the functional outcome and quality of life of laparoscopic-assisted IPAA are not different from conventional IPAA [128, 129, 135].

In many centres, the laparoscopic approach systematically includes a Pfannenstiel incision for completion of the proctectomy as well as a double stapling anastomosis technique and a protective loop ileostomy [135, 138, 147, 150–152]. However some authors have proposed a safe ileostomy omission in selected laparoscopic cases [51, 132, 134].

Since 2001, in our Colorectal Surgery Unit we have prospectively used the laparoscopic approach in all consecutive patients. We have shown the feasibility of every technical option already described: preservation of the right arcade for mesenteric lengthening, close rectal wall proctectomy, endoanal mucosectomy with hand-sewn anastomosis and, finally, systematic omission of the protective loop ileostomy [112].

Another major potential advantage of the laparoscopic approach is decreased formation of postoperative adhesions. Since laparoscopic procedures result in a more gentle manipulation of tissues, less bleeding, and less contamination with foreign bodies than conventional surgery, there have been many claims that laparoscopy also reduces the incidence of postoperative adhesions [51, 153, 154].

Decreased postoperative adhesive disease could in turn reduce the rate of small-bowel obstruction after IPAA [51, 137, 145, 155–158]. Moreover, as will be discussed later, this could also influence fertility after IPAA in young women with FAP [159].

These theoretical advantages of laparoscopic RPC over conventional open surgery, although not yet evidence-based, constitute a serious trend and have a great chance of being confirmed by subsequent randomised trials. And, last but not least, since abdominal surgery precedes development of desmoid tumour (DT) in 68–83% of FAP patients, there may be a link between surgery and DT development [160–165]. However, there is no evidence to suggest that the extent or type of surgery influences DT development [161, 166–168]. Hence, it is not known whether minimally invasive techniques lower the risk of postoperative intraabdominal DT, but the concept is attractive [21, 127]. Finally, randomised studies are needed to define adequately the future role of the laparoscopic approach in RPC [51, 111, 130, 150]. However, preliminary reports are promising.

Mortality–morbidity

IPAA for FAP is a safe operation, since the reported mortality rate in large published series ranges from 0% to 1% [4, 6, 12, 44, 78, 169].

In contrast to mortality, overall morbidity complication rates remain significant and range from 10% to 25% for FAP patients, which is far better when compared with those patients who have undergone the operation for UC [4, 6, 12, 16, 33, 169, 170].

The most commonly encountered complications among patients undergoing IPAA are:

Small-bowel obstruction (SBO): 10–15% [4, 7, 12, 170]: The risk of SBO after IPAA is high, although most do not require surgical intervention.

In a large series of 1178 IPAA, including only 66 FAP patients, Mac Lean et al. [171] reported a cumulative risk of SBO of 31.4% and a need for surgery of 7.5% at 10 years.

In patients requiring laparotomy, the obstruction was most commonly due to pelvic adhesions (32%), followed by adhesions at the ileostomy closure site (21%). This increased risk of SBO related to construction of an ileostomy has already been suggested by others [44, 106, 109, 113, 121, 172, 173]. On the other hand, the laparoscopic approach for IPAA might reduce postoperative adhesion formation and hence the risk of SBO [51, 137, 145, 155–158].

Anastomotic leak and pelvic sepsis: 0–9% [4, 6, 12, 25, 78, 122, 170]: For FAP, the main risk factor for pouch-related septic complication is anastomotic tension when IPAA is performed [122].

Anastomotic stricture: 4–12% [4, 6, 12, 174]: Non-fibrotic strictures respond well to anal dilatation, whereas fibrotic strictures are more commonly associated with intraoperative or postoperative complications, often necessitating future surgical therapy.

Pouch failure requiring excision: 0–12% [4, 6, 12, 58, 78, 122, 170, 175, 176]: Pouch excision is associated with a high morbidity [177]. However, repeat surgery for pouch salvage is possible in the majority of patients in specialised centres, with an acceptable outcome [54, 55, 168, 176, 178, 179].

