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File ID	155365
Filename	Chapter 4: Quality of life and consequences for social life for familial adenomatous polyposis

SOURCE (OR PART OF THE FOLLOWING SOURCE):

Type	Dissertation
Title	The psychosocial and behavioral impact of familial adenomatous polyposis
Author	K.F.L. Douma
Faculty	Faculty of Social and Behavioural Sciences
Year	2009
Pages	197
ISBN	978#90#75575#24#8

FULL BIBLIOGRAPHIC DETAILS:
http://dare.uva.nl/record/323948

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Quality of life and consequences for social life for familial adenomatous polyposis

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Submitted

ABSTRACT

Purpose

Familial adenomatous polyposis (FAP) is characterized by the development of multiple adenomas in the colorectum that can lead to colorectal cancer. The purpose of this study was to document the impact of FAP on health-related quality of life (HRQL) and on practical aspects of daily life.

Methods

Participants were 525 individuals (response rate 64%) from 145 families at high-risk for FAP. A battery of self-report questionnaires was used to assess generic (SF-36 Health Survey) and condition-specific (EORTC QLQ-C38, Inflammatory Bowel Disease Questionnaire) HRQL, and the consequences of FAP for daily life (e.g. employment).

Results

Self-reported HRQL, assessed by the SF-36, was comparable to that of an age-, gender-, and education-adjusted sample from the general Dutch population. Surgically-treated FAP-patients had significantly lower scores on several HRQL domains (e.g. social functioning, bodily pain) compared to at-risk individuals, non-carriers and not surgically-treated FAP-patients. Type of surgery (colectomy with IRA versus protocolectomy with IPAA) was not associated significantly with HRQL, with the exception of body image. Within the surgically treated group, post-surgical complications and comorbidity significantly affected HRQL. Twenty five to 40% of respondents reported that FAP had limited their recreational activities and affected their working life.

Conclusions

Surgically treated FAP-patients report lower HRQL than those not who have not undergone an operation. However, HRQL was not found to be associated significantly with the type of surgery performed. It is important that patients and caregivers be well informed about the practical and social consequences of FAP.

INTRODUCTION

Familial adenomatous polyposis (FAP) is a hereditary cancer syndrome characterized by the development of multiple (>100) adenomas in the colorectum from the age of 10 years onward. Without treatment, virtually all individuals with FAP will develop colorectal cancer. Therefore, prophylactic surgery is advised, usually between the ages of 15 and 25 years [1;2]. The two main surgical options for FAP are colectomy with ileorectal anastomosis (IRA) or proctocolectomy with ileal pouch-anal anastomosis (IPAA).

There is evidence that each type of familial cancer has a different psychosocial impact [3]. Similar to other hereditary cancer syndromes, FAP is expected to affect health related quality of life (HRQOL) and social aspects of life, such as education, work, leisure time activities and relationships. However, FAP is characterized by an earlier age at onset than, for example, hereditary breast and overian cancer (HBOC) and Lynch syndrome. DNA testing, and invasive procedures such as endoscopic screening and prophylactic surgery are carried out at a younger age.

Previous studies of the HRQL of FAP-patients have focused primarily on the impact of the two main surgical options for FAP. Some of these studies have reported better HRQL after IRA [4;5], while others found no difference [6;7] or better HRQL after IPAA [8;9]. Comparisons of the HRQL of FAP-patients with that of the general population have also yielded mixed results [8;10]. Factors found to be associated significantly with poorer HRQL include employment status and feelings of hopelessness [10]. Two studies reported limitations in social life (family relations, work and sports) due to FAP [11;12]. Clinical observations, anecdotal reports and qualitative studies also indicate that FAP-patients and their families are concerned about the negative effects of the disease on practical aspect of daily life (e.g. work, employment, insurance) [13-18].

The studies conducted to date on the HRQL of FAP-patients have had a number of limitations. Most studies included a heterogeneous patient sample (e.g. including FAP and ulcerative colitis or Crohn's disease) [19-27] or small sample sizes [6;7;10-12;28-30]. Only half of the studies used (semi-)standardized questionnaires [6;8-10;28;29;31]. Those studies investigating the HRQL impact of different types of surgery tended to have larger samples, but only included carriers and did not report on the HRQL of at-risk individuals or of non-carriers. Furthermore, they did not explore other potentially important clinical variables such as the presence of desmoid tumors, time since surgery, DNA diagnosis, or age at diagnosis [5;8;9;31].

