

## Different surgical strategies in the treatment of familial adenomatous polyposis : what's the role of the ileorectal anastomosis ?

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### Abstract

Prophylactic (procto-) colectomy is the treatment of choice to reduce the risk of colorectal cancer in FAP patients with multiple adenomas. Because patients present at young age, rectum-sparing surgery is sometimes advocated, so that there is no pelvic dissection with impact on quality of life, preserved pelvic innervation and sexual function and fertility. The main disadvantage of a total colectomy with an ileorectal anastomosis (IRA) is a rectal cancer risk of 50% at the age of 50 years and a cumulative risk of 25,8% after 25 years of follow-up. Therefore, this procedure should be reserved for patients with an unaffected rectum. There should be no discussion to perform a primary IPAA in patients with multiple rectal adenomas (> 20) or those with a severe dysplastic or large (> 3 cm) rectal adenoma or a cancer elsewhere in the colon. A patient with an IRA should undergo yearly follow-up by rectoscopy. (*Acta gastroenterol. belg.*, 2011, 74, 435-437).

### Introduction

Familial adenomatous polyposis (FAP) is an autosomal-dominant inherited colorectal cancer syndrome, characterized by an early onset of hundreds to thousands of adenomas throughout the colon and rectum, usually in adolescence. It is caused by a germline mutation in the adenomatous polyposis coli (APC) gene, located on chromosome 5q21.

FAP patients develop polyps in the second and third decades of life and if left untreated, they will definitely develop colorectal cancer, usually by the age of 40 years (1).

Prophylactic (procto-) colectomy is the treatment of choice to reduce the risk of colorectal cancer (CRC) in FAP patients with multiple adenomas. Three different surgical options are a total colectomy with ileorectal anastomosis (IRA), a proctocolectomy with a (continent) ileostomy or a proctocolectomy with an ileal pouch-anal anastomosis (IPAA) (2). The aim of this article is to discuss the different surgical possibilities, with emphasis on the role of IRA.

### Statement of the problem

Prophylactic surgery in the treatment of FAP should focus on the removal of the entire mucosa from the

caecum up to the dentate line at the anal verge, because the adenoma-carcinoma sequence can occur in every epithelial cell of the colon and the rectum. Because patients present around the age of 20 years, rectum-sparing surgery is often advocated, so that there is no pelvic dissection with better quality of life, preserved sexual innervation and fertility (3,4). Questions with regard to rectal cancer risk, fertility and choice of operation will be addressed in this paper.

### Rectal cancer risk

The risk of developing rectal cancer is not the same in all FAP patients. Correlations have been made between number of polyps and the risk of colorectal cancer (5). A number of 1000 synchronous adenomas is used to define a severely affected colon and 20 synchronous adenomas to define a severely affected rectum. Even the presence of a rectal adenoma with severe dysplasia or a large (> 3 cm) adenoma with predominantly villous histology is enough for a 'high risk rectum'. The difference between a 'high' and a 'low' risk rectum has surgical implications. After IRA, thus leaving the rectum behind, there is a rectal cancer risk of 50% at the age of 50 years and a cumulative risk of 25,8 % after 25 years of follow-up (6-8). Even for IPAA using a double-stapled anastomosis where mucosectomy is not routinely performed, the retained mucosa has the potential to undergo the adenoma-carcinoma sequence (9,10). For both IPAA and IRA there is a need for endoscopic follow-up. The authors recommend yearly pouchoscopy or rectoscopy after IPAA or IRA, respectively (11-13).

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## Fertility

There is increasing evidence that pelvic dissection and ileal pouch surgery will decrease female fertility. This fact should be taken into account when counselling patients for surgery. Pouch surgery for FAP is often carried out in women who are still in their fertile years and of whom many have not given birth at the time of surgery. It is important to know whether these women have normal fertility before surgery, or if their fertility is impaired compared to other women. Three studies have shown that reproductive fitness of female FAP patients before surgery is comparable to or even better than a reference population (14-16). After pelvic surgery, however, there are studies indicating that fertility will be impaired (17,18). Data for fertility after IPAA for FAP are limited. Only one co-operative study from the Nordic Polyposis registers showed that fertility after IRA was normal, but it was significantly reduced after IPAA. After 2 years of unprotected intercourse the success rate of becoming pregnant was 86% before surgery, 87% after IRA, but only 61% after IPAA (16). This shows that IPAA in FAP patients has an important effect on fertility. Therefore, in cases of young compliant women with a low rectal cancer risk, it should be possible to perform IRA and to allow childbearing, with conversion to IPAA at a later stage (19).