Despite the complexity of the operation, IPAA for patients with FAP is safe and entails an acceptable risk of complications. Although most of these complications are not life-threatening, they often necessitate re-hospitalisation and repeat surgery for the patient [5]. Fortunately, increased experience has significantly reduced the incidence of complications [180].

Functional results

Persons undergoing IPAA should be advised that, although the surgery will preserve faecal elimination via the anus, functional outcomes are not comparable with bowel elimination via an intact colon and rectum [15]. There are numerous descriptive reports focusing on functional outcomes following IPAA including both UC and FAP, with a majority of UC patients [33]. Stool frequency ranges from 4 to 6 stools/24 hours, with a frequency of night-time faecal elimination of 0–1 episode [4, 6, 12, 15, 30, 37, 78, 170, 181]. It may take upwards of 1 year for a patient to achieve reasonable stability with respect to bowel function and frequency. A 10-year follow-up found daytime and night-time stool elimination consistent with patterns established by the end of postoperative year 1.

Normal daytime faecal continence is found in 80–95% of patients, faecal spotting at night in 32–42% and faecal soiling at night in 1% [4, 6, 12, 15, 30, 37, 78, 170, 181]. Some argue that pouch function in FAP improves between the first and fifth year after the operation [181]. A long-term follow-up study in 1156 IPAA patients, including 37 FAP patients, showed that functional results do not deteriorate over time as regards continence [182].

Providing that the patients are highly motivated to accept the consequences of an IPAA (more frequent bowel movements, as well as the risk of soilage and incontinence), most of them are satisfied with the functional results [7].

Quality of life

There are numerous studies showing that patient satisfaction level is high after IPAA [4, 6, 37, 78,

183–192]. In the large Cleveland Clinic series, including both UC and FAP, overall long-term quality of life after IPAA was excellent [78, 182]. Quality of life was good to excellent in 99% of FAP patients [78]. Around 98% of patients would recommend the surgery to others. In a series of 187 FAP patients, the Mayo Clinic group showed that patients were satisfied with the outcome of the operation and the quality of life achieved. Daily activities ranging from social, home, travel, sports, and sexual activities were affected minimally, with only 2% of patients reporting adverse outcomes in these areas [7].

Some studies have suggested that better functional results in FAP were not equated with better quality of life, thus highlighting the influence of non-pouch-related factors on quality of life after ileal pouch formation [184–187]. In that respect, preoperative diagnosis (UC vs. FAP) has an impact on quality of life [184, 188–192]. In fact, overall satisfaction with respect to daily activities is higher in UC when compared with FAP. This difference is not surprising since patients with FAP are generally healthy with minimal or no restriction on their lifestyle before surgery, in contrast to UC patients who are often ill and symptomatic. Therefore, restrictions on lifestyle after surgery may be perceived as more severe by FAP patients [4].

On the other hand, in a prospective, age-related analysis of surgical results, functional outcome and quality of life after IPAA, Delaney et al. [186] showed that although functional outcome after IPAA is not as good as in older patients, there were only minor differences in quality of life, health, energy and happiness between age groups, with a slight benefit for those under 45 years, which is in fact the more frequent age for IPAA in FAP.

Finally, regarding IPAA in teenage FAP patients, the impact of IPAA on quality of life was favourable in the majority of them [37].

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%) [4–8, 12, 16, 33, 78, 169, 181, 193–204]. The reason for this very low incidence of pouchitis in FAP is still unknown [4]. 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 [4, 205].

Adenomas, dysplasia and cancer after IPAA

Even after a prophylactic RPC with IPAA, FAP patients are still at the risk of developing adenomas with dysplasia or adenocarcinomas, either in the anal canal or in the ileal pouch itself. At present the prospect of actual cancer seems unlikely, but with the increased length of follow-up after IPAA more and more reports of pouch polyps are coming out with the occasional report of pouch cancer.

