REVIEW ARTICLE

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Dispelling misconceptions in the management of familial adenomatous polyposis

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Abstract

Patients with familial adenomatous polyposis require surgical intervention at some point in their lives. The diagnosis is often apparent from their phenotype and family history, however, this is not always the case. Many factors can influence the surgical strategy although the polyposis burden and distribution remain the main consideration. While prophylactic removal of the rectum and colon is often required, sparing the rectum at the index surgery is safe in select patients. This article aims to dispel misconceptions in the diagnosis and treatment of patients with familial adenomatous polyposis.

Introduction

Familial adenomatous polyposis (FAP) is an autosomal dominantly inherited disease that presents with numerous colonic adenomatous polyps.[1] It arises due to a germline mutation in the tumour suppressor gene *APC*. Classical FAP presents with hundreds and often thousands of adenomas that are impossible to control endoscopically. Without colectomy, colorectal carcinoma (CRC) is inevitable, at a mean age of 39 years.[2] Prophylactic screening and surgery in affected individuals aims to minimize this risk. While prophylactic proctocolectomy is considered by some to be routine for patients with FAP, this approach has been challenged by a desire to preserve the rectum in the interests of quality of life and reducing operative risk.

Patients with FAP may also suffer from desmoid disease which can pose serious challenges for the treating clinician. The risk or presence of desmoid disease can influence surgical strategy. It has been suggested by some that the development of desmoid disease in patients with an ileorectal anastomosis may prevent future proctectomy and therefore a proctocolectomy should be offered originally, instead of the ileorectal anastomosis.[3]

Because FAP is such a rare disease, the experience of individual surgeons may be skewed by unusual presentations and outcomes. The course of patients with the disease may be unpredictable and management decisions may be made on the basis of anecdote, 'myth', and fear of possible (but unlikely) adverse consequences, rather than evidence. Choice of surgery and the role of desmoid disease are just two examples. The aim of this review is to settle some of the misconceptions in the diagnosis and management of FAP.

Will all patients with familial adenomatous polyposis have a family history of polyposis or CRC?

Family history is an important clue to the diagnosis of FAP. The autosomal dominant pattern of inheritance, along with high penetrance, and strong phenotypic expression, means that FAP is the most readily recognizable of the hereditary colorectal cancer

syndromes. The disease typically presents with hundreds or thousands of colorectal polyps as well as certain characteristic extra-colonic and extra-intestinal manifestations. In up to 25% of cases, however, there will be no family history prior to diagnosis.[4]

In the majority of these patients with no family history of FAP, it is believed that a new mutation has occurred at conception, but other reasons for the lack of family history include ignorance (through adoption, non-paternity, death of the affected parent or the deliberate withholding of the diagnosis by family members) and somatic mosaicism.[5] In cases of mosaicism, the *APC* mutation is present only in certain tissues (such as the ovary or sperm cells) and will not be detected by sequencing of DNA from white blood cells. Unlike patients with a family history of FAP who are aware of their risk and so can be tested for the presence of the mutation, patients without a family history usually present with symptoms and have a high risk of CRC being present at the time of diagnosis.[6]

An unpublished study at the Cleveland Clinic compared 50 FAP probands without a family history of FAP with 73 probands with a family history that acted as controls. In this study, patients without a family history were more likely to have colectomy at a younger age (31 versus 33), more likely to have a profuse polyposis at diagnosis (34.9% versus 19.3%), were more likely to undergo a proctocolectomy, and were almost twice as likely to have colorectal cancer, although no difference in overall survival was observed.

Probands have a significantly higher mortality rate when compared with patients with a family history of FAP. Koskenvuo *et al.* showed that probands had a mortality rate of 34.9 per thousand person-years versus 8.3 per thousand person-years for 'call-up' patients who were being screened because of a family history of FAP. This difference was attributed to death from CRC being higher in the probands than the 'call-up' patients (29% versus 3.8%) with no difference in death from other causes between the two groups.[7] This demonstrates worse colorectal cancer outcomes in probands and supports colonoscopic screening in FAP families in order to reduce mortality from CRC.

