Self-Surveillance for Genetic Predisposition to Cancer: Behaviors and Emotions

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**Purpose/Objectives:** To describe the kinds of self-monitoring activities and the emotional responses associated with those activities in patients with a genetic predisposition to multiple endocrine neoplasia type 2a (MEN2a) or familial adenomatous polyposis (FAP).

**Research Approach:** Thematic analysis of the transcripts of patient interviews conducted for two previous grounded theory investigations of participation in lifelong surveillance for patients with cancer predisposition syndromes and their family members.

**Setting:** In the original studies, participants were recruited through a high-risk gastrointestinal cancer clinic (for FAP) and pediatric and adult endocrinology clinics (for MEN2a) at two eastern U.S. medical centers and by patient referral.

**Participants:** 58 transcripts of interviews with 29 patients; 17 diagnosed with FAP or the variants of Gardner syndrome and attenuated FAP and 12 patients diagnosed with MEN2a.

**Methodologic Approach:** Informants participated in two hour-long, in-depth interviews and completed a self-administered sociodemographic questionnaire.

**Main Research Variables:** Types of self-surveillance activities.

**Findings:** Patients engage in an elaborate set of self-surveillance activities that are grouped into five categories of behavior: Medication Appraisal, Phenotype Tracking, Intake and Output Monitoring, Laboratory and Treatment Recording, and Tracking of Visits. Self-surveillance behaviors are grouped independent of type of syndrome, penetrance, age, or gender of the patient. Each category comprises a variety of behaviors that correspond with treatment recommendations and understanding of the disorder.

**Conclusions:** Self-surveillance may be driven by a combination of anticipation and the need for control and understanding.

**Interpretation:** Findings from the study could be used to create an assessment tool to evaluate the extent to which patients are involved in day-to-day self-monitoring. Clinicians may use the categories to better understand patients’ knowledge deficits and the emotional impact of enhanced vigilance. Self-surveillance activities performed by patients with MEN2a and FAP also may be performed by patients with other cancer predisposition syndromes.

The importance of surveillance to controlling cancer is explicit. Guidelines for lifelong cancer risk management and prevention invariably comprise periodic medical evaluation and patient instruction about the value of adherence to follow-up guidelines. Another element of lifelong management is patient self-monitoring or self-surveillance. Although self-surveillance occurs out of the purview of clinicians, it plays a significant role in lifelong health promotion and disease prevention. Patients who engage in self-surveillance interpret what they observe and decide to either self-treat, seek help, or avoid professional intervention. The observations and interpretations patients make of their physical signs and feelings are the sources of essential data that often are sought and used by clinicians to make medical judgments.

How patients think about the implications of their genetic predisposition and the meaning of lifelong surveillance for cancer affects their ability to accept and practice behaviors that promote health. Surveillance guidelines, however, do not elaborate on how, or the extent to which, patients should engage in self-surveillance activities. Although nurses, justifiably, presume that self-surveillance may contribute to good health care, significant gaps remain in the knowledge of patients’ experiences. The purpose of this article is to describe the kinds of self-monitoring activities performed by patients with one of two types of cancer predisposition syndromes and the emotions associated with engaging in the self-surveillance activities. The
article will include a brief overview of two cancer predisposition syndromes and offer recommendations on how the information can be tested further and translated to practice.

Background

Lifelong medical management of a person’s genetic predisposition to cancer is guided by recommendations for periodic physical examinations and laboratory testing for phenotypic expression of a genetic mutation. Syndromes, by definition, are complex disorders that may affect multiple organ systems. The expression of a cancer predisposition syndrome is unpredictable. Although cancer is inevitable, it occurs at different ages and in different ways for different family members. When people are diagnosed with a genetic mutation that predisposes them to cancer, they are certain of one thing: The watching and waiting for the expression of the genetic disorder will begin. Two types of cancer syndromes illustrate the complexity of lifelong surveillance: multiple endocrine neoplasia type 2a (MEN2a) and familial adenomatous polyposis (FAP).