Since residual rectal mucosa carries a much higher risk of malignant transformation than ileal pouch mucosa, it is essential to distinguish adenomas arising in the anal canal from true adenomas originating in the ileal pouch itself [8].

Risk of neoplasia in the anal canal

The fate of retained at-risk – *APC* mutation carrying – epithelial cells in the anal canal after IPAA has already been discussed. To date, it is almost impossible to estimate the risk of dysplasia and cancer in this retained anorectal mucosa. However, this risk is theoretically and logically greater after the double-stapled technique, which preserves the ATZ zone, than after the mucosectomy technique, where it has been shown that small islets of columnar epithelial cells can also be left behind [4, 5, 16, 21, 42, 49, 52, 55, 66, 81, 83, 85, 92, 206]. Some reports in fact indicate that histologically examined anorectal mucosal strippings taken at the time of proctocolectomy for FAP already contained dysplasia in 75–100% of cases [5, 81, 92].

There are reports of adenomas arising in the anal canal after either mucosectomy or double-stapled anastomosis [52, 66, 207–209]. The incidence of dysplastic polyps in the residual ATZ after double-stapled IPAA has been estimated to be up to 28–31% compared with 10–14% after endoanal mucosectomy, which is nearly less than half [52, 66].

Regarding the major cause of concern, i.e. true invasive adenocarcinoma in the anal canal, only 8 cases have been reported to date, 4 after incomplete mucosectomy with 1–2 cm of mucosa having been left behind, and 4 after double-stapled anastomosis [39, 79, 81, 87, 210, 211].

One of these cancers originated from a true rectal stump of at least 5 cm left behind after what could actually be called a double-stapled pouch-rectal anastomosis in an obese male patient with a narrow pelvis. This illustrates the risk of leaving lower rectum in situ in very difficult cases [39, 66]!

Therefore, close follow-up by video-endoscopy with biopsy of all suspicious lesions, destruction of small polyp by fulguration (provided there are fewer than 10 polyps) and transanal excision of large polyps are recommended [66]. If no polyps are found, we recommend annual examination. If adenomas is already present, a six-month follow-up would be advisable. If serious neoplasia occurs – high-grade dysplasia or carpeting of the mucosa – the ATZ can usually be stripped transanally and the pouch advanced to the dentate line with a redo-IPAA [21, 39, 52, 66, 208, 212]. Finally, invasive adenocarcinoma arising in the anal canal after IPPA has to be treated as for a low rectal carcinoma by abdominoperineal excision of the pouch and anal canal and conversion to an end ileostomy [40, 213].

Risk of neoplasia in the ileal pouch

In FAP, adenomas may occur in the ileum, but they are not common, and the incidence of small-bowel cancer outside the duodenum is low [2–4, 214–216]. Moreover, the long-term fate of the mucosal lining of an ileal reservoir in patients with FAP is not completely known [5]. Therefore, until recently, the risk of pouch polyposis and pouch cancer had not been seriously considered [214].

Now, pouch polyps and polyposis following IPAA for FAP have also been observed: FAP patients may develop adenomas or microadenomas in the reservoir itself. Several articles document a prevalence of polyps in the pouch, especially after long-term follow-up, of between 8% and 62% [8, 66, 80, 209, 217–222].

The risk of developing one or more adenomas at 5, 10, 15 years has been calculated and was 7%, 35% and 75%, respectively [8]. Parc et al. [8] showed that FAP patients with pouch adenomas were more likely to have duodenal and ampullary adenomas, raising the notion of a specific FAP phenotype. However, no correlation could be found between this phenotype and the site of the *APC* gene mutation.

The age of a pouch is clearly important in the development of adenomas, and older pouches warrant more careful follow-up because adenomas become very common after ten years [221]. Further investigations and longer follow-up will therefore be required to determine whether pouch adenoma is inevitable in all patients with IPAA or specific to a subgroup of patients [8]. The exact impact of pouch polyposis will not be fully understood until most FAP patients with ileal reservoirs reach a mean follow-up of 20, 30 or 40 years [21].