The authors seek genetic testing for an *APC* or *MYH* mutation in any patient presenting with >10 synchronous colorectal adenomas regardless of family history of polyposis or CRC.[8]

Should all patients with FAP undergo prophylactic proctocolectomy to prevent future neoplasia?

The primary goals of colorectal surgery in FAP are cancer prevention and treatment, and maximizing quality of life. As colorectal surgery is often pre-emptive and is usually considered in young patients, there are increasing patient expectations for technical perfection.[9] The extent of resection depends on a balance of reducing cancer risk while preserving bowel function and quality of life. Surgical options include total proctocolectomy (TPC) (with restoration of gastrointestinal tract or with end ileostomy (EI)) or colectomy with ileorectal anastomosis (IRA). Both options have oncologic and functional implications. While colectomy and IRA maintains better bowel function, there is risk of cancer developing in the rectum. Conversely, restorative TPC with ileal pouch anal anastomosis (IPAA) nearly eliminates colorectal cancer risk but results in worse functional outcome compared to an IRA.

Before the advent of the ileal pouch (1978),[10] the choice of operation was between a TPC and EI or a total colectomy (TAC) and IRA. Because most patients rejected the notion of a prophylactic procedure that gave them a permanent ileostomy, many underwent an IRA when they would have had an IPAA had that been available to them. Indeed the secondary proctectomy rate following TAC and IRA was 33% at a median interval at 212 months during this 'pre-pouch' era.[11] This proctectomy rate is unacceptably high. However, a follow-up study from the Cleveland Clinic showed that more appropriate selection of patients for IRA in the 'post pouch' era led to a proctectomy rate of only 2%.[12] These data are supported by Bussey *et al.* from St Mark's.[13] From 173 patients who had IRA, only 11 patients required future proctectomy for rectal cancer, and six underwent proctectomy for adenoma and benign conditions.

Morbidity and quality of life data should be considered when choosing an operative strategy. A meta-analysis of 12 observational studies compared the morbidity and functional data of restorative proctocolectomy (RPC) with those following a colectomy and IRA. There was no difference in immediate bleeding or septic complications but RPC with IPAA was associated with a higher 30-day reoperation rate, worse stool frequency and more 24-h incontinence.[14] No difference was observed in nocturnal stool frequency and day-time incontinence. In addition, pelvic surgery is associated with increased sexual and urinary dysfunction,[15] decreased fecundity in females,[16] a higher chance of impotence in men and reduced quality of life scores following IPAA compared with IRA.[17] All of these factors should be considered when suggesting an operative strategy.

Polyp burden predicts future proctectomy risk. A study by Church *et al.* observed 94 patients with less than five rectal adenomas and less than 1000 colonic polyps who underwent IRA and were followed for a median of 12 years. No patient required secondary proctectomy.[12] Conversely, 35% of 74 patients who had >20 rectal polyps and underwent a IRA, required future proctectomy. Indeed simple proctoscopic examination with less than five rectal adenomas nearly always correlates with mild disease. In such cases, an IRA is appropriate.

APC genotype can also assist in the surgical planning for FAP patients. The two genotypes that predict future proctectomy are:

(1) 1309 mutation.
(2) 1328 mutation.[18]

Therefore patients carrying these mutations (and 1309 is a 'hotspot' for FAP families) should have a RPC.

The authors advocate a TAC and IRA for select patients with FAP who have a low risk of future intestinal neoplasia and/or should not undergo the increased risk and questionable function of a RPC. The objective criteria for rectal sparing are:

(1) No rectal cancer.
(2) No advanced rectal neoplasia (tubulovillous adenoma, large >1 cm, high grade dysplasia).
(3) Low polyp burden in the rectum (<20 adenomas).
(4) Absence of profuse colonic polyposis.
(5) Absence of colonic cancer.
(6) Absence of 1309 or 1328 mutations on genotype.

Should patients with FAP undergoing a restorative proctocolectomy have a hand-sewn anastomosis?

Following removal of the colon and rectum in individuals with FAP, an ileal pouch is created and anastomosed to the anus. The most commonly fashioned pouch is a J pouch. Management of the distal anorectal mucosa is still debated.