Prototypical Cancer Predisposition Syndromes

FAP and MEN2a have autosomal dominant inherited patterns; thus, all carriers of the altered gene will develop disease, and each carrier has a 50% chance of passing the genetic mutation to offspring. Both are uncommon cancer syndromes, but each may be considered a prototype for practice in medical genetics because specific causative mutations are known, reliable molecular genetic tests can identify individuals at risk, and treatments are available, such as prophylactic surgery, to prevent cancer occurrence (Gimm & Dralle, 1999).

Early identification of individuals with mutations to the adenomatous polyposis coli (APC) gene (with FAP) or the rearranged transfection (RET) gene (with MEN2a) leads to early treatment and enhanced health monitoring, which has been associated with decreased morbidity and mortality (Cruz-Coerrea & Giardiello, 2002; Giardiello et al., 1997). MEN2a and FAP may be diagnosed prenatally or early in life by molecular genetic testing. Once a diagnosis is made, cancer prevention strategies such as prophylactic surgery are effective, but patients must participate in lifelong surveillance for precursor lesions, second primary neoplasias, and complications from treatment. The experiences of patients with MEN2a and FAP with regard to lifelong self-surveillance may be relevant for patients with other single-gene disorders that have multiple physical effects (cancer and noncancer) that are expressed variably and require complex care over a lifetime, such as Von Hippel Lindau syndrome, breast and ovarian cancer syndrome, and Marfan syndrome.

Characteristics and management of multiple endocrine neoplasia type 2a: MEN2a is a cancer syndrome that is caused by a mutation to the RET proto-oncogene on chromosome 10 (Mulligan et al., 1993; Pausova et al., 1996). Genetic testing is available to members of high-risk families for detection of carrier status (Howe et al., 1992; Learoyd et al., 1997; Ledger, Khosla, Lindor, Thibodeau, & Gharib, 1995; Mulligan et al.). The syndrome leads to abnormal cell growth and malignant and benign tumors in endocrine glands. Affected individuals may develop medullary thyroid carcinoma (MTC) and pheochromocytoma (adrenal tumor) (Modigliani et al., 1995).

MTC is the main cause of morbidity and mortality and develops in nearly all mutation carriers (Chi & Moley, 1998; Frank-Raue, Hoppner, Buhr, Herfarth, & Raue, 1997; Lairmore, Frisella, & Wells, 1996). Even though MTC is slow growing and indolent, it can metastasize to local and distant sites before it is clinically detectable (Gimm & Dralle, 1999). Approximately 25,000 cases of thyroid cancer were expected to occur in 2005 (Jemal et al., 2005). MTC represents about 10% of those cancers, and 25% of that 10% are inherited (Randolph & Maniar, 2000). A precursor lesion of MTC may be present at birth in some carriers, but the mean age at diagnosis of MTC in carriers is 20 years, much earlier than the mean age at diagnosis of MTC in patients who do not carry the RET mutation, which is 35 years (Halling et al., 1997). A carrier of MEN2a may undergo prophylactic or curative thyroidectomy as early as five years old (Lairmore et al.).

Lifelong surveillance is advised to detect disease and to monitor for consequences associated with medical management after thyroidectomy, parathyroidectomy, or adrenalectomy (Suzbin, Kacker, Kakani, Komisar, & Blaugrund, 1996). Biochemical screening for an elevated serum calcitonin level is used to detect recurrent cancer and new tumors as well as to monitor the effectiveness of postoperative thyroid hormone-replacement therapy (Fitze, 2004; Skinner, DeBenedetti, Moley, Norton, & Wells, 1996). Despite efficacy of hormone-replacement therapy, clinicians face some problems determining optimum dosage and combating nonadherence (Frank-Raue et al., 1997). In summary, surveillance for MEN2a is complex, involves monitoring multiple organ systems over a lifetime, and is an important means of cancer control (Easton et al., 1989; Telander, Zimmerman, van Heerden, & Sizemore, 1986; Wells et al., 1982, 1994).

