

Impact of familial adenomatous polyposis on young adults: Quality of life outcomes

Author/Contributor:

Andrews, Leslie; Mireskandari, Shab; Jessen, J; Thewes, Belinda; Solomon, Michael; Macrae, F; Meiser, Bettina

Publication details:

Disease of the colon and rectum
v. 50
pp. 1306-1315
0012-3706 (ISSN)

Publication Date:

2007

Publisher DOI:

<http://dx.doi.org/10.1007/s10350-007-0259-9>

License:

<https://creativecommons.org/licenses/by-nc-nd/3.0/au/>

Link to license to see what you are allowed to do with this resource.

Downloaded from <http://hdl.handle.net/1959.4/38960> in <https://unsworks.unsw.edu.au> on 2022-06-29

Impact of familial adenomatous polyposis (FAP) on young adults:

Quality of life outcomes

Andrews L,¹ Mireskandari S,^{2,3} Jessen J,⁴ Thewes B,²

Solomon M,⁵ Macrae F,⁶ Meiser B^{2,7}

¹ Hereditary Cancer Clinic, Prince of Wales Hospital, Sydney, Australia

² Psychosocial Research Group, Prince of Wales Hospital, Sydney, Australia

³ Prince of Wales Clinical School, University of New South Wales, Sydney, Australia

⁴ School of Biomedical Science, Faculty of Medicine, University of Newcastle, Newcastle, Australia

⁵ Discipline of Surgery, University of Sydney, Sydney, Australia

⁶ Department of Colorectal Medicine and Genetics, The Royal Melbourne Hospital, Australia

⁷ School of Psychiatry, University of New South Wales, Sydney, Australia

Correspondence to: Dr Lesley Andrews, Hereditary Cancer Clinic, Prince of Wales Hospital, Randwick, NSW 2031, Australia, Tel 0061-2-9382-2551, Fax 0061-2-9382-2588, e-mail: lesley.andrews@sesiahs.health.nsw.gov.au

Running title: Quality of life in young adults with FAP

Key words: familial adenomatous polyposis, quality of life, impact of prophylactic surgery

ABSTRACT

PURPOSE: This study utilized a novel questionnaire to assess quality of life and psychological adjustment amongst young adults aged 18 to 35 years old with a diagnosis of, or at risk of developing Familial Adenomatous Polyposis (FAP). **METHODS:** Eighty-eight participants (25 males, 63 females) were recruited through four Australian Hereditary Bowel Cancer Registries. **RESULTS:** The average age of participants was 28 years and the average age of these participants at the time of their last genetic consultation was 23 years. Seventy-one participants (81%) had clinical FAP, of whom 57 had undergone either an ileorectal anastomosis (IRA) or formation of an ileal pouch with anal anastomosis (IPAA) to prevent colorectal cancer. The IPAA group had significantly more adverse outcomes for physical functioning, body image, sexual impact and negative affect compared to the no-surgery group; and significantly more negative outcomes for physical functioning and negative affect compared to the IRA group. Amongst the total sample, a small proportion (11.4%) had avoidance scores indicative of a significant stress response, and being single was associated with higher levels of avoidance responses about FAP ($z=-3.19, p=0.001$). **CONCLUSIONS:** FAP may have a negative impact across a broad range of life domains. Being single is an important risk factor for adverse psychological outcomes. Timing of surgery and choice of surgery may minimize the negative impact on physical and psychological functioning. Psychological interventions may be required for a small proportion of those affected by FAP, and ongoing access to genetic services may help to identify and address the needs of this group.

BACKGROUND

Familial Adenomatous Polyposis (FAP) is a hereditary cancer syndrome with the onset of multiple adenomatous polyps in the colon typically occurring in adolescence. Children in families with an identified APC gene mutation are offered predictive testing at around the age of puberty. For those identified with a mutation, or those at 1 in 2 risk in a family where the mutation has not been identified, regular colonic surveillance with sigmoidoscopy or colonoscopy is recommended from around the same age. Prophylactic colectomy with either an ileorectal anastomosis (IRA) or formation of an ileal pouch with anal anastomosis (IPAA) is performed to prevent colorectal cancer, typically by the middle of the third decade. A temporary ileostomy may be required postoperatively. Rarely, this may be permanent.