## Choice of procedure

Since the introduction of the IPAA operation for FAP in the late 1970s, the choice between IPAA and IRA in FAP still remains controversial. Therefore, some attempts have been made to design the best surgical strategy for each FAP patient.

Most patients undergo prophylactic surgery in their second or third decade (3,4). During this period of life, important decisions with regard to relationships and career are made. Because of this and because these patients are usually asymptomatic, it is very important to choose an operation with a low morbidity- and a very low mortality-rate. It should interfere with normal life as little as possible (4). Results of the Dutch Polyposis Registry, comparing IRA with IPAA, show that there were no significant differences with respect to quality of life (4). Two other studies regarding health-related quality of life showed, despite poorer functional results, no significant differences between the IRA and the IPAA groups (20,21).

In approximately 20 to 30% of patients with FAP, the rectum is either spared of polyps or has a significantly decreased number (22). In patients with a low rectal polyp count ('low risk rectum'), a total colectomy with IRA or ileostomy could be a valid alternative. This approach avoids deep pelvic dissection and avoids any risk for autonomic nerve damage and would result in a reduced risk for anastomotic leakage. In most patients an ileostomy can be avoided. The main reason to perform a

total colectomy with IRA (rather than a proctocolectomy with IPAA) is the better functional result (i.e., decreased stool frequency, less night-time leakage), together with preserved fertility in young females and sexual function in males (5,23,24).

There are two important conditions before an IRA can be performed. These are a low rectal cancer risk and the absence of or predisposition to develop a desmoid tumour. Without any discussion, in patients with severe rectal (> 20 adenomas) or colonic (> 1000 adenomas) polyposis or those with a high grade dysplastic rectal adenoma, a cancer anywhere in the large bowel or a large (> 3 cm) rectal adenoma a primary IPAA should be performed (25-27). The risk of rectal cancer after IRA is linked to the severity of polyposis and therefore IRA is a reasonable option in mildly affected patients, in young women before childbearing and includes all those with attenuated FAP (25-27). Thus, indications for IRA are a low rectal polyp count (< 20) and a motivated patient, who is willing to undergo close follow-up with a rectoscopy every year.

With regard to the surgical procedure itself, the mortality rate is comparable for both procedures and is very low at 0.5-1% (8,28). Morbidity after IPAA remains more significant than after IRA, but increased surgical experience has significantly decreased the incidence of complications. In the recently published experience of the Cleveland Clinic, specifically addressing the question of IPAA vs. IRA, authors propose a decisional algorithm based on composite parameters as severity of polyposis, degree of rectal involvement and genetic data. They currently report 80% of the patients with attenuated FAP were treated with IRA and 81% of patients with severe FAP with IPAA whereas 63% and 37% of the patients with mild FAP were treated with IRA and IPAA, respectively (29).

The overall prevalence of desmoid tumours in FAP is 15% and clusters in families (30). These benign tumours are locally invasive fibromatosis with a major cause of morbidity and mortality. Traditionally, desmoid tumours have been associated with trauma, such as abdominal surgery, e.g. prophylactic colectomy. A recent meta-analysis did not show any significant difference between IRA and IPAA in terms of the incidence of intra-abdominal desmoids (31). It has been described, however, that conversion of IRA to IPAA was impossible due to desmoid disease, with deaths related to rectal cancer (32). Consequently, the threat or existence of an intra-abdominal desmoid tumour should be a contra-indication. Whether at time of pouch surgery it is possible to create a pouch will be dependent upon the presence or absence of a desmoid lesion. Therefore a preoperative MRI of the abdomen is indicated to detect the presence of a mesenteric desmoid tumour. Family history, presence of osteomata and germline mutations after codon 1399 have been identified as independent risk factors for desmoid occurrence (30). CT-scanning is the best modality for making the diagnosis, but MRI

scanning using T2 weighted imaging may give an indication as to how the tumour is likely to behave, with a bright signal indicating high water content and association with rapid growth (33,34). Thus, in cases with a family history of desmoid disease, genetic predisposition or existence of a desmoid tumour on MRI, an IPAA should be performed.

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