Most pouch polyps reported have been small tubular adenomas with mild dysplasia, and it has been suggested that these are unlikely to progress to pouch cancer [214]. However, as in the colon and duodenum in FAP, it has recently been postulated that there is likely to be an adenoma–carcinoma sequence in the ileal pouch [214]. This is in fact suggested by the description by Beveridge et al. [214] of two large villous adenomas in a pouch displaying all the risk factors of malignant transformation: size, sessile nature, severity of dysplasia and villous architecture. Three cases of true pouch adenocarcinomas have in fact been reported after IPAA [213, 223, 224]. All reported cases of pouch cancer in FAP had a high cancer risk at the time of IPAA [39].

In the meantime, all these findings strengthen the recommendation for careful regular endoscopic monitoring of FAP pouches and the evaluation of management and treatment strategies for pouch adenomas [214]. Long-term monitoring by pouchoscopy in all patients with IPAA for FAP is mandatory. Early detection may allow control by means of medical therapy and an endoscopic or limited surgical procedure [209].

Pouch polyposis has been treated successfully by oral Sulindac, at a dosage of 150–200 mg twice daily [21]. Large pedunculated polyps are easily removed by snare polypectomy and argon plasma coagulation [214]. Frank malignancy in a pouch has to be treated by pouch excision and terminal ileostomy [40, 213].

Recommendations

All FAP patients have a genetic and lifelong risk of developing polyps in any residual rectal mucosa as well as in ileal mucosa. Consequently, they should undergo regular endoscopic monitoring (yearly) of both pouch and anastomotic areas in the anal canal after IPAA, regardless of the type of anastomosis, whether hand-sewn or double-stapled (Table 1) with biopsy of all suspicious lesions, destruction of small polyps by fulguration, provided there are fewer than ten polyps, or transanal excisions of larger polyps.

Fertility, pregnancy, childbirth

Fertility

Patients with FAP are generally young and fit, and many are childless at the time of operation and the desire for future pregnancies is also important [225, 226].

IPAA operation does not jeopardise pregnancy and childbirth but some reports have suggested that it could impair fertility [154, 227–233]. However, until recently, knowledge about the fertility of women suffering from FAP was sparse and inconclusive [225, 234]. In fact, the majority of the reports about fertility after IPAA were related mainly to women with UC, in whom a dramatic decrease in fertility of 80% has been demonstrated [235–238].

Recently, Olsen et al. [225] have shown that fertility dropped to 54% following IPAA in FAP patients, although it was much greater than the postoperative fertility of women with UC. To date, the reasons for this difference between FAP and UC are still not known and will need further investigation. Notwithstanding this difference, it has been postulated that this adverse effect of IPAA on fertility, for both FAP and UC, could to some extent be a result of surgical technique and postoperative pelvic adhesions [159, 225, 232, 235–240]. It seem 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 [159, 225, 235–238].

Gynaecological surgeons have been studying the link between pelvic adhesions with infertility for a long time and have shown that moderate to severe pelvic adhesions may be responsible for 40% of infertility [154, 241, 242].

In fact, two previous studies carried out after coloproctectomy and IPAA or terminal ileostomy with

Table 1. Prophylactic surgery for FAP patients in our practice (modified from Soravia and Cohen [5]).

Factor	IRA	IPAA
Indications for surgery	< 20 rectal polyps AAPC	Most of FAP patients >1000 colonic polyps Cancer anywhere in the large bowel > 20 rectal adenomas Rectal polyp carpeting Severe dysplastic rectal adenoma Large (> 3 cm) rectal adenoma Resectable rectal cancer Desmoid in 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: 2 x/year	Pouch endoscopy: 1 x/year; 2 x/year if polyps

AAPC = attenuated adenomatous polyposis coli; CRC = colorectal cancer; FAP = familial adenomatous polyposis; IPAA = restorative proctocolectomy with ileal pouch-anal anastomosis and mucosectomy; IRA = total colectomy with ileorectal anastomosis; QOL = quality of life.

hysterosalpingograms found complete unilateral or bilateral obstruction of the Fallopian tubes in 52% of patients [232, 240].

