

Preoperative and Postoperative Quality of Life in Patients with Familial Adenomatous Polyposis

ORIGINAL CONTRIBUTION

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PURPOSE: This study was designed to prospectively examine functional outcome, quality of life, and patients' personal experiences and adjustment to functional changes during the first year after prophylactic surgery.

METHODS: Twenty-one consecutive patients with familial adenomatous polyposis were examined before proctocolectomy (T0), on ileostomy reversal (T1), and 6 (T2) and 12 months (T3) after surgery by means of standardized questionnaires and interviews.

RESULTS: Average physical and mental health declined profoundly after proctocolectomy, followed by a steady improvement after 6 and 12 months. The majority of patients reported the ileostomy period as particularly distressing. After one year, 75 percent of patients reported complete recovery in terms of physical, emotional, and social functioning, whereas one-quarter of patients did not regain their former level of functioning. Despite substantial improvement in pouch functions, functional impairment persists because of frequent bowel movements, resulting mainly in restricted social activities. Ten percent of patients reported impaired sex life, irrespective of gender.

CONCLUSIONS: The majority of patients with familial adenomatous polyposis were found to adjust favorably to functional impairment while maintaining satisfactory quality of life. Complementing standardized quality of life measures by patients' personal experiences may help to identify vulnerable patients in need of psychosocial support.

KEY WORDS: Familial adenomatous polyposis; Quality of life; Prospective study; Preoperative and postoperative comparison; SF-36.

Familial adenomatous polyposis (FAP) is an autosomal dominant syndrome caused by a germline mutation of the APC gene. Affected individuals develop multiple adenomatous polyps all over the colon, which result in the development of colorectal cancer mostly before aged 40 years. Restorative prophylactic proctocolectomy (PPC) has become the surgical treatment of choice to minimize the risk of developing colorectal cancer, to preserve fecal continence, and to provide an acceptable functional outcome. Reconstruction by means of an ileal pouch-anal anastomosis (IPAA) has gained wide acceptance, whereas subtotal colectomy with ileorectal anastomosis (IRA) is still considered as an equivalent alternative by some authors.¹⁻⁷ In the Department of Surgery of Heidelberg University, the surgical procedure of choice is the prophylactic proctocolectomy IPAA, with a temporary protective ileostomy to allow sufficient time for the anastomosis to heal and to prevent complications, such as pelvic sepsis.^{8,9} In recent years, quality of life (QoL) after proctocolectomy increasingly attracted surgeons' attention in addition to the functional result. Health-related QoL is considered an important validation criterion in measuring the outcome of surgery from the patients' perspective.¹⁰⁻¹²

Only a few studies have prospectively analyzed the quality of life of patients with FAP.¹¹ Validated measures to assess QoL were rarely used, and patients' personal experience has attracted little attention.¹⁴ This study was designed to prospectively assess QoL in patients with FAP before and during the first year after proctocolectomy and IPAA. A particular focus of interest relates to the personal experiences during the transient ileostomy period and to the process of adaptation to the functional changes in patients' everyday life. The patients often are young and in a vulnerable phase of life developing from teenagers to adults. In addition to patients' personal experiences elicited by repeated personal interviews, QoL data were collected by using standardized questionnaires. This combined approach is expected to yield valuable information, which may influence clinicians' practice on particular issues that are unique to this period.

Presented at the Consortium for Psychooncology, Cologne, Germany, December 1 to 2, 2005, and at the German Cancer Congress, Berlin, Germany, March 22 to 26, 2006.

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PATIENTS AND METHODS

The patient sample consisted of 21 patients with FAP from 19 different kindreds between aged 16 and 43 years who underwent restorative prophylactic proctocolectomy and IPAA with transient ileostomy from June 2003 to July 2004 in the Department for General Surgery, University of Heidelberg Medical Center. The protocol was approved by the Institutional Review Board (046/2004). All patients who underwent PPC because of FAP during a one-year period were consecutively recruited. Twelve men (mean age, 29.3 years) and nine women (mean age, 22.1 years) participated in the study. Two patients (9.5 percent) refused to participate.

The patients included in the study were at least 15 years old, had clinically proven FAP with no signs of bowel cancer, and no mental condition that precluded them from completing questionnaires and undergoing repeated personal interviews.