Parks and Nichols from St Mark's suggested removing the all anorectal mucosa to the dentate line and performing a hand-sewn (HS) anastomosis.[10] The benefit of this approach is that it may remove all risk mucosa. However, we would caution that an incomplete mucosectomy can result in islands of at risk mucosa, and several cases of anal transitional zone cancer following mucosectomy have been described.[19] The alternative is to retain the distal anal segment and staple the ileal pouch to the top of the anal canal. The advantages of this technique are that it is quicker to perform, easier to survey and involves less manipulation of the anal sphincter complex with the potential for improved function.

A single institution series assessed 119 patients who underwent IPAA: 77 had a stapled anastomosis while 42 underwent HS anastomosis. Worse day and night time seepage, incontinence rates and increased pad usage were observed in the mucosectomy group when compared with the stapled approach.[20] However, reduced neoplasia risk was observed in the mucosectomy group (28% versus 14%). Importantly, the same group reported on adenocarcinomas arising following both HS and stapled anastomosis. Neither approach was free from risk.[21] Data from St Mark's registry support the decreased risk of distal adenomas in patients with a mucosectomy.[22] One hundred and forty patients underwent IPAA. Twenty-seven percent of the mucosectomy group developed adenomas when compared with 54% in the stapled group. Importantly, no high-risk adenomas were reported in this study. A more recent Norwegian study compared patients with FAP who underwent IPAA with/without mucosectomy and also found an increased rate of adenoma at the anastomotic site when a mucosectomy was not performed.[23]

Lovegrove *et al.* performed a meta-analysis comparing mucosectomy with stapled anastomosis following RPC. Twenty-one studies with 4000 patients were included in the analysis. Worse nocturnal continence in the mucosectomy group was found when compared with the stapled group. No difference in postoperative complications, the number of daily bowel motions and requirement for anti-motility drugs were observed. The study is limited in that it did not differentiate between ulcerative colitis and FAP patients when comparing the two approaches.[24]

The authors' practice is to perform a stapled ileoanal pouch anal anastomosis, unless there is obvious polyposis or dysplasia that extends down towards the dentate line. When performing a stapled IPAA it is wise to minimize the length of the retained columnar epithelium, the 'columnar cuff' to approximately 1 cm. This allows for easy removal of the columnar cuff if carpeting with polyps subsequently occurs.

Should FAP patients with known desmoid disease have a proctocolectomy rather than total colectomy and ileorectal anastomosis?

Desmoid tumours (DT) present a significant challenge to patients with FAP. They cause morbidity arising from local compression and can lead to sequelae including bowel obstruction, perforation, ureteric compression with resultant renal failure and major vascular issues. While desmoid disease can arise *de novo* (3% incident rate at primary surgery),[25] the majority of intra-abdominal and abdominal wall lesions occur following index colectomy (up to 80%).[26–28]

Patients with a high risk of DT can be identified by a positive family history of DT as well as a suggestive genotype.[28–31] Some studies have shown female gender to be an independent risk factor for development of DT in FAP.[28,32,33] Elayi *et al.* have described a scoring system to help identify such high-risk patients.[34] For patients that are prone to develop desmoids, it may be prudent to delay surgery.[33]

There is significant controversy as to which surgical strategy is more likely to predispose to desmoid tumour. It has been suggested that ileal pouch reconstruction may result in more desmoid tumour by placing tension on the ileal mesentry.[35] This theory is supported by location of DT in the ileal mesentery. A study by Vitellaro *et al.* demonstrated an increased risk of desmoid tumour in IPAA compared with IRA with a hazard ratio of 1.67.[36] However, numerous other studies have shown no difference in rate of DT between patients who have undergone an IRA or IPAA.[28,37–40]

Burgess *et al.* specifically investigated whether DT cause more morbidity if they occur following in IPAA when compared to an IRA, finding that there was no difference in complications between the two groups. This finding suggests that presence or risk of desmoid tumour should not preclude IPAA.