Patients with an IRA have an average of 3-4 bowel movements per day, and 72-98% are continent, during the day.¹ Those with an ileal pouch have an average of 4-5 bowel movements daily, and 60-98% are continent during the day.¹ While these figures may be acceptable outcomes from the surgical perspective, little is known about their impact on adolescents and young adults who are at a critical stage of maturation and development, particularly with regard to sexual identity. Several studies have compared the functional impact of IRA to that of IPAA amongst young adults with FAP (see Soravia¹ for a summary of findings). Those studies that assessed quality of life essentially found that quality of life and/or patient satisfaction were similar for the two procedures; however in these studies quality of life was measured using generic, one-item measures of quality of life with unknown psychometric properties.¹⁻³ Two other studies compared the impact on quality of life of IRA to that of IPAA amongst patients with FAP unselected for age and found no differences between groups, using generic rather than disease-specific quality of life measures.⁴

Attitudes and psychosocial adjustment to FAP

There are only limited studies on attitudes and psychosocial adjustment to FAP,⁵ and psychosocial impact of cancer in young adults.⁶⁻⁸ A study by Miller et al.⁵ compared psychosocial adjustment of young adults to that of older adults who had FAP and had undergone bowel surgery. Overall scores of affect and well-being were lower than those reported by healthy community samples in normative studies.³ Younger participants (aged 40 years or less) were significantly more likely to report negative affect scores, compromised attractiveness as a result of surgery, and anger at their diagnosis compared to older participants.⁵ This study did not explore needs for support or information, but interestingly found that those with more accurate knowledge of their condition experienced significantly lower fear about future health, were significantly more likely to have positive affect scores and were happier about their health. These findings suggest that education about FAP is associated with positive impact on psychosocial outcomes.

While this study provided some important insights into the experience of people with FAP, several shortcomings need to be acknowledged. The results of this study were largely descriptive and it included adults of all ages, with limited exploration of the needs of younger participants. Furthermore, as the study was conducted almost 20 years ago, much has changed in regards to the surgical treatment and clinical management of FAP, especially since the advent of genetic testing for individuals who are at risk. Although the study aimed to explore the impact of FAP on emotional status and well-being, psychological questionnaires with better psychometric properties than those used in this study are now available.

Psychosocial Impact of Cancer on Young Adults

It is well documented that younger people have a significantly higher risk of poor psychological adaptation to chronic illnesses,⁶ cancers,⁷ and hereditary cancer.^{5,8} Poor psychological adaptation of younger people, compared with older adults, may be a result of limited experience in developing coping skills to deal with adversity, a societal expectation that illness and disability are associated with advancing age, or the developmental stressors associated with adolescence and young adulthood. Freeman has identified five developmental issues that confront the adolescent including: the need to develop a sense of identity, the need to differentiate themselves from parents and other authority figures, the need to develop satisfying peer relationships, courtship and the need to develop sexual relationships, and career planning.⁹ A diagnosis of FAP, the need for regular bowel screening, and surgical procedures during adolescence have the potential to interfere with these developmental tasks, yet to date this relationship has not been systematically studied. The nature and treatment of FAP also brings with it many challenges to body image and self-esteem, at a time when young people are developing a sense of self and may already be experiencing a precarious sense of self-esteem and body image.

The limited literature available suggests that further research is required to determine more specific impacts of FAP surgery on various life domains. Such data is important in optimizing pre-surgical and genetic counseling, with regard to both timing and content with a view to improving emotional adjustment. The current study aims to assess the impact of being diagnosed with FAP or being at 1 in 2 risk, on a number of life domains, and explore the impact of different surgical procedures and sociodemographic variables on psychological adjustment.

MATERIALS AND METHODS

This study was undertaken as part of a larger study, in which attitudes to genetic testing, support and information needs and experiences of discrimination were also assessed.¹⁰ Eligible participants were individuals aged between 18–35 years with either a clinical or a genetic diagnosis of FAP, or at 1 in 2 risk of developing FAP based on family history. Participants were identified through four Australian Hereditary Bowel Cancer Registries.