Patients were contacted immediately on admission to the surgical ward. After provision of thorough written and personal information about the goals of the study, written informed consent was obtained from all participants.

Data collection took place at four fixed time points: before proctocolectomy (T0), during the hospital stay for ileostomy closure (usually 2 to 3 months after PPC) (T1), three months after ileostomy reversal (T2), and one year after proctocolectomy (T3). All patients underwent laparoscopic proctocolectomy with temporary ileostomy.

Instruments

All instruments were psychometrically tested with established validity and reliability. For global instruments, German population norms were available for comparison; they were complemented by disease-specific measures.

The SF-36 Medical Outcome Survey (MOS Short Form SF-36)¹⁵ is used worldwide and was chosen to assess overall health-related QoL. Patients' data were compared with healthy population norms from the German version of the SF-36.¹⁶ It is composed of four domains that measure physical health (general health perception, physical function, physical role function, physical pain) and four domains that measure mental health (psychic well-being, social function, emotional role function, and vitality). Each of these four domains can be summed up to two scales regarding "physical health" and "mental health": scores range from 0 to 100, with higher scores indicating a better quality of life. The sum score mean is 50/10 (M/SD).

The Oresland-Score was used to quantify overall function after pouch operation¹⁷ (number of daytime/nighttime bowel movements, urgency, seepage, use of protection pads, perianal soreness, medication for stool

regulation, dietary restriction, social restrictions, further stress factors, life changes). Items are summed up (score range, 0–14); higher score indicates poor function.

The Hospital Anxiety and Depression Scale (HADS) was designed to quantify general distress, anxiety, and depression in patients with physical diseases.¹⁸ Subscale scores for depression and anxiety (7 items each) range from 0 to 21 for each subscale.

Optimism and self-efficacy were recorded by a short-form questionnaire originally developed by Scheier and Carver.^{19,20} Higher scores represent high self-efficacy and optimism.

Perceived social support was measured by using the F-SozU-K, a 14-item questionnaire.²¹ The items were added up and a mean was calculated with high scores reflecting higher degrees of perceived support.

Furthermore, five illness-specific questions were added that addressed topics not covered by the former questionnaires. From clinical experience, these questions are considered to be important in the everyday life of patients with FAP (restrictions, disturbance caused by frequent bowel movements, changes in social habits, relationships, sex life).

A semistructured interview was performed face-to-face or by telephone at every time period of data collection to grasp important and sensitive information on patients' personal experiences that cannot be adequately evaluated when relying solely on questionnaires.

Statistical analysis was performed by using the SSPS® software (SPSS Version 11.5, Chicago, IL). Descriptive analysis is presented in the form of percentage or mean/standard deviation (M/SD). With regard to the small sample size, nonparametric tests were used throughout: Spearman's correlation coefficient, Wilcoxon's test to calculate changes in dependent samples over time, and Mann-Whitney *U* test to compare data in pairs. Comparison of the study sample with the population norm was calculated by using the one-sample *t*-test. Statistical significance was set at $P < 0.05$ (two-sided).

RESULTS

Interview Results

The majority of patients were asymptomatic and FAP was diagnosed within the previous year because of a known family history (81 percent). Despite being aware of their risk of being affected, 62 percent were shocked on the definite diagnosis. Fifty-seven percent considered their family experience with FAP as being helpful in coming to terms with their disease, whereas a smaller proportion (19 percent) felt burdened by the familial experience. Nearly all patients felt well supported by families and friends and were optimistic about their ability to cope with the upcoming challenges after surgery.

Before surgery, 91 percent reported good or excellent physical health. However, 38 percent reported considerable fear and depressed mood shortly before the operation, and nearly all patients (90 percent) were afraid of the ileostomy.

Three months after PCC, and before ileostomy closure, the majority of patients expressed relief that the period with the ileostomy had come to an end and expected the return to their normal lives. Two-thirds had to empty the ileostomy bag more than five times per day and more than one-third at least twice during the night. Local inflammation around the ileostomy occurred in 57 percent of the patients, and 81 percent needed medication for stool regulation.