Previous authors have stated that a proctectomy should be performed to prevent a scenario where a desmoid occurs after an IRA and blocks access to the rectum, preventing its later removal should this be necessary.[3] Guidelines suggest that the presence of intra-abdominal DT should prompt the surgeon to offer proctocolectomy and IPAA to alleviate concerns that a proctectomy and/or IPAA may not be able to be safely performed in the future.[3,41] The Cleveland Clinic experience suggests that this fear is unfounded. Between 1950 and 2008, 67 patients underwent a proctectomy and IPAA after prior IRA for FAP. Twenty-six had intra-abdominal DT. In no case did the desmoid disease affect the ability to perform a proctectomy, although in 13 cases the desmoid disease changed the surgical approach. In eight patients, desmoid disease precluded IPAA and five of these patients underwent EI instead. Two patients underwent pouch-low rectal anastomosis, while in one case there was no proctectomy performed as the mesenteric desmoid was excised and a new IRA performed.[42]

The authors believe that concern about a high desmoid tumour risk is a valid reason to perform an IRA and should not be an independent indication for a prophylactic proctectomy.

Does laparoscopic colectomy increase the risk of desmoid disease?

Prophylactic surgery for FAP is increasingly done though a minimally invasive approach.[43] A single institution experience published by Vitellaro *et al.* reviewed the outcomes of 602 patients undergoing open prophylactic colectomy, and 60 patients who underwent laparoscopic prophylactic colectomy. They demonstrated a marked reduction in desmoid tumour in laparoscopic group, estimating a rate of DT of 13% at 5 years following open compared with 5% in the laparoscopic colectomy group.[36] Noteworthy in this study is that the majority (57/73) of laparoscopic cases were to perform IRA, meaning that caution should be used in translating findings to the laparoscopic IPAA cohort of patients.

The Cleveland Clinic's data suggest that laparoscopic IRA is the least likely of all prophylactic procedures to result in desmoid tumour formation. Desmoid tumour occurred in 3.8% of patients who had undergone laparoscopic IRA versus 15.8% in the open IRA group, and desmoids occurred in 46.2% of patients who underwent laparoscopic IPAA versus 46.3% when open IPAA was performed. This suggests that laparoscopic colonic surgery reduces the risk of desmoid tumour, whilst laparoscopic rectal dissection and ileal pouch formation confers the greatest risk of desmoid tumour.[35]

Laparoscopic IPAA has been shown to be safe when compared with open surgery.[44] Larson *et al.* presented a case-matched series of 300 patients with 100 laparoscopic cases demonstrating earlier return of bowel function, reduced requirements for narcotics and equivalent complication rates.[45] The same author, using a validated survey tool to address functional outcomes including sexual dysfunction, cosmesis and quality of life, found comparable outcomes in both groups.[46] However, there were few FAP cases in that study and the wider applicability of laparoscopy to FAP patients remains to be tested. A systematic review of laparoscopic compared to open IPAA including patients with FAP or ulcerative colitis revealed no difference in mortality or complications between the groups and reoperation and readmission rates were similar, although operative times were significantly longer with laparoscopy.[47] As expected, cosmesis scores were considerably higher in the laparoscopic studies. Early case series have demonstrated that single incision laparoscopic IPAA can be performed safely by well-trained laparoscopic surgeons.[48]

Despite knowledge that laparoscopic colectomy and IRA is safe, cost effective and cosmetically appealing, there are limited outcome data, specifically for patients with FAP. An initial series of 16 FAP patients by Milsom *et al.* demonstrated good short-term outcomes with laparoscopic colectomy,[49] and this has been substantiated by a recent comparative series from St Mark's Hospital, where fewer complications were demonstrated in the laparoscopic group.[50]

In summary, prophylactic laparoscopic colectomy is safe when compared with open surgery and offers advantages of improved cosmesis and shorter return to bowel function. Laparoscopic rectal dissection and IPAA confers the highest risk of desmoid tumour and thus should be avoided. The authors practice is to perform laparoscopic TAC and IRA in cases of rectal preservation, and laparoscopic colectomy with open proctectomy and IPAA in cases when proctocolectomy is desired.