Potentially eligible individuals were initially contacted by a letter from their registry, inviting them to participate in the study. Individuals were asked to return an enclosed preference card to indicate their interest in participation. The research team telephoned individuals who agreed to be included in the study, to ensure their eligibility and confirm mailing details. Packages including a questionnaire, consent form and a reply-paid envelope were then mailed out to these participants. Reminder calls were made and replacement questionnaires were mailed as necessary. To assess test-retest reliability of the disease-specific novel quality of life measures, a second questionnaire was mailed two weeks after the first questionnaire to all those participants who had completed the first questionnaire.

Measures

A questionnaire was purposively designed, based on the review of relevant literature and also a previous qualitative study involving in-depth interviews with young adults with or at risk for FAP, the results of which will be reported separately. The following measures were administered:

Demographic characteristics. Information regarding age, sex, marital status, and education level were collected. In addition, individuals were asked whether they had children presently and if they planned to have children (or more children) in the future.

Medical/family history. Details of FAP diagnosis, genetic counseling, genetic testing and genetic testing results were collected. In addition, FAP family history details were collected, which included the first person in the family diagnosed with FAP, family genetic testing results, as well as deaths from bowel cancer in the family. Data on screening type and frequency as well as any information on FAP-related surgery was also collected.

Quality of life measure. The Quality of Life measure consists of six subscales: (i) Surgery-related impact (7 items); (ii) Body image (3 items); (iii) Sexual impact (4 items, 2 which are unique to females and/or males); (iv) Negative affect (3 items); and (v) the Physical functioning (10 items) and (vi) Role-emotional subscales (3 items). Scales (i) to (v) were purposively designed for this study and subjected to an analysis of internal and test-retest reliability to ascertain their psychometric properties. With the exception of the Sexual Impact subscale, all subscales were found to have excellent internal consistency and test-retest reliability with the following Cronbach's alpha levels and test-retest correlations: (i) Surgery-related impact ($\alpha = 0.74$, $r=0.60$); (ii) Body image ($\alpha = 0.80$, $r=0.72$); and (iv) Negative affect ($\alpha = 0.76$, $r=0.75$). With regard to the Sexual Impact subscale, test-retest correlation for the female and the male versions were $r=0.86$ and $r=0.64$ respectively. Cronbach's α amongst the female version of the scale was excellent ($\alpha = 0.74$), while Cronbach's α amongst males was poor ($\alpha = 0.22$). There was no variance amongst responses, with all male participants reporting that FAP had not at all affected erection and/or ejaculation; it is a feature of correlations in general that if the range

of the variables is limited, correlations tend to be low. Subscales (v) and (vi) were taken from the MOS 36-item Short-Form Health Survey (SF-36).¹¹

Intrusive and avoidance responses about FAP. The Impact of Event Scale (IES)¹² was used to measure (i) intrusive and (ii) avoidance responses about FAP, that is (i) involuntary thoughts, troubled dreams, strong waves of feelings and (ii) denial of the meanings of the event, blunted sensation, behavioral inhibition, respectively. This 15-item scale is a validated measure of responses in relation to a specific stressor (in this case concern about either having been diagnosed with or being at risk of developing FAP).

Life Impact. Seven five-point Likert-scales items (ranging from ‘Very positively’ to ‘Very negatively’) were specifically designed for this study to assess the overall impact of a diagnosis or being at risk of FAP on education, employment, friendships and social activities, relationships with family members, intimate relationships, attitudes towards long-term relationships or marriage, and attitudes towards having children. The scale was found to have good internal consistency with a Cronbach’s alpha level of ($\alpha = 0.65$) and excellent test-retest reliability ($\alpha = 0.79$).

Statistical analysis

Data was analyzed using SPSS 12.0 (Statistical Program for the Social Sciences). Descriptive statistics were used to describe the sample in terms of sociodemographic, clinical and psychological characteristics. For the newly developed Quality of Life and Life Impact of FAP scales, test-retest correlations and Cronbach’s alpha were calculated. For the Quality of Life subscales, scores were summed within scales and re-scaled from 0 to 100, so that a high scale score represents a higher response level and/or less symptoms. Bivariate tests were carried out to

assess associations between age, sex, and surgery and marital status, on the one hand, and intrusive and avoidance responses about FAP, Quality of Life and Life Impact of FAP, on the other. Due to the skewed distribution of the continuous quality of life and psychological outcome variables, non-parametric Mann Whitney *U* tests were used for the predictor variables age, sex and marital status. Kruskal Wallis tests were performed to compare the three surgery groups (IRA, IPAA and no-surgery), followed by Mann Whitney *U* tests for significant Kruskal Wallis tests.