Strong feelings of disgust and shame were reported by the majority of patients. Some patients felt unable to handle their stoma bag themselves. Quite a few patients tried to decrease stool volumina by reducing food (and liquid) intake. One teenage girl who was unable to eat had to have her ileostomy closed prematurely because of a critical weight loss.

Two-thirds of the patients felt definitely limited in their everyday life, especially when visiting public events; 43 percent felt disturbed by these limitations. Decrease in sexual activity was reported by one-quarter of the patients. Sixty-two percent described the ileostomy period not as bad as expected, whereas 20 percent experienced it even worse than anticipated.

Three months after ileostomy closure, 52 percent reported having adapted to the pouch within 6 weeks, and 29 percent within 8 to 12 weeks. However, the high stool frequencies were disturbing for most patients.

Seventy-six percent evaluated their psychic constitution as satisfactory and 24 percent were dissatisfied. Profound changes were experienced by 19 percent of patients at work, by 33 percent in leisure time, and by 10 percent in their social contacts. The search for toilets led to restricted leisure activities, such as sports or social events, resulting in reduced spontaneity. Furthermore, most patients felt limited because of the dietary restrictions. Altogether patients were relieved after ileostomy closure.

One year after proctocolectomy, 86 percent reported physical health as good or very good and only 14 percent were dissatisfied; 76 percent also emphasized their mental health as good and 24 percent mentioned anxiety and fears, *e.g.*, future complications. One patient was diagnosed with a desmoid tumor.

Overall, 52 percent described their life as having changed for the worse after the operation, with only one patient judging his life quality better than before. Forty-three percent had experienced deterioration concerning their working life and 38 percent concerning their leisure activities. Ten percent of both sexes complained about severe problems in their sexual life. Thirty-eight percent admitted that the illness and the operations still played a central role in their life. The

excellent support of family members and friends was emphasized by 95 percent of patients.

Altogether, the ileostomy period was judged to be the most difficult. After ileostomy closure, many patients successfully adjusted to the changes in bowel function, although coming to terms with the resulting impairments in nearly every life domain required strong coping efforts. When asked, 38 percent would advise future patients to think positive, to seek information about the disease and the operation, and to search for contact and exchange with patients who have already undergone surgery.

Functioning

The postoperative course of pouch-related symptoms and pouch function was assessed using the Oresland-Score at three months (t2) and nine months (t3) after ileostomy closure.

After three months the pouch function was generally poor, with average scores at 5.7/3.8 (M/SD). Six months later, function had significantly improved, with average scores at 4.3/3.3 ($P=0.002$). Changes over time for specific items are shown in Figure 1. During the six-month postoperative period, a remarkable improvement of function was observed, with bowel frequencies decreasing both during the day and at night, as well as urgency, seepage, and perianal soreness improving. This resulted in less impairment in social activities. Patients' efforts in handling the changed situation seemed to become more effective with time, as demonstrated by an increasing proportion of patients using dietary restrictions. Despite these remarkable improvements, functional impairment persists, as shown by bowel frequencies exceeding five in every second patient at daytime, and two or more in every third patient at nighttime (Fig. 1).

Course of Health-Related Quality of Life

The course of patients' physical health over time was assessed by means of the SF-36 "physical health" mean summary score. A considerable decline is observed at t1 ($P=0.003$, Wilcoxon's) after PPC and ileostomy compared with t0. Three months after ileostomy closure, (t2) physical health improved significantly ($P=0.013$, Wilcoxon's) compared with t1. This was followed by a further non-significant improvement six months later (t3). One year after proctocolectomy (t3), patients had regained their mean preoperative level of self-reported physical health ($P=0.272$). Although men did not differ significantly from women with regard to their physical condition, women did not achieve their preoperative level of physical health.

Patients with FAP started from a high level of physical health before surgery (t0) comparable to that of the general German population. At the time of ileostomy closure (t1) patients' physical condition was significantly worse than that of the general population ($P=0.015$).