The primary objective of the current study was to investigate the HRQL of FAP-family members, and to identify the sociodemographic and clinical factors associated significantly with HRQL. Also investigated were the consequences of FAP for several practical aspects of daily life, including education, employment, and insurance.

MATERIALS AND METHODS

Study sample

Potential participants were identified through the pedigrees of families registered at the Netherlands Foundation for the Detection of Hereditary Tumours (NFDHT). Eligible participants were those: 1) aged 16 years or older, 2) from families registered at the NFDHT; and 3) with either a FAP diagnosis (clinically and/or genetically proven), at risk of inheriting FAP, or proven non-carriership.

Procedures

Invitation letters were sent to contact persons within the families who were typically those who had assisted in drawing the family pedigrees at time of registration at the NFDHT, and were often the key figure within the family with regard to counseling issues. The contact persons were asked to: 1) complete a self-report questionnaire themselves; and 2) assist in inviting other family members to participate in the study by mail. This procedure was adopted to comply with the privacy regulations of the NFDHT. In some families, more than one contact person was recruited because of the large number of family members (or branches within the family).

Questionnaires were mailed between October, 2005 and January, 2007, with a reminder letter sent to all potential participants after 2 weeks. Self-reported clinical data were confirmed by medical record audits whenever possible. All participants gave informed consent. The study was approved by the ethical committees of both the Netherlands Cancer Institute and the NFDHT.

Measures

Sociodemographic variables

Gender, age, level of education, employment and number of children were assessed by self-report.

Clinical characteristics

Data on group (FAP diagnosis, at risk or non-carrier), surgery type, time since surgery, time since DNA diagnosis, personal cancer history, age at diagnosis, desmoid tumours, complications due to surgery, and first-degree relatives with cancer were collected through self-report and, when possible, were confirmed by medical record audits provided by the NFDHT. To correct for the possible confounding effects of comorbidity, respondents were asked whether they had any other chronic health conditions (e.g. diabetes or cardiovascular disease).

Psychosocial factors

QUALITY OF LIFE

Generic HRQL

Generic HRQL was assessed with the Dutch language version of the SF-36 Short Form Health Survey (SF-36) [32]. The SF-36 is composed of 36 questions organized into eight scales: general health perceptions (GH), physical function (PF), role limitations due to physical and emotional health (RP, RE), social function (SF), mental health (MH), bodily pain (BP) and vitality (VT). Scale scores range from 0 to 100. A higher score indicates a better health status. The SF-36 also yields two component summary scores, one for physical (PCS) and one for mental health (MCS).

Condition-specific HRQL

Selective subscales of the Dutch language version of the EORTC-QLQ-C38 [33] were used to assess HRQL issues specific to FAP. The QLQ-C38 was developed for use in colon cancer, and therefore many of the items are also relevant for FAP. We administered the items/scales assessing body image, sexual function, stoma-related problems (where applicable), defecation problems, and weight loss. Scale scores range from 0 to 100, with a higher score indicating better functioning or more symptoms.

We also employed the Social Functioning (SF) subscale of the Dutch language version of the Inflammatory Bowel Disease Questionnaire (IBDQ) [34]. Scores for the IBDQ SF range from 5 to 25. A higher score reflects a better level of functioning.

The EORTC-QLQ-C38 and the IBDQ were administered to FAP-patients only.

CONSEQUENCES FOR SOCIAL LIFE

The questionnaire also included a series of questions relating to problems with education, work, hobbies, sport activities, friendships and relationships, and with health, life or disability insurance.

Data analysis

Descriptive statistics were used to characterize the study sample. The scores on the SF-36 scales were compared with age-, gender-, and education-adjusted normative data available for the general Dutch population [32].

Analysis of variance was used to compare HRQL outcomes between the four risk groups (FAP-patients with and without surgery, at-risk and non-carriers). When significant differences between these risk groups were observed, additional univariate analyses (Student's t-tests, ANOVA, Pearson's r, Kruskal Wallis, Spearman's rho and Mann-Whitney tests) were performed to evaluate which additional sociodemographic and clinical variables were related significantly to the relevant subscales of the HRQL measures.