A strategy to reduce such adhesions would involve intraoperative measures to preserve tubal patency and normal anatomical relationships [159, 238, 243]. Instillation of adhesion-prevention gels/anti-adhesives barriers has been suggested [244, 245]. Although various materials are available to prevent pelvic adhesions, there are no data regarding efficacy in improving fertility, and further research is necessary in this area [246].

Oophoropexy has also been proposed [159, 237]. But these measures have not been widely implemented yet and are not part of our routine practice.

The laparoscopic approach could be another highly promising tool in the future to tackle post-IPAA adhesion formation and therefore infertility as well [14, 159]. In an extensive review of the literature, Gutt et al. [154] found that all clinical investigations and most experimental studies reported a reduction of adhesion formation after laparoscopic surgery compared with open surgery. Whether laparoscopy also significantly reduces formation of pelvic adhesions and could therefore influence fertility after IPAA is the subject of ongoing studies [159].

Whatever the case, this reduction in fecundity in women with FAP undergoing IPAA should be discussed with patients before surgery [5, 225, 238]. Whether this should change the surgical strategy to one of IRA and secondary IPAA in women keen to have a family is uncertain [226, 238].

It is not yet clear how much emphasis should be placed on a woman's reproductive career when offset by the potential risk of rapid progression of rectal polyposis in 'high-risk' FAP individuals [226].

Pregnancy–delivery

The majority of studies available to date involve UC patients, which means an extrapolation when discussing FAP patients.

Despite the first report of a successful pregnancy and delivery after IPAA in 1984 by Pezim [227] recommending Caesarean section, all published studies to date found that IPAA is compatible with a safe pregnancy and normal vaginal delivery [159, 228, 230, 239, 247–249].

Pregnancies are usually uneventful, without complications and carried to term. Small-bowel obstruction in particular seems to be rare [159]. Pregnancy does not adversely affect pouch function. The number of daily bowel movements increases only modestly, the increase being noted in the last trimester of pregnancy and persisting for about 3 months after delivery [159, 228, 230, 239, 249, 250].

The type of delivery does not affect pouch function post partum. The presence of an ileal pouch does not mandate Caesarean section, and such a distinction should be based on obstetric considerations [231, 248]. If necessary, a mediolateral episiotomy can be performed safely.

However, since the long-term effects (i.e. 20–40 years) of vaginal delivery on pouch function are unknown, we still have to be careful when advising a vaginal delivery in a primipara with an ileal pouch [251]. We must be aware that there is increasing evidence for occult sphincter injury and pelvic floor innervation damage after a first vaginal delivery [249, 251, 252].

The result of several 'hits' including ageing, obstetric injury and surgical reconstruction could lead to an increased risk of incontinence. Such damage could be

more devastating to a patient with a pelvic pouch [249, 251].

When counselling FAP patient, we should keep in mind that the full impact of the delivery route on pouch function will not be known before many years.

Urinary and sexual function

Given that the majority of patients who develop FAP do so either during or before their prime reproductive years, the impact of IPAA on urinary and sexual function in men and women is a most important consideration [4, 6, 229, 247].

Urinary and sexual dysfunction does arise and is likely to be technique-dependent [247]. By using the 'close rectal wall' dissection technique for proctectomy, as described above, urinary and sexual dysfunction can be almost completely avoided [4, 6, 129]. In a series of 171 FAP patients, the St-Antoine's group reported a 1.7% rate of transient postoperative dysuria and urinary retention, 0.6% transient impotence and 0% retrograde ejaculation [6]. We experienced the same results in our unit [129]. This is clearly in contrast to results after the TME-like conventional dissection carried out in the anatomical plane between the mesorectum and the presacral fascia.