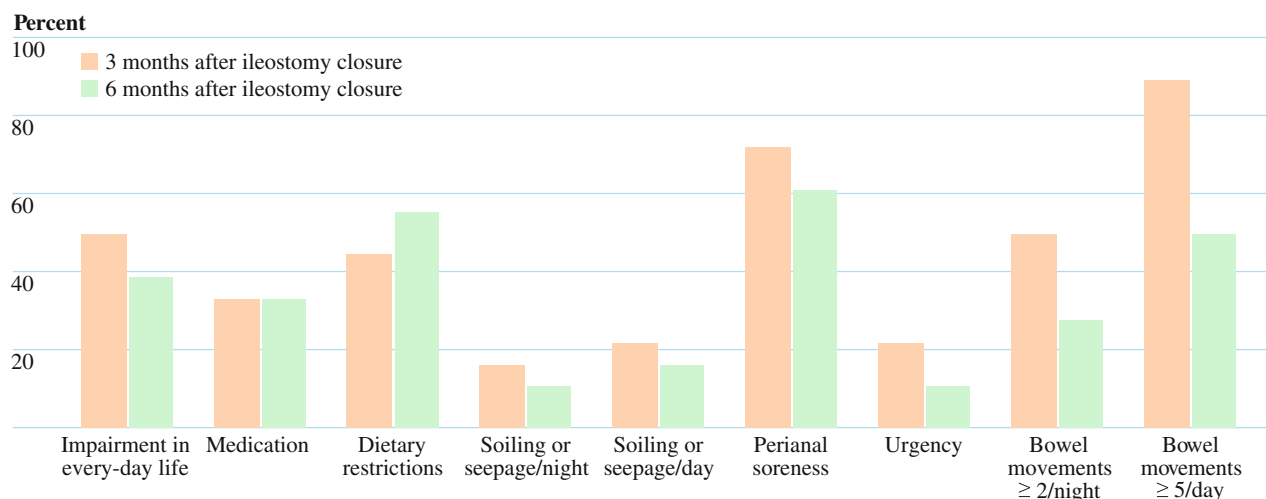


FIGURE 1. Pouch functions three and six months after ileostomy closure (Oresland-Score).

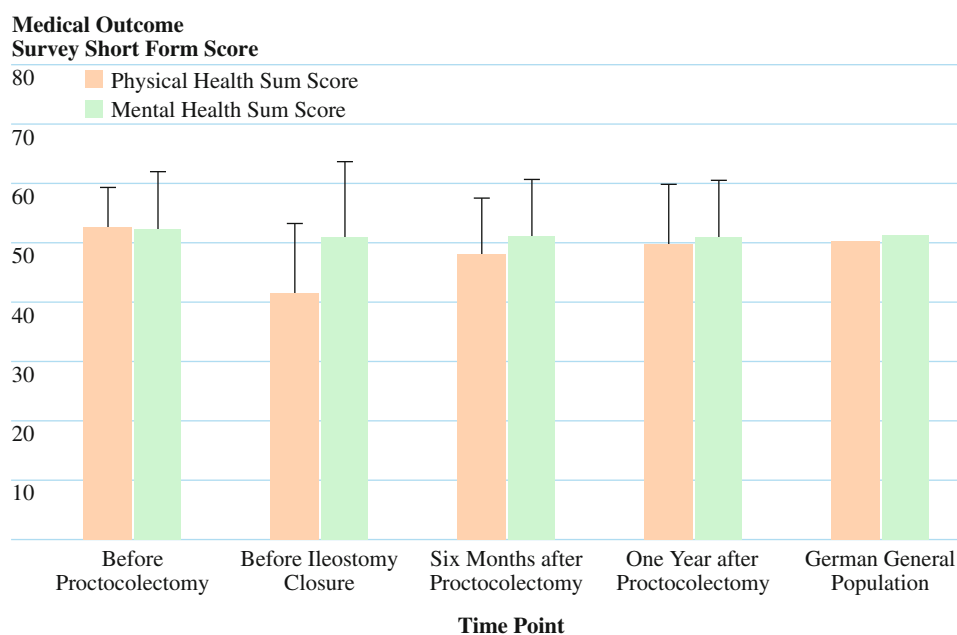
However, three months (t2) later patients' physical health had again attained the preoperative level (Fig. 2).

Patients' mental health remained basically unchanged during the one-year period, aside from a slight, nonsignificant decrease after proctocolectomy. One year postoperatively (t3) patients had reached their preoperative mean level. Women's mental health score was lower at t1 and t3 compared with men's, although the difference did not reach statistical significance. As disclosed in personal interviews, some of the women suffered from psychologic

distress, such as difficult family relationships in one case, and a recurrent desmoid tumor in another case. However, mental health was at each time period comparable to that of the control sample (Fig. 2).