RESULTS

Sociodemographic and medical characteristics

Of 265 registrants sent invitations to participate, 108 were lost to contact and 11 were found to be ineligible. Of the remaining 146 registrants successfully contacted, 116 consented to be contacted and, of those, 88 returned completed questionnaires, resulting in an overall participation rate of 60.3%. Sixty-one participants also returned the second questionnaire.

Table 1 provides a summary of the sociodemographic and medical characteristics of participants. Twenty-five participants were male (28.4%) and 63 (71.6%) were female. Twenty-eight participants (32.9%) were single, and 57 (67.1%) married or in a *de facto* relationship. The average age of participants was 27.6 years (SD=4.9, range=18-35 years). Seventy-one participants (80.7%, 22 males and 49 females) were clinically affected by FAP and 17 (19.3%, 3 males and 14 females) were clinically unaffected by FAP. Of the 71 participants clinically affected by FAP, only 14 (15.9%) had not undergone surgery. Thirty-three participants had had a total colectomy and IRA, and 22 a restorative proctocolectomy with IPAA. Fifty-four

participants (63.5%) had consulted with a genetic counselor or clinical geneticist regarding FAP. The average age at the last genetic consultation was 22.6 years (SD=6.4, range=8-34 years).

[Insert Table 1 about here]

Physical impact of surgery

Table 2 demonstrates the number and percentages of participants reporting at least some degree of negative impact on physical functioning separately for those who underwent IRA and IPAA. It shows that 72.7% of participants who had undergone IRA reported frequent bowel movements within the past 4 weeks, compared to 100.0% of those who had undergone IPAA.

Body image and sexual functioning

Table 2 also shows the number of participants reporting at least some degree of negative impact on several aspects of body image and sexual functioning. FAP or its treatment had a negative impact on perceived physical attractiveness in one quarter (25.0%) of all participants, in 20.2% on their feelings of femininity or masculinity, in 26.2% on their sexual desire, in 23.8% on sexual enjoyment, and 51.2% were dissatisfied with their body. While men did not report any limitations in regards to achieving and maintaining an erection or ejaculation, 30.0% of women did report experiencing pain during intercourse and 14.3% reported difficulty to orgasm.

[Insert Table 2 about here]

Psychological Adjustment

Scores greater than 20 on each of the IES subscales are indicative of a significant stress response in relation to having FAP or being at risk of developing FAP. Accordingly, 4.5% and 11.4% of

participants had scores on the intrusion and avoidance subscales of the IES respectively indicative of a significant stress response.

Life impact of FAP

Ten percent (10.3%) reported that FAP had affected their education either ‘somewhat’ or ‘very’ negatively; 18.2% their friendships and social activities; 10.3% their relationships with other family members; 18.1% their intimate relationships and 10.6% their attitudes towards long-term relationships or marriage. Twenty-six percent (25.9%) of participants reported being less or not at all willing to have children as a result of their being at risk of developing or being diagnosed with FAP.

Comparisons of quality of life and psychosocial outcomes

Bivariate analyses explored associations between dependent variables (quality of life, life impact and psychological outcomes), on the one hand, and each of the potential predictors and confounding variables (surgery status, age, sex and marital status), on the other. With regard to age and sex, no statistically significant associations were found with any of the outcome variables, except that men showed a greater surgery-related impact than women ($z = -2.68, p = 0.007$).

Table 3 presents a summary of the results of statistical tests comparing intrusion and avoidance responses about FAP, Quality of Life and Life Impact of FAP outcomes scores by surgery group. Kruskal Wallis tests showed significant surgery group differences for Physical functioning ($\chi^2=9.2, p=0.010$); Body image ($\chi^2=23.2, p<0.001$); Sexual impact ($\chi^2=11.2, p=0.004$) and Negative affect ($\chi^2=6.6, p=0.037$). With regard to Body image, Sexual impact and Negative

affect, Table 3 shows that functioning is highest for the no-surgery group, followed by the IRA group, with functioning being lowest for the IPAA group. No significant differences were observed for comparisons between the IRA group and the no-surgery group. Mann Whitney *U* tests show significant differences between the IPAA group compared to the no-surgery group for all four outcome variables; and significant differences between the IPAA group compared to the IRA group for Physical functioning and Negative affect.