Multiple regression analysis was carried out to determine which variables were associated significantly with HRQL outcomes at the multivariate level. All variables that showed at least a marginal ($p<.10$) association with HRQL outcomes at the univariate level were entered into the regression analysis. The multilevel method was used because, in some cases, the data from individual respondents were correlated (i.e., nested within families).

Effect sizes were estimated with Cohen's d statistic. An effect size between 0.10 and 0.30 is assumed to be small, between 0.30 and 0.50 to be moderate, and 0.50 or higher to be large [35].

Questionnaires with more than 80% missing items were excluded from all analysis. Half scale mean substitution (for the SF-36 and the IBDQ) or median substitution (EORTC QLQ-C38) were used for missing scale items.

All analyses, except the multilevel analyses, were carried out with SPSS version 15.0. For multilevel analyses, MLwiN version 2.02 was used. A p-value of .05 was considered significant

RESULTS

Sample characteristics

Detailed information on the recruitment of individual family members into the study is presented elsewhere [36]. In total, we received 530 completed questionnaires (64% response rate). Five questionnaires were excluded because more than 80% of the data were missing. Compared with respondents (n=525), non-respondents (n=297) were significantly more likely to be male (58% vs 46%; $p<0.01$) and younger [41.3 (SD=15.8) years vs 43.6 (SD=14.1) years); $p<0.05$].

Table 1. Characteristics of the respondents (*n*=525)

	Mean (range)	SD	N	%
Age	43.6 (16-84)	14.1		
Gender:				
Male			242	46
Female			283	54
Level of education:				
Primary school/basic vocational school			175	33
High school			269	51
College or university			79	15
Missing			2	-
Personal cancer history:				
Yes			45	9
No			480	91
Actual risk:				
FAP-patient[a] who had surgery			296	56
FAP-patient[a] who had no surgery yet			45	9
At risk for FAP[b]			50	10
Non-carrier			134	26
Type of most recent surgery (n=296)				
IRA			136	46
IPAA			112	38
stoma			42	14
Other			6	2
Time since last surgery:				
No surgery, because non-carrier			134	26
No surgery (yet)			95	18
0 - 1 year			16	3
1 - 2 years			16	3
2 - 5 years			33	6
5 - 10 years			73	14
More than 10 years			158	30
Time since receiving DNA test result				
No DNA testing			163	31
0 -1 year			8	2
1 - 2 years			27	5
2 - 5 years			41	8
5 - 10 years			96	18
More than 10 years since DNA test result			14	29
Awaiting DNA test result			9	2
Missing			27	5
Age at diagnosis				
19 years or younger			141	27
Older than 19 years			200	38
Not applicable			184	35

[a] Diagnosis confirmed by positive DNA test result and/or by clinical observation of more than 100 polyps.

[b] Individuals without polyps at risk of inheriting FAP (with inconclusive DNA test result or no DNA testing (yet)).

Table 1 displays the sociodemographic and clinical characteristics of the study sample. Approximately two-thirds of the sample was composed of FAP-patients, of whom 296 had been surgically treated and 45 had not. Five FAP-patients had a genetic diagnosis without having polyps. Fifty-nine percent of the respondents was diagnosed with FAP at age 20 or older, while 41% was younger than 20 years at time of diagnosis.

Generic HRQL

Statistically significant differences were observed between the FAP sample and the general population sample for the SF-36 scales Bodily Pain (p=.000, d=0.18), Role Emotional (p=.006, d=0.13), General Health (p=.001, d=0.14) and Mental Health (p=.001, d=0.14), with the FAP sample reporting better functioning. It should be noted, however, that the effect sizes were consistently small.

Table 2 shows the differences in mean scores on the SF-36 scales between FAP-patients with or without surgery, those at risk of FAP, and non-carriers. Surgically treated FAP-patients scored significantly lower on most of the SF-36 scales, with the exception of Mental Health and the Mental Component Score.