Characteristics and management of familial adenomatous polyposis: FAP arises from germline mutations of the APC gene on chromosome 5 (5q21) (Giardiello et al., 1997; Kinzler et al., 1991). FAP includes familial polyposis, hereditary polyposis of the colon, and Gardner syndrome. All patients with FAP develop multiple adenomatous polyps in the colon and rectum that inevitably develop into adenocarcinomas (Phillips, Spigelman, & Thompson, 1994). Some families that are affected with a mild form of FAP, called attenuated FAP, develop fewer polyps at later ages.

FAP occurs in approximately 1 of every 8,300–14,025 live births, affecting both genders equally (Bussey, 1975). Although most cases are inherited, approximately 20%–40% arise from de novo mutations without a family history of disease (Lal & Gallinger, 2000). FAP represents about 5% of all colorectal cancers (Cruz-Coerrea & Giardiello, 2002). Therefore, about 7,300 cases were expected in 2005 (Jemal et al., 2005). The syndrome leads to the development of hundreds to thousands of polyps in the colon and rectum, usually beginning in adolescence (Phillips et al., 1994; Rhodes & Bradburn, 1992).

The diagnosis of FAP is made by discovering more than 100 adenomatous polyps or through genetic testing in a known FAP family. Once a person is diagnosed, an analysis of the family should be done to determine the likelihood that other relatives have the condition. Genetic testing can identify the specific abnormality in the affected individual and then allow healthcare professionals to look for that defect in other family members. Children of patients with FAP should undergo genetic testing or endoscopic screening by age 10. If genetic tests are positive in those children, endoscopic screening should continue every one to two years. If genetic testing is negative, they can be spared the intensive screening (Coyle, 2002; Jo & Chung, 2005).
Approximately 15% of carriers develop polyps by 10 years of age, and 90% develop polyps by age 30. If genetic testing is not performed, the diagnosis of FAP among individuals with family history is confirmed by the presence of 100 or more adenomatous polyps on colonoscopic examination (King, Dozois, Lindor, & Ahlquist, 2000; Petersen, 1996). Benign adenomas progress to colorectal cancer by the fifth decade or earlier (Hyer & Fell, 2001). Colectomy usually is advised at the time of diagnosis (Cruz-Correa & Giardiello, 2002) and performed during adolescence or early adulthood.

FAP also is associated with a number of benign and malignant extra-colonic lesions. People with FAP have an increased lifetime risk of duodenal (5%–11%), pancreatic (2%), thyroid (2%), brain (less than 1%), and liver cancers. Benign lesions include congenital hypertrophy of the retinal pigment epithelium that may develop into tumors of the fundus (Shields, Shields, & Singh, 2000). Carriers may develop osteomas, jaw lesions, supernumerary teeth, epidermoid cysts, desmoid tumors, gastric adenomas, mesenteric fibromatosis, and other lesions (Giardiello et al., 1993; Gurbuz et al., 1994; Offerhaus et al., 1992; Sondergaard et al., 1987). Those lesions require lifelong monitoring and medical management (Matloff, Brierley, & Chimera, 2004).

As with MEN2a, members of families with FAP participate in enhanced surveillance as soon as their carrier status is known or if clinical evidence exists with or without a family history. Because the mutation may be detected in only 60%–80% of cases and de novo mutations may exist, people in high-risk families without positive genetic test results are advised to participate in enhanced colorectal cancer screening (Hyer & Fell, 2001; Nandakumar, Morgan, Silverberg, & Steinhagen, 2004). Flexible sigmoidoscopy is started annually at 10–12 years of age, every two years at age 25, and every three years at age 35 (Cruz-Correa & Giardiello, 2002; National Comprehensive Cancer Network, 2005). Even patients with negative results on mutation testing should undergo sigmoidoscopy at 15, 25, and 35 years of age with standard screening thereafter, because errors in molecular testing have been reported (Giardiello et al., 1997). Once polyposis has been established, complete colonoscopy is performed every 6–12 months and upper endoscopy at least every three to four years. In addition, patients may be treated with nonsteroidal anti-inflammatory drugs to attenuate adenoma formation. Patients on chemoprevention also must be monitored for signs of adverse reactions such as abdominal pain, diarrhea, allergic reaction, and dyspepsia (Ishikawa, 2004; Steinbach et al., 2000).