[Insert Table 3 about here]

Table 4 shows the results of bivariate analyses between marital status and all outcome variables. Significant differences were found for marital status and avoidance responses about FAP ($z = -3.18, p=0.001$), with participants who were single showing significantly higher levels of FAP avoidance responses.

[Insert Table 4 about here]

DISCUSSION

This study used a novel, FAP-specific quality of life measure and consequently was not designed to provide quality of life data compared to general population norms. It should be noted though that the mean scores on the Physical functioning and Role-emotional functioning subscales are very similar to those found amongst the general Australian population,¹³ indicating that these aspects of quality of life as measured by a generic measure do not appear to be compromised. However, findings using the measures of FAP-specific quality of life demonstrate the extensive domains of psychosocial functioning that may be adversely affected by FAP. The unique aspect

of this study has been the combination of multiple items to develop an FAP-specific questionnaire relating to six domains – education, social functioning, family relationships, intimate relationships, long-term relationships and childbearing, combined to provide overall impact scores. Such a tool will be useful for future studies for comparison of FAP-related interventions and treatment. This study provided preliminary data on the reliability of the FAP-specific questionnaire, however further validation work involving a larger sample would be warranted in any future studies using this instrument.

Diagnosis and surgery for FAP typically occur in late adolescence or early adulthood at a time when sexuality and sexual relationships are being established. In this study of young adults, those who had undergone an IPAA had more adverse physical functioning, body image, sexual impact and negative affect compared to the no-surgery group; and lower physical functioning and negative affect compared to those who had an IRA. This has important implications for the timing and choice of surgery. The finding that being single is associated with higher levels of distress is also relevant to the timing of elective surgery. This study provides some evidence for considering IRA as a first procedure in young adults, with a second procedure to remove the rectum being deferred until later ages when the impact may be less, particularly if the patient is not in a long-term relationship.

Our findings demonstrate differences in the impact on quality of life of IRA versus IPAA and are in contrast to results from previous studies that have used generic quality of life measures.¹⁻⁴ We used a disease-specific measure, which may be potentially more sensitive. Our findings require replication in samples with different patient characteristics and further investigation to determine the basis of the lower functioning and potential for improvement.

While no male participants in the current study reported difficulties with sexual functioning, 3% of all females reported pain with intercourse, with a smaller proportion experiencing difficulty with orgasm. While the reasons for these gender differences may be physiological, they may also represent a more adverse psychological impact of surgery on females. Post-operative counseling for females may improve adjustment. Additionally, if a more negative sexual impact is found to be related to body image due to large incisions and resultant scars, utilization of minimally invasive techniques to reduce scarring may result in better psychological outcomes.

The mean levels of intrusion and avoidance responses were comparable to those found in other studies involving individuals at risk for hereditary cancer (e.g.¹⁴). While a minority reported intrusive thoughts about FAP indicative of a significant stress response, a greater percentage demonstrated clinically significant levels of avoidance ideation. This aspect of distress was not associated with previous surgery, and may be amenable to psychological interventions. As almost 40% of participants had never consulted a genetics service, and of those who did, the average time since the last consultation was on average five years ago, a larger study may be able to determine if genetic counseling could reduce the high level of distress experienced by this subgroup. These findings highlight the weakness in reporting mean levels of distress, which may disguise significant distress in a small number of individuals. Such individuals could be targeted for counseling to reduce stress if protocols were in place to identify those in need of these interventions.

An additional analysis demonstrated a significant association between marital status and avoidance responses about FAP. Single people in the current sample had higher mean levels of avoidance responses about FAP than those who were married. This result corroborates findings

from a related study that found better health-related quality of life amongst married adults diagnosed with FAP and desmoid tumour.¹⁵ Taken together, these findings demonstrate that single young adults with FAP are more vulnerable and in greater need of psychological interventions.

The conclusion of this study is that there is a minority of individuals with high levels of FAP related distress affecting multiple aspects of their lives, and that undergoing surgery and being single are important risk factors for adverse psychological and sexual outcomes. These findings indicate that genetic and supportive counseling services should be available at the time of diagnosis, as well as before and after surgery. Follow up procedures should be available at all clinics to ensure that these individuals are offered support as they pass through different life stages and need to address different problems.