Regarding the subscales of the SF-36 (Fig. 3), physical functioning ($P=0.004$), physical role limitation ($P=0.059$), bodily pain ($P=0.028$), and vitality/energy ($P=0.028$) significantly decreased between t0 (preoperatively) and t1 (3 months postoperatively). Thereafter, the scores improved continuously up to t3, reaching similar levels to

FIGURE 2. Medical Outcome Survey Short Form (SF-36) results during follow-up. Comparison to the German population norm.



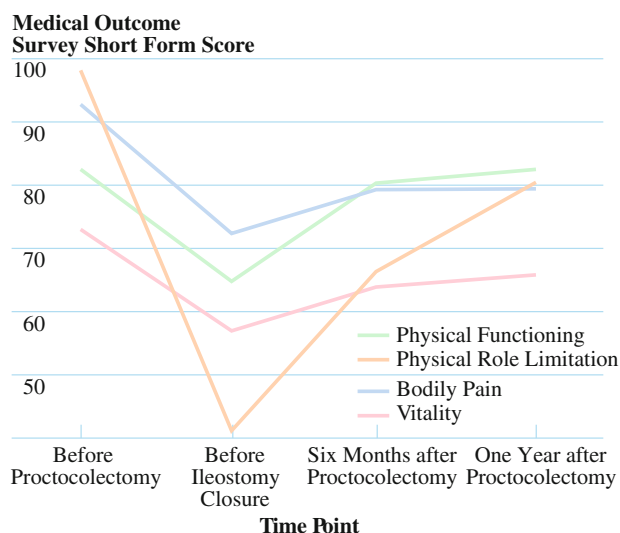


FIGURE 3. Changes in Medical Outcome Survey Short Form (SF-36) subscores.

those before surgery for most subscores. Only the subscales “physical role limitation” ($P=0.041$) and bodily pain ($P=0.037$) did not regain the preoperative level.

Because biased results cannot completely be ruled out caused by the relatively small sample size when relying solely on means, the proportion of patients who had not regained their former functioning level at t3 was calculated. Percentages of patients who did not regain their baseline level are displayed in Figure 4.

One year after proctocolectomy, three-quarters of patients demonstrate full recovery in terms of physical role, physical functioning, and social functioning, whereas one-quarter of the patients apparently need a longer time

period to fully recover. Every second patient suffers more pain, and one of three patients report impaired vitality compared with the time before proctocolectomy.

Personal Attributes and Resources

Anxiety and depression as measured by the HADS remained unchanged over time; at no time period did patients’ mean levels differ from those of the normal population after adjustment for sex and age. Patients’ personal resources, such as optimism and social support, significantly exceeded those of a general population sample.

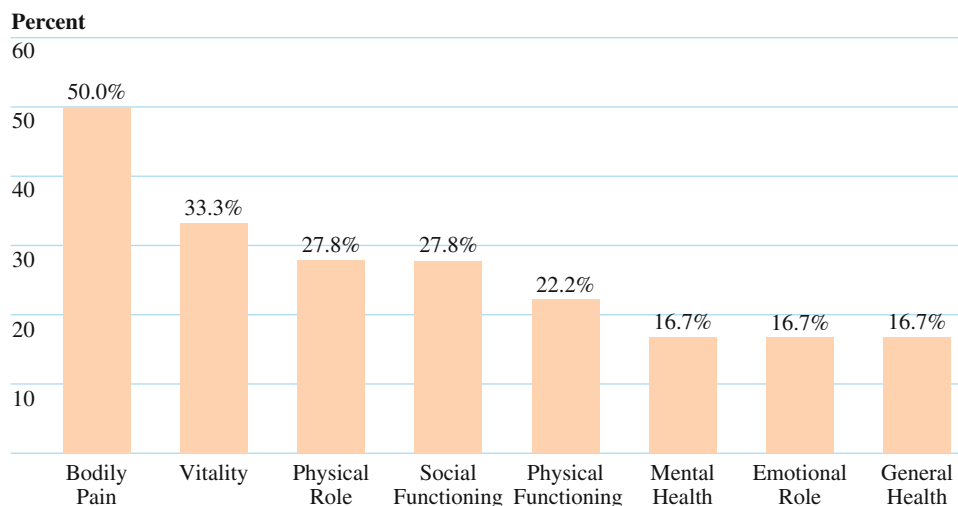
DISCUSSION

This prospective study was designed to examine functional outcome, QoL, and personal experiences of patients with FAP who underwent prophylactic proctocolectomy and IPAA with a protective ileostomy. Apart from one study that reported on a large sample of patients with FAP,¹¹ there is limited knowledge about the course of QoL in patients with FAP subsequent to preventive surgery.¹⁴