Discussion

Identification and management of patients with FAP can be challenging for clinicians. A series of management issues revolving around colorectal surgery have evolved and have been discussed here in the light of available evidence. Surgical strategy in FAP continues to evolve with more emphasis on maintaining quality of life. This requires thoughtful consideration of all factors impacting on each individual patient: their genotype, their family history, their extra-colonic manifestations, their comorbidities, their age and gender, and the stages they are at in their physical, social, academic and mental development. It is not just about a blanket approach of TPC for everyone. We hope that our review helps guide decision-making by clarifying these management issues.

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References

1. Kinzler KW, Nilbert MC, Su LK *et al.* Identification of FAP locus genes from chromosome 5q21. *Science* 1991; **253**: 661–5.
2. Arvanitis ML, Jagelman DG, Fazio VW, Lavery IC, McGannon E. Mortality in patients with familial adenomatous polyposis. *Dis. Colon Rectum* 1990; **33**: 639–42.
3. Vasen HFA, Moslein G, Alonso A *et al.* Guidelines for the clinical management of familial adenomatous polyposis (FAP). *Gut* 2008; **57**: 704–13.
4. Bisgaard ML, Fenger K, Bulow S, Niebuhr E, Mohr J. Familial adenomatous polyposis (FAP): frequency, penetrance, and mutation rate. *Hum. Mutat.* 1994; **3**: 121–5.
5. Hes FJ, Nielsen M, Bik EC *et al.* Somatic APC mosaicism: an underestimated cause of polyposis coli. *Gut* 2008; **57**: 71–6.
6. Jarvinen HJ. Epidemiology of familial adenomatous polyposis in Finland: impact of family screening on the colorectal cancer rate and survival. *Gut* 1992; **33**: 357–60.
7. Koskenvuo L, Pitkaniemi J, Rantanen M, Lepisto A. Impact of screening on survival in familial adenomatous polyposis. *J. Clin. Gastroenterol.* 2016; **50**: 40–4.
8. Grover S, Kastrinos F, Steyerberg EW *et al.* Prevalence and phenotypes of APC and MUTYH mutations in patients with multiple colorectal adenomas. *JAMA* 2012; **308**: 485–92.
9. Brennan MF. Pre-emptive surgery and increasing demands for technical perfection. *Br. J. Surg.* 2003; **90**: 3–4.
10. Parks AG, Nicholls RJ. Proctocolectomy without ileostomy for ulcerative colitis. *Br. Med. J.* 1978; **2**: 85–8.
11. Church J, Burke C, McGannon E, Pastean O, Clark B. Risk of rectal cancer in patients after colectomy and ileorectal anastomosis for familial adenomatous polyposis: a function of available surgical options. *Dis. Colon Rectum* 2003; **46**: 1175–81.
12. Church J, Burke C, McGannon E, Pastean O, Clark B. Predicting polyposis severity by proctoscopy: how reliable is it? *Dis. Colon Rectum* 2001; **44**: 1249–54.
13. Bussey HJ, Eyers AA, Ritchie SM, Thomson JP. The rectum in adenomatous polyposis: the St. Mark's policy. *Br. J. Surg.* 1985; **72**(Suppl.): S29–31.