The retrospective design of this study of young adults provides some insights into the cumulative impact of FAP from adolescence. Whilst the results of this study provide some insights into the impact of FAP on young adults, it raises many additional questions and highlights the need for further research. A prospective multi-centre study of adolescents would provide much needed data on the impact of FAP and the optimal psychosocial management amongst this population.

Acknowledgments

We are most grateful for the valuable contribution of all those participated in this study. This study has been funded by the Cure Cancer Foundation of Australia. The authors would like to thank the following individuals for their assistance with participant ascertainment and

recruitment: NSW and ACT Hereditary Cancer Registers, especially Freddy Sitas, Sarah Doak and Udani Abeyapala; Queensland Familial Bowel Cancer Registry, especially Ngaire Knight and Tricia Keevers; South Australia Familial Cancer Unit, especially Dr Graeme Suthers; and the Victorian Family Cancer Register, especially Marita Black, Toulia McArdle and Kate Minkoff. Bettina Meiser is supported by a Career Development Award from The National Health and Medical Research Council of Australia (ID 350989).

Table 1. Summary Characteristics of Participants (N=88)

Variable	Surgery (N=57) N (%)	No-surgery (N=31) N (%)	Total Sample (N=88) N (%)
Age (years):			
18-23	15 (26.3)	7 (23.3)	22 (25.3)
24-29	18 (31.6)	11 (36.7)	29 (33.3)
30-35	24 (42.1)	12 (40.0)	36 (41.4)
Sex:			
Male	18 (31.6)	7 (22.6)	25 (28.4)
Female	39 (68.4)	24 (77.4)	63 (71.6)
Marital Status:			
Single	18 (31.6)	10 (33.3)	28 (32.2)
<u>Separated/divorced</u>	<u>1 (1.8)</u>	<u>1 (3.3)</u>	<u>2 (2.3)</u>
Married/De Facto	38 (66.7)	19 (63.3)	57 (65.5)
Children:			
No	30 (52.6)	17 (56.7)	47 (54.0)
Currently Pregnant	1 (1.8)	2 (6.7)	3 (3.4)
Yes	26 (45.6)	11 (36.7)	37 (42.5)
FAP Status:			
Clinically Unaffected (N=17)			
Mutation carrier	0 (0.0)	2 (11.8)	2 (11.8)
At 1 in 2 Risk	0 (0.0)	15 (88.2)	15 (88.2)
Clinically Affected (N=71)			
Had surgery	57 (100.0)	-	57 (81.4)
No surgery yet	-	14 (100.0)	14 (19.7)
Type of surgery (N=57):			
Total Colectomy & IRA	33 (57.9)	-	33 (57.9)
Restorative Proctocolectomy with IPAA	16 (28.1)	-	16 (28.1)
Proctocolectomy & ileostomy	1 (1.8)	-	1 (1.8)
Colectomy & IRA and later proctocolectomy & IPAA	5 (8.8)	-	5 (8.8)
Don't know what type surgery	2 (3.5)	-	2 (3.5)

- Deleted: 9
- Deleted: 3
- Deleted: 4
- Deleted: 1
- Deleted: 6
- Deleted: 6
- Deleted: 30
- Deleted: 4
- Deleted: 5

Table 2. Number of participants reporting negative impact on several aspects of physical functioning, body image and sexual functioning^a