Variables associated with physical functioning for surgically-treated FAP-patients

As FAP-patients who had undergone surgery scored consistently lower than the other groups on the SF-36 scales, and particularly the physical health-related scales, we performed additional subgroup analyses to determine which sociodemographic, clinical variables explained differences in SF-36 scores *within* the surgically treated group. As shown in table 3, the multilevel regression analysis showed that those with higher levels of physical functioning (PCS) were significantly more likely to be male (β=0.272), to have had no complications during surgery (β=0.405), and not to have had any comorbid conditions (β=0.557). This multivariate model explained 20% of the variance in PCS scores ($p<.01$).

Tabel 2. Mean scores and standard deviations for the SF-36, EORTC-QLQ-C38 and IBDQ.

Scales	FAP-patient		At-risk	Non-carrier		
	surgically treated	without surgery				
	n= 296	*n*= 45	*n*=50	*n*=134		
	mean (SD)	mean (SD)	mean (SD)	mean (SD)	Total n	p
SF-36						
General health perceptions	62.1 (25.2)	71.2 (18.3)	76.1 (19.8)	73.9 (21.0)	513	**.000**
Physical functioning	80.9 (22.4)	91.3 (15.2)	90.6 (17.8)	90.2 (18.4)	516	**.000**
Physical role limitations	64.9 (42.5)	91.1 (20.7)	90.3 (26.4)	87.8 (29.6)	515	**.000**
Emotional role limitations	81.2 (36.1)	93.3 (23.1)	94.4 (21.0)	93.1 (20.6)	513	**.000**
Social functioning	81.4 (23.8)	90.3 (17.7)	93.6 (13.5)	88.6 (18.6)	519	**.000**
Mental health	78.7 (16.5)	80.9 (16.4)	81.8 (15.4)	80.5 (15.2)	515	.470
Bodily pain	74.6 (27.8)	87.1 (20.2)	87.7 (18.0)	85.2 (21.0)	517	**.000**
Vitality	63.2 (22.6)	68.6 (19.2)	72.0 (17.4)	68.3 (20.3)	515	**.011**
Physical component scale (PCS)	45.9 (11.6)	52.4 (8.3)	53.7 (7.7)	51.9 (9.4)	506	**.000**
Mental component scale (MCS)	52.1 (9.8)	53.4 (8.6)	54.1 (8.6)	53.3 (8.0)	506	.350
EORTC-QLQ-C38						
Body Image	85.4 (20.5)	91.9 (16.1)	94.0 (13.1)	92.3 (13.1)	525	**.000**
Sexual functioning	42.2 (23.2)	41.9 (22.7)	47.6 (23.7)	45.7 (21.2)	433	.487
Sexual enjoyment	69.6 (24.1)	72.0 (28.4)	71.7 (25.2)	75.0 (22.9)	353	.359
Male sexual problems	11.6 (22.3)	0.0 (0.0)	9.3 (18.3)	4.8 (13.9)	168	.086
Female sexual problems	17.9 (26.9)	9.7 (17.0)	8.9 (12.4)	12.1 (17.6)	185	.609
Defecation problems	19.0 (13.0)	10.6 (12.1)	n/a	n/a	282	**.000**
Stoma-related problems	18.2 (17.1)	n/a	n/a	n/a	51	n/a
Weight loss	10.1 (20.4)	6.7 (15.2)	11.3 (21.9)	8.7 (18.2)	525	.707
IBDQ						
Social functioning	21.8 (4.5)	24.0 (2.3)	n/a	n/a	335	**.000**

SF-36 scale scores could range between 0-100. The PCS and MCS are based on the mean scores and standard deviation of a general population sample which is standardized at a mean of 50 and a SD of 10. The EORTC-QLQ-C38 scores could range between 0-100.The Social Functioning subscale of the IBDQ could range between 0- 25. Significant differences are shown in bold.

Condition-specific quality of life

FAP-patients who had had surgery reported significantly worse body image, poorer social functioning and more defecation problems than the other groups (Table 2). No significant differences were observed between groups in sexual functioning.

Variables associated with condition-specific HRQL for surgically-treated FAP-patients

Standard linear regression analyses were carried out for Body Image (BI) and Defecation problems (DF), because they were not family dependent, while multilevel regression analysis was carried out for disease-related Social functioning (SF). FAP-patients who reported compromised BI were significantly more likely to have had a stoma (β=0.254). The model, which was not found to be significant (p=.117), explained 9% of the variance in body image (Table 3).