In a comprehensive review of all published papers, Colwell and Gray [33] reported rates of sexual disturbance in males of 0.5–1.5% for erectile dysfunction and of 3–4% for ejaculatory dysfunction after IPAA. In males, denervation of the pelvic plexus is itself postulated to contribute to erectile and ejaculatory dysfunction [33]. Furthermore, when using a true TME technique for FAP patients with resectable rectal cancer or high-grade dysplasia, rates of urinary and sexual disturbance of nearly 10% or more have been reported [17, 38, 247].

Sexual dysfunction in women after proctectomy is less well-described, in part because of reluctance by both physicians and patients to discuss such matters [229, 247], and in part because female sexual dysfunction is much harder to measure. In their extensive review, Colwell and Gray [33] found a dyspareunia rate of 3–22%. In addition, the fear of leakage of stool inhibited sexual relations in 3% of women. Colwell and Gray [33] concluded that sexual dysfunction affects both genders, but women are at greater risk of this adverse effect than men.

Theoretically, sexual dysfunction in women after proctectomy could result from injury to the autonomic nerves, but it has also been postulated that mechanical problems secondary to anatomical changes within the pelvis produced by removal of the rectum could contribute to dyspareunia [33, 229, 247]. For instance, Metcalf et al. [229] found that patients after proctectomy and terminal ileostomy have a greater incidence of dyspareunia than those with an ileal pouch posterior to the vagina. This underlines the importance of distortion of the pelvic anatomy.

However, the incidence of these urinary and sexual complications may decrease, as it has been reported that the incidence of surgical complications is declining because of increasing experience with and standardisation of the IPAA procedure [6, 20, 50, 103, 182, 194, 253]. As regards recommendations for current clinical practice, FAP patients should be advised of anticipated functional outcomes of the IPAA affecting urinary and sexual function [4, 33].

Surgical 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 the multiple parameters into account, as listed in Table 1.

IRA vs. IPAA

We believe that the first parameter to be taken into account for the selection of an operation should be based upon the perceived risk of cancer development in the residual rectum [4, 6, 254, 255]. To this end, we feel that there is no argument that patients with severe rectal (>20 adenomas) or colonic (>1000 adenomas), or those with a severe dysplastic rectal adenoma, a cancer anywhere in the large bowel or a large (>3 cm) rectal adenoma should have a primary IPAA [21, 86, 256, 257].

In those undergoing IRA, the major cause of concern remains the risk of rectal cancer, despite strict and rigorous endoscopic monitoring of the rectum. But, since some reports have highlighted the risk of adenoma formation in ileal pouches, both IRA and IPAA require lifelong monitoring of the rectum or pouch, because both are at risk of developing adenomas [8, 21, 86, 221, 256, 258].

At 10, 15, 20 and 25 years after IRA surgery, the cumulative risk of developing rectal cancer by years of follow-up after surgery were 3.9%, 10.4%, 12.1% and 25.8%, figures largely confirmed by other studies [2, 4, 8, 259–272]. Moreover, despite lifelong regular and careful rectal examination with systematic destruction of all newly formed polyps, rectal cancer cannot always be detected at an early stage, and so threatens the patient's life and ultimately may require a proctectomy with a permanent abdominal stoma [1, 17, 209, 265, 272].

The risk of rectal cancer after IRA is strongly linked to the severity of colorectal polyposis at presentation, and IRA is a reasonable option in mildly affected patients (<20 rectal adenomas, <1000 colonic adenomas) and includes all those with attenuated FAP [21, 86, 256].

As regards the surgical procedure itself, the mortality rate is comparable for both procedures and is very low at 0.5–1% [6, 273].