Scale	Ileorectal Anastomosis (N=33)		Ileal Pouch with Anal Anastomosis (N=21)		No-surgery (N=31)		Total sample ^c (N=85)	
	N	%	N	%	N	%	N	%
<i>Physical functioning</i>								
Frequent bowel motions	24	72.7	17	100.0	N/A	N/A		
Loss of bowel control	6	20.0	5	27.8	N/A	N/A		
Staining of underwear	4	14.3	5	29.4	N/A	N/A		
Abdominal bloating	10	40.0	6	54.5	N/A	N/A		
Excessive wind	9	39.1	5	55.6	N/A	N/A		
Problems with urinations	25	75.8	13	68.4	N/A	N/A		
<i>Body image</i>								
Physical attractiveness	6	18.2	13	65.0	2	6.5	21	25.0
Less feminine/masculine	4	12.1	9	45.0	4	12.9	17	20.2
Body dissatisfaction	18	90.0	15	45.5	10	32.3	43	51.2
<i>Sexual functioning</i>								
Sexual desire	10	30.3	11	55.0	1	3.2	22	26.2
Sexual enjoyment	9	27.3	10	50.0	1	3.2	20	23.8
Erection (male)	0	0.0	0	0.0	0	0.0	0	0.0
Ejaculation (male)	0	0.0	0	0.0	0	0.0	0	0.0
Intercourse Pain (female) ^b	6	30.0	8	66.7	1	5.6	15	30.0
Orgasm ability (female) ^b	3	16.7	4	30.8	0	0.0	7	14.3

^a Five-point Likert-type scales were used with the following response options: 'Not at all', 'A little bit', 'Moderately', 'Quite a bit' and 'Extremely'. Participants were categorized into those reporting no adverse effects versus those reporting at least some degree of adverse effects.

^b Women who reported 'not applicable' were omitted from the calculation of percentages. ^c Participants who reported that they did not know what surgery they had (2) and/or that they had a proctocolectomy and ileostomy (1) were excluded from the analysis. Those reporting a colectomy & IRA and later proctocolectomy & IPAA (5) were included in the IPAA group.

Table 3. Comparisons of Quality of Life, Life Impact of FAP and FAP-specific distress outcomes scores by surgery group

Scale	Ileorectal Anastomosis (N=33)		Ileal Pouch with Anal Anastomosis (N=21)		No-surgery (N=31)		Total sample ^d (N=85)	
	M	SD	M	SD	M	SD	M	SD
<i>Intrusive and avoidance responses about FAP^a</i>								
Intrusive responses about FAP	4.8	6.6	5.6	6.4	3.9	5.5	4.7	6.1
Avoidance responses about FAP	5.6	7.3	7.8	8.5	7.2	10.4	6.7	8.8
<i>Quality of Life^b</i>								
Physical impact of surgery	78.4	15.9	72.5	15.4	N/A	N/A	N/A	N/A
Physical functioning	96.1	7.9	88.1	16.5	90.0	23.2	91.9** [†]	17.1
Body image	89.9	16.1	72.1	23.0	94.1	9.4	87.2*	18.1
Sexual impact	86.2	21.6	77.5	26.2	91.0	19.0	85.8** [†]	22.2
Negative affect	79.7	23.5	70.2	20.7	82.8	16.4	78.5*	20.7
Role-emotional Functioning	88.9	28.5	76.7	34.4	83.9	29.7	84.1	30.4
<i>Life Impact of FAP^c</i>								
	2.8	0.4	3.0	0.7	2.8	0.3	2.8	0.4

^a Range 0 to 35 for the Intrusion Subscale and 0 to 40 for the Avoidance Subscale, with higher scores indicating more intrusive and/or avoidance responses. ^b Scales range in score from 0 to 100, and a high scale score represents a higher response level and/or lower level of symptoms. ^c A high scale score represents a more negative impact of FAP (range 1 ['Very positively'] to 5 ['Very negatively']). ^d Participants who reported that they did not know what surgery they had (2) and/or that they had a proctocolectomy and ileostomy were excluded from the analysis. Those reporting a colectomy & IRA and later proctocolectomy & IPAA (5) were included in the IPAA group. * $p < 0.05$ differences between the IPAA group compared to the no-surgery group. ** $p < 0.01$ differences between the IPAA group compared to the no-surgery group. [†] $p < 0.05$ differences between the IPAA group compared to IRA group.