Defecation problems among surgically treated FAP-patients was associated significantly with having had surgical complications (β=0.238), and having comorbid conditions, such as cardiovascular diseases and diabetes mellitus (β = 0.151). This multivariate model explained 11% of the variance in defecation problems (p=.000).

Higher disease-related SF among surgically treated FAP-patients was associated significantly with a higher education level (β=0.418), having children (β=0.336), not having had complications (β =.351) and not having comorbid conditions (β = 0.313). This multivariate model explained 18% of the variance in social functioning (p<.01).

Consequences for daily life

As shown in Table 4, 41% of FAP-patients reported that FAP had influenced their employment status. Most commonly reported reasons for working less were: feeling to ill, hospital stays and fatigue. Most commonly reported reasons for working more were: wanting to feel useful, no longer wanting to be seen in a patient role, and financial needs.

Between 14% and 25% of respondents reported that FAP had consequences for other areas of daily life (education, sports, hobbies, relationships).

Table 3. Multivariate regression analyses on HRQOL for surgically-treated FAP-patients

Variables	Physical component scale[a]		Defecation problems[b]		Social Functioning IBDQ[c]		Body Image[d]	
	Standardized β	*p*	Standardized β	*p*	Standardized β	*p*	Standardized β	*p*
Gender	.272	.006						
Complications	.405	<.001	.238	.000	.351	<.001		
Comorbid conditions	.557	<.001	.151	.02	.313	.003		
Education level[e]					.418	.01		
Children					.336	.002		
Type of surgery: IRA vs stoma[f]							.254	.03
Explained variance (%)	20%	<.01	11%	.000	18%	<.01	9%	.117

[a] Variables significant at the univariate level and entered into the model were gender, age, education level, age at diagnosis, cancer history, type of surgery, complications and comorbid conditions.

[b] Variables entered were age, complications and comorbid conditions.

[c] Variables entered were education level, having children, cancer history, complications, comorbid conditions and type of surgery.

[d] Variables entered were complications, desmoid tumours, first degree relatives with cancer, having had DNA-testing, type of surgery.

[e] Only the results for the categories high versus low education are shown as it was the contrast between these two educational levels that was significant at the univariate level.

[f] Only the results for the contrast between IRA and stoma are shown as this was the only contrast that yielded significant results.

Table 4. Consequences for social life in FAP-patients (*n*=341)

Has FAP had consequences for your ….?	N (%)*	Reasons, if yes:	N (%)
Education:			
No	287 (86%)		
Yes	46 (14%)		
		Longer study duration	20 (43%)
		Other study than originally planned	17 (37%)
		Education not yet completed	16 (35%)
Work:			
No	180 (59%)		
Yes	124 (41%)		
		Partly or completely work disabled	73 (59%)
		Worked less, but not disabled	30 (24%)
		Worked more	5 (4%)
		Worked both less and more at different points in time	16 (13%)
Hobby's:			
No	276 (82%)		
Yes	60 (18%)		
		Spends less time	43 (72%)
		Changed type	43 (72%)
		Spends more time	10 (17%)
Sport activities:			
No	251 (75%)		
Yes	84 (25%)		
		Spends less time	68 (81%)
		Changed type	64 (76%)
		Spends more time	4 (5%)
Friendships:			
No	279 (83%)		
Yes	59 (17%)		
		Lost friends	46 (78%)
		Friendships less intense	28 (47%)
		Friendships more intense	20 (34%)
Relationship:			
No	238 (78%)		
Yes	66 (22%)		
		Difficulty starting a relationship	31 (47%)
		Relationship ended	28 (42%)
		More intense	27 (41%)

Table 4. *Continued*

Has FAP had consequences for your?	N (%)*	Reasons, if yes:	N (%)
Insurance**		**Type of insurance, if yes**	
Higher premium because of FAP			
No	251 (75%)		
Yes	82 (25%)		
		Life insurance	52 (65%)
		Disability insurance	9 (11%)
		Health insurance	15 (19%)
		Other[a]	24 (30%)
Insurance or mortgage denied because of FAP or DNA test result?			
No	290 (87%)		
Yes	43 (13%)		
		Life insurance	27 (63%)
		Disability insurance	3 (7%)
		Health insurance	10 (23%)
		Other[a]	10 (23%)

*Shown are only the top 2 or 3 consequences. Numbers do not always add to 341, because some respondents were not yet in the working age group (n=27), not all respondents had a partner (n=29) or because of missing values.