14. Aziz O, Athanasiou T, Fazio VW *et al.* Meta-analysis of observational studies of ileorectal versus ileal pouch-anal anastomosis for familial adenomatous polyposis. *Br. J. Surg.* 2006; **93**: 407–17.
15. Slors FJ, van Zuijlen PP, van Dijk GJ. Sexual and bladder dysfunction after total mesorectal excision for benign diseases. *Scand. J. Gastroenterol. Suppl.* 2000: 48–51.
16. Olsen KØ, Juul S, Bülow S *et al.* Female fecundity before and after operation for familial adenomatous polyposis. *Br. J. Surg.* 2003; **90**: 227–31.
17. Gunther K, Braunrieder G, Bittorf BR, Hohenberger W, Matzel KE. Patients with familial adenomatous polyposis experience better bowel function and quality of life after ileorectal anastomosis than after ileoanal pouch. *Colorectal Dis.* 2003; **5**: 38–44.
18. Wu JS, Paul P, McGannon EA, Church JM. APC genotype, polyp number, and surgical options in familial adenomatous polyposis. *Ann. Surg.* 1998; **227**: 57–62.
19. Brown SR, Donati D, Seow-Choen F. Rectal cancer after mucosectomy for ileoanal pouch in familial adenomatous polyposis: report of a case. *Dis. Colon Rectum* 2001; **44**: 1714–5.
20. Remzi FH, Church JM, Bast J *et al.* Mucosectomy vs. stapled ileal pouch-anal anastomosis in patients with familial adenomatous polyposis: functional outcome and neoplasia control. *Dis. Colon Rectum* 2001; **44**: 1590–6.
21. Ooi BS, Remzi FH, Gramlich T, Church JM, Preen M, Fazio VW. Anal transitional zone cancer after restorative proctocolectomy and ileoanal anastomosis in familial adenomatous polyposis: report of two cases. *Dis. Colon Rectum* 2003; **46**: 1418–23; discussion 22–3.
22. von Roon AC, Will OC, Man RF *et al.* Mucosectomy with handsewn anastomosis reduces the risk of adenoma formation in the anorectal segment after restorative proctocolectomy for familial adenomatous polyposis. *Ann. Surg.* 2011; **253**: 314–7.
23. Wasmuth HH, Tranø G, Myrvold HE, Aabakken L, Bakka A. Adenoma formation and malignancy after restorative proctocolectomy with or without mucosectomy in patients with familial adenomatous polyposis. *Dis. Colon Rectum* 2013; **56**: 288–94.
24. Lovegrove RE, Constantinides VA, Heriot AG *et al.* A comparison of hand-sewn versus stapled ileal pouch anal anastomosis (IPAA) following proctocolectomy. *Ann. Surg.* 2006; **244**: 18–26.
25. Hartley JE, Church JM, Gupta S, McGannon E, Fazio VW. Significance of incidental desmoids identified during surgery for familial adenomatous polyposis. *Dis. Colon Rectum* 2004; **47**: 334–8; discussion 9-40.
26. Gurbuz AK, Giardiello FM, Petersen GM *et al.* Desmoid tumours in familial adenomatous polyposis. *Gut* 1994; **35**: 377–81.
27. Sturt NJ, Gallagher MC, Bassett P *et al.* Evidence for genetic predisposition to desmoid tumours in familial adenomatous polyposis independent of the germline APC mutation. *Gut* 2004; **53**: 1832–6.
28. Bertario L, Russo A, Sala P *et al.* Genotype and phenotype factors as determinants of desmoid tumors in patients with familial adenomatous polyposis. *Int. J. Cancer* 2001; **95**: 102–7.
29. Slowik V, Attard T, Dai H, Shah R, Septer S. Desmoid tumors complicating Familial Adenomatous Polyposis: a meta-analysis mutation spectrum of affected individuals. *BMC Gastroenterol.* 2015; **15**: 84.
30. Nieuwenhuis MH, Lefevre JH, Bülow S *et al.* Family history, surgery, and APC mutation are risk factors for desmoid tumors in familial adenomatous polyposis: an international cohort study. *Dis. Colon Rectum* 2011; **54**: 1229–34.
31. Church J, Xhaja X, LaGuardia L, O'Malley M, Burke C, Kalady M. Desmoids and genotype in familial adenomatous polyposis. *Dis. Colon Rectum* 2015; **58**: 444–8.