Table 4. Comparisons of Quality of Life, Life Impact of FAP and FAP- Specific Distress Outcome Scores by Marital Status

Scale	Married (N=57)		Single (N=30) ^d		Total sample (N=88)		p value
	M	SD	M	SD	M	SD	
<i>Intrusive & avoidance responses about FAP^a</i>							
Intrusive responses about FAP	3.7	5.2	6.1	7.3	4.6	6.0	0.22
Avoidance responses about FAP	4.5	6.4	11.1	11.1	6.8	8.8	0.001
<i>Quality of Life^b</i>							
Surgery-related impact	74.5	15.1	82.8	16.2	78.1	15.9	0.06
Physical functioning	92.5	15.8	91.2	19.2	92.1	16.9	0.54
Body image	88.1	18.0	85.3	18.5	87.3	18.3	0.48
Sexual impact	88.6	19.2	81.3	26.4	81.6	24.4	0.25
Negative affect	77.7	20.4	79.2	21.3	78.4	20.5	0.51
Role-emotional functioning	82.5	32.2	88.5	25.6	84.7	30.0	0.42
<i>Life Impact of FAP^c</i>	2.9	0.5	2.8	0.4	2.9	0.4	0.45

^a Range 0 to 35 for the Intrusion Subscale and 0 to 40 for the Avoidance Subscale, with higher scores indicating more intrusive and/or avoidance responses. ^b Scales range in score from 0 to 100, and a high scale score represents a higher response level and/or lower level of symptoms. ^c A high scale score represents a more negative impact of FAP (range 1 ['Very positively'] to 5 ['Very negatively']). ^d Two separated/divorced participants are included in this group.

Deleted: 28

Formatted: Superscript

Formatted: Font: 8 pt

References

1. Soravia C, Klein L, Berk T, O'Connor B, Cohen Z, McLeod R. Comparison of ileal pouch-anal anastomosis and ileorectal anastomosis in patient with familial adenomatous polyposis. *Dis Colon Rectum* 1999;42:1028-1034.
2. Fazio V, Ziv Y, Church J, et al. Ileal pouch-anal anastomoses complications and function in 1005 patients *Annals of Surgery* 1995;222:120-127.
3. Nyam D, Brilliant P, Dozois R, Kelly K, Pemberton J, Wolff B. Ileal pouch-anal canal anastomosis for familial adenomatous polyposis. *Annals of Surgery* 1997;226:514-521.
4. Van Duijvendijk P, Slors J, Taat C, et al. Quality of life after total colectomy with ileorectal anastomosis or proctocolectomy and ileal pouch-anal anastomosis for familial adenomatous polyposis. *British Journal of Surgery* 2000;87(5):590-596.
5. Miller HH, Bauman LJ, Friedman DR, DeCosse JJ. Psychosocial adjustment of familial polyposis patients and participation in a chemoprevention trial. *Int J Psychiatry Med* 1986;16(3):211-30.
6. Cassileth B, Lusk E, Strouse T, et al. Psychosocial status in chronic illness. A comparative analysis of six diagnostic groups. *New Engl J Med* 1984;311(8):506-511.
7. Vinokur AD, Threatt BA, Vinokur-Kaplan D, Satariano WA. The process of recovery from breast cancer for younger and older patients. Changes during the first year. *Cancer* 1990;65(5):1242-54.
8. Lerman C, Kash K, Stefanek M. Younger women at increased risk for breast cancer: Perceived risk, psychological well-being and surveillance behaviour. *J Nat Cancer Inst* 1994;Monographs No. 16:171-176.
9. Freeman E. Interaction of Pregnancy, Loss and Developmental Issues in Adolescents. *J Contemp Soc Work* 1987;68(1):38-46.

10. Andrews L, Mireskandari S, J J, et al. Impact of familial adenomatous polyposis on young adults: Attitudes to genetic testing, and support and information needs. Accepted by *Genetics in Medicine* on 29/6/2006
11. Ware JE, Sherbourne CD. The MOS 36-item Short-Form Health Survey (SF-36): I. Conceptual Framework and item selection. *Med Care* 1992;30:473-483.
12. Horowitz MJ, Wilner N, Alvarez W. Impact of Event Scale: A measure of subjective stress. *Psychosom Med* 1979;41:209-218.
13. Australian Bureau of Statistics. National Health Survey: SF-36 Population Norms. ABS Catalogue No. 4399.0. Canberra, 1997.
14. Meiser B H, Collins V, Gaff C, St John J. Psychological impact of genetic testing for hereditary non-polyposis colorectal cancer. *Clin Genet* 2004;66:502-511.
15. Espen M, Berk T, Butler K, Gallinger S, Cohen Z, Trinkhaus M. Quality of life in adults diagnosed with familial adenomatous polyposis and desmoid tumour. *Dis Colon Rectum* 2004;47:687-696.