** Missing data for 8 individuals.

[a] For example, funeral insurance.

Approximately one-quarter of the FAP-patients reported having had to pay higher premiums, primarily for life insurance. Thirteen percent indicated that they had been denied a policy, again primarily for life insurance. No significant association was found between insurance problems and age at FAP diagnosis or having undergone DNA testing (data not shown).

DISCUSSION

The results of our study indicate that the HRQL of FAP family members is comparable to that of the general Dutch population. This was also found in a previous study by Hassan *et al*.[8]. However, Esplen *et al.* reported that FAP-patients had poorer HRQL than general population peers[10]. The inclusion of only a selective subgroup of patients (i.e those with desmoid tumours), small numbers, and use of different measures in the Esplen study may explain these discrepant results.

FAP-patients who have had surgery report significantly poorer physical functioning and disease-related social functioning, and more body image and defecation problems than those who have not been surgically treated. Further, among surgically treated patients, surgical complications and comorbidity explained a significant

percentage of the variance in HRQL. Interestingly, type of surgery itself was not associated significantly with most HRQL outcomes, with the exception of body image. Individuals with a stoma reported poorer body image than those who had undergone an IRA. Other studies have yielded mixed results with regard to the effect of type of surgery on HRQL. Some have found that HRQL outcomes are better following IRA[5], others have reported better results following IPAA[8;9], and still others have found no differences[6;7]. Differences in measures used (non-standardized versus standardized measures)[5], methods of data collection (telephone interview versus questionnaire)[5;6], and sampling frame (patients from one clinic only versus a nationwide sample) [8] may explain, at least in part, these discrepant results.

Although, in general, the HRQL of FAP-patients was not significantly compromised, approximately 40% reported that FAP had affected their working life (i.e., partial or complete disability), and 25% indicated that it had limited their recreational activities. Two other studies have reported a similar range of FAP-related restrictions in daily life[11] [12].

Approximately one-quarter of the sample indicated having had some difficulty in obtaining (affordable) insurance due to FAP. However, although some patients reported having to pay higher insurance premiums, this was not found to be related to genetic testing.

Several limitations of the current study should be noted. First, the study was cross-sectional in nature, and thus we can only speak of associations, and not causal relatioship. Second, although normative data were available for the SF-36 Health Survey, there is no such comparative information for the EORTC QLQ-C38 or the IBDQ, thus limiting comparisons with other patient populations.

The current study also had a number of specific strengths. First, we surveyed not only FAP-patients, but also their family members, thus providing a more complete picture of the impact of FAP within the family. Second, our approach to data analysis (multilevel statistics) took explicitly into consideration the fact that more than one individual from any given family could be included in the study sample. Adjusting the data for this type of statistical dependency can be important [37]. Third, the study had a relatively large sample size, which facilitated a number of subgroup analyses. Finally, ours is among the first studies to examine not only the association between surgical type and HRQL, but also the influence of other clinical variables such as DNA status, complications, etc.

In summary, the results of this study indicate that FAP-family members have an HRQL similar to that of the general population. However, FAP-patients who have had

surgery have significantly poorer HRQL than those who have not yet undergone surgery, those at-risk, and non-carriers. Type of surgery, however, was not found to be associated significantly with HRQL outcomes, with the exception of compromised body image in those with a stoma. Rather, complications due to surgery and the presence of comorbid conditions (e.g. cardiovascular disease, diabetes mellitus) appear to drive the HRQL of this subpopulation of patients. Patients need to be informed and counseled about the social and practical consequences of FAP, particularly with regard to work and insurance issues. To our knowledge, FAP-patients are not provided with psychosocial counseling before and after surgery. Although only a minority may need such formal counseling, we would recommend that it be offered to all patients on a routine basis at time of surgery.

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