Morbidity after IPAA remains more significant than after IRA, but increased surgical experience has signif-

icantly decreased the incidence of complications [6, 257, 274]. The rate of SBO is the same in both groups and is the major cause for repeat surgery [4, 6, 273]. The risk of desmoid tumour development is also the same after both types of surgery [166, 209, 275]. The functional results after IPAA improve over time and, as regards ultimate function, there appears to be little to choose between IPAA and IRA [5, 6, 12, 181, 190, 209, 255, 276–278]. And even if IPAA is a more complex procedure with more morbidity and functional results are slightly better after IRA, Van Duivendijk et al. [279, 280] have shown that there is no difference in the quality of life experienced by the patients after both type of operations.

In our colorectal surgery unit, we recommend IPAA for almost all our FAP patients, and base our recommendation on the risk of rectal cancer after IRA and equivalent quality of life after the two operations (Table 1). But we accept that IRA could still be a reasonable option in mildly affected patients (<20 rectal adenomas), in attenuated FAP and in young women before childbearing (Table 1) [4].

Secondary IPAA after IRA

Since the cumulative risk of developing a rectal cancer is related to age, Nugent and Phillips [272] have proposed performing a primary IRA in patients under the age of 30 years, with a subsequent proctectomy and IPAA by the age of 45 years, if necessary. The risk of cancer with IRA may therefore be overcome by this surgical strategy. Moreover, secondary proctectomy with IPAA following IRA may be safe and has an outcome similar to that of primary proctocolectomy with IPAA [281–285].

However, conversion of IRA to IPAA is not always feasible because of malignant disease or abdominal DT precluding further rectal stump management [4, 54, 86, 276, 281]. 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 DT. Two of these patients died from rectal cancer [281]. This may argue in favour of IPAA as the first surgical step in FAP treatment [4, 281], particularly since the incidence of DT has not been proven to be higher after IPAA compared to IRA. Indeed, our preference is to advocate IPAA in cases with a familial history of DT (Table 1). When a DT is found at the time of the first laparotomy, IPAA should be seriously considered if technically feasible [4, 209].

Genetics and surgery

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.

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 [4, 21, 272, 286–290]. There is a particular 'hot spot' mutation at codon 1309 that always causes severe disease, usually with thousands of polyps.

Vasen et al. [259] 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) [79, 259].

However, Giardiello et al. [289] has 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 kindreds carrying the some APC mutation but also within kindreds [243, 291].

Bertario et al. [290] also found a significantly increased risk of early colorectal cancer (CRC) associated with two areas before codon 1250: 514–713 and 976–1067. This means that caution should be exercised against setting strict surgical guidelines based on mutational analysis.

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 [4, 292, 293]. 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 [9, 292, 294]. Evidence indicating a much lower rate of CRC in AFAP families than in classic FAP families has been reported in recent years [9, 292]. In all AAPC kindred, a predominance of right-sided colorectal adenomas and rectal polyps sparing was observed [294]. Accordingly, if surgery for otherwise intractable polyps is indicated, total colectomy with IRA is recommended (Table 1) [9, 292, 294].

But since the natural history of AAPC is not well documented yet, and therefore the exact risk of colorectal cancer remains unknown, caution should again be exercised when choosing IRA, and lifelong rectal monitoring should be mandatory [9, 292, 294].

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 [291, 295]. 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 [291, 295].

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) [4, 167, 296].

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 [243, 294]. 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 [4].

Conclusions

In conclusion, since there is a significant risk of rectal cancer after IRA, and despite the controversies around technical issues, we believe that a complex procedure such as IPAA should be currently proposed to the majority of FAP patients as the operation of choice. It is safe, it allows a complete removal of the diseased colorectal mucosa, it carries an acceptable risk of complications, and it offers predictable functional results with a high level of patient satisfaction.

Continued technical advances and greater surgical experience can only further improve function, outcome and patient satisfaction. But it is crucial that surgeons treating the disease are aware of the alternative surgical options, all of which continue to have an important role for specified subsets of patients.

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