Starting from perfect general health before surgery, PPC led to a profound though transient decline in patients’ overall QoL, as assessed by the SF-36, predominantly in terms of physical condition. After ileostomy closure, a steady recovery at fast pace was observed during the subsequent months. Overall, patients with FAP had regained their former physical and mental health status one year after surgery. These findings are in accordance with those reported by Fazio *et al.*¹¹ who found patients’ QoL to improve steadily within the two years after PPC.

Even in the presence of “normal” SF-36 sum scores suggesting perfect health, it became apparent from interviews that half the patients had experienced adverse

FIGURE 4. Proportion of patients not recovering within the first year after prophylactic proctocolectomy/ileal pouch-anal anastomosis.



effects and substantial deterioration in their lives after PCC and IPAA with regard to work, social activities, and sexual life. The ileostomy period was perceived as particularly difficult by the majority of patients. Patients reported on strong feelings of disgust and shame because of the loss of a sense of control and physical integrity, resulting in a severely disturbed body. Similar perceptions were described in a recent qualitative study.²² Possibly, the ileostomy experience negatively interferes with a fragile body image in this stage, only gradually consolidating during adolescence.²³ This may explain maladaptive responses, such as anorectic reactions in teenage patients, leading to dehydration or critical weight loss. There is evidence from interview findings that passive and avoidant coping prevails during the transient ileostomy period, as opposed to active and confronting coping strategies when there is no escape from the enduring changes after ileostomy closure. As suggested by some authors, the decision against a protective ileostomy should be based on clinical selection criteria,^{4,24} which could favorably be complemented by appraisal of the teenage patient's developmental and psychosocial condition.

After continence was restored, patients complained in particular about high stool frequencies and the loss of spontaneity because of depending on sanitary facilities. Functional outcome, evaluated by the Oresland-Score, improved significantly within six months after ileostomy reversal. The proportion of patients reporting five or more bowel movements during the daytime decreased from 90 percent after three months to 50 percent six months later. Similarly, nighttime bowel movements decreased and impaired continence was less frequently reported. These findings are in line with those reported by Oresland one year after operation.¹⁷ Above the overall improvements, the time required to adapt to the pouch varied considerably among patients: 50 percent achieved this within six weeks, whereas one-third needed two to three months. This information on interindividual variability may be useful for future patients and encourage them to have patience until bowel function has stabilized.

The profound change in bowel habits after PPC/IPAA exerts a major challenge to patients' coping strategies, requiring considerable resources and ability to adjust successfully. The majority of patients show favorable personal and social resources as indicated by the absence of psychologic morbidity preoperatively and postoperatively, high levels of optimism and self-efficacy, and excellent social support. With the aid of these resources, most patients successfully adapt to the challenges and impairments while regaining their former health state. Support from their families is considered crucial by patients, with the majority emphasizing family support as most helpful. It is generally assumed that FAP families develop a strong sense of commitment and coherence through previous experiences with the inherited disorder, thus facilitating patients' adjustment.¹³ Data from the interviews in this study partly support this assumption,

with 50 percent of the patients considering experiences in the family as facilitating the adjustment to their own disease. On the other hand, a number of patients considered their family history as an additional burden (e.g., through cancer-related death of a parent).

CONCLUSIONS

After proctocolectomy because of FAP, patients experience the time with an ileostomy as a period with poor life quality. After this period, most patients successfully adapt to the new situation within a short time, and one year after proctocolectomy they evaluate their quality of life similar to the general population, even with considerable functional impairment. Complementing standardized QoL measures by patients' personal experiences may favorably add to further improve functional outcome and QoL of patients with FAP.

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