32. Lotfi AM, Dozois RR, Gordon H *et al.* Mesenteric fibromatosis complicating familial adenomatous polyposis: predisposing factors and results of treatment. *Int. J. Colorectal Dis.* 1989; **4**: 30–6.
33. Durno C, Monga N, Bapat B, Berk T, Cohen Z, Gallinger S. Does early colectomy increase desmoid risk in familial adenomatous polyposis? *Clin. Gastroenterol. Hepatol.* 2007; **5**: 1190–4.
34. Elayi E, Manilich E, Church J. Polishing the crystal ball: knowing genotype improves ability to predict desmoid disease in patients with familial adenomatous polyposis. *Dis. Colon Rectum* 2009; **52**: 1762–6.
35. Vogel J CJ, LaGuardia L. Minimally invasive pouch surgery predisposes to desmoid tumor formation in patients with familial adenomatous polyposis. In: *The American Society of Colon and Rectal Surgeons Annual Meeting Abstracts, April 30–May 5, 2005, Philadelphia, Pennsylvania*. Philadelphia: Dis. Colon Rectum, 2005; 662.
36. Vitellaro M, Sala P, Signoroni S *et al.* Risk of desmoid tumours after open and laparoscopic colectomy in patients with familial adenomatous polyposis. *Br. J. Surg.* 2014; **101**: 558–65.
37. Nieuwenhuis MH, De Vos Tot Nederveen Cappel W, Botma A *et al.* Desmoid tumors in a dutch cohort of patients with familial adenomatous polyposis. *Clin. Gastroenterol. Hepatol.* 2008; **6**: 215–9.
38. Lefevre JH, Parc Y, Kernéis S *et al.* Risk factors for development of desmoid tumours in familial adenomatous polyposis. *Br. J. Surg.* 2008; **95**: 1136–9.
39. Sinha A, Tekkis PP, Neale KF, Phillips RKS, Clark SK. Risk factors predicting intra-abdominal desmoids in familial adenomatous polyposis: a single centre experience. *Tech. Coloproctol.* 2010; **14**: 141–6.
40. Heiskanen I, Jarvinen HJ. Occurrence of desmoid tumours in familial adenomatous polyposis and results of treatment. *Int. J. Colorectal Dis.* 1996; **11**: 157–62.
41. Burgess A, Xhaja X, Church J. Does intra-abdominal desmoid disease affect patients with an ileal pouch differently than those with an ileorectal anastomosis? *Dis. Colon Rectum* 2011; **54**: 1388–91.
42. Church JM, Xhaja X, Warrier SK *et al.* Desmoid tumors do not prevent proctectomy following abdominal colectomy and ileorectal anastomosis in patients with familial adenomatous polyposis. *Dis. Colon Rectum* 2014; **57**: 343–7.
43. da Luz MA, Church JM, Burke CA. The evolution of prophylactic colorectal surgery for familial adenomatous polyposis. *Dis. Colon Rectum* 2009; **52**: 1481–6.
44. Vitellaro M, Bonfanti G, Sala P *et al.* Laparoscopic colectomy and restorative proctocolectomy for familial adenomatous polyposis. *Surg. Endosc.* 2010; **25**: 1866–75.
45. Larson DW, Cima RR, Dozois EJ *et al.* Safety, feasibility, and short-term outcomes of laparoscopic ileal-pouch-anal anastomosis. *Ann. Surg.* 2006; **243**: 667–72.
46. Larson DW, Davies MM, Dozois EJ *et al.* Sexual function, body image, and quality of life after laparoscopic and open ileal pouch-anal anastomosis. *Dis. Colon Rectum* 2008; **51**: 392–6.
47. Ahmed Ali U, Keus F, Heikens JT *et al.* Open versus laparoscopic (assisted) ileo pouch anal anastomosis for ulcerative colitis and familial adenomatous polyposis. *Cochrane Database Syst. Rev.* 2009; CD006267.
48. Geisler DP, Kirat HT, Remzi FH. Single-port laparoscopic total proctocolectomy with ileal pouch-anal anastomosis: initial operative experience. *Surg. Endosc.* 2011; **25**: 2175–8.
49. Milsom JW, Ludwig KA, Church JM, Garcia-Ruiz A. Laparoscopic total abdominal colectomy with ileorectal anastomosis for familial adenomatous polyposis. *Dis. Colon Rectum* 1997; **40**: 675–8.
50. McNicol FJ, Kennedy RH, Phillips RKS, Clark SK. Laparoscopic total colectomy and ileorectal anastomosis (IRA), supported by an enhanced recovery programme in cases of familial adenomatous polyposis. *Colorectal Dis.* 2012; **14**: 458–62.