Psychosocial Impact of Lifelong Surveillance

The literature on cancer self-surveillance is written predominantly from physiologic and technical perspectives. Monitoring one’s own disease early in life may generate negative emotions, including denial, anger, and anxiety, and may alter a young person’s image of self or relationships in the family (Fryer, 2000; Harper & Clarke, 1990; Wertz, Fanos, & Reilly, 1994). Investigations of the psychosocial factors associated with participation in surveillance activities, such as adherence to monitoring guidelines, perceived quality of life, and psychological threat of cancer risk, confirm the complex psychosocial nature of self-surveillance.

Adherence to follow-up guidelines: A substantial body of literature dating back to the 1970s exists on compliance and predictors of screening behavior that provides insight into the potential emotional consequence of self-surveillance. Findings point to patients’ concerns about the effectiveness of treatment, the discomfort of some screening tests, and the fear of negative findings as the principal mediators of compliance with screening guidelines and that compliance decreases over time, especially if extra effort is involved (Beeker, Kraft, Southwell, & Jorgensen, 2000; Blalock, DeVellis, & Sandler, 1987; Matthews & Hingson, 1977; Neilson & Whynes, 1995). However, some aspects of self-surveillance that are not yet known may be instrumental to long-term compliance.

Cleiren, Oskam, and Lips (1989) surveyed patients with MEN2a and their relatives about follow-up services and superficially looked at issues related to surveillance. The authors described how families were dissatisfied with annual screening. They recounted patient complaints of insufficient information, inadequate handling of children, and delays in receiving medical reports. Respondents reported difficulties talking about fears and uncertainties. The researchers concluded that the patients had different expectations of their screening needs than clinicians and advised that patients and family caregivers should be equally informed and involved in ongoing care. They suggested that follow-up care is complex and that communication between patients and professionals must be evaluated and revised for optimum therapeutic value. In the report, a description of what patients were doing at home with regard to watching and waiting was conspicuously lacking. Observations from self-monitoring of physical feelings may not match clinicians’ reports of abnormal laboratory work and the need to retest. For example, a patient with MEN2a who has had a thyroidectomy might feel healthy even after missing a dose or more of calcium supplements or thyroid hormone replacement. Patient dissatisfaction may stem from a missed opportunity to satisfactorily compare findings from self-surveillance with findings from formal medical surveillance.

Perceived quality of life: Day-to-day living with disease is the principle correlate of quality of life for patients. In a meta-analysis of more than 20 years of research on the relationship between type of colorectal surgery (stoma versus nonstoma) and quality of life, Sprangers, Taal, Aaronson, and te Velde (1995) reported that groups experienced long-term effects of treatment in four categories of functioning: physical, psychological, social, and sexual. All categories of effects require lifelong self-monitoring that may affect quality of life. However, the authors did not specifically address self-monitoring activities with regard to long-term effects and did not propose a theoretical link among categories.

Information-seeking behavior is fundamental to day-to-day living and is related to self-surveillance in that it helps patients to interpret their observations (e.g., why they are feeling a particular emotion or sensation) and what actions, if any, they must take. Sabay, Gray, and Fitch (2000) described patients’ (N = 20) experiences attending a gastrointestinal follow-up clinic for colorectal cancer. The investigators reported that, overall, patients were satisfied with their treatment, yet some were dissatisfied with information on long-term management. Although patients relied on cancer specialists for specific cancer-related information, they relied on primary care providers to fill gaps in information about other health issues and day-to-day care. As in other studies, the authors did not qualify self-surveillance as part of long-term management, and patients’ participation in lifelong self-monitoring was not explored. The measuring of patients’ dissatisfaction
with information on long-term management might have inadvertently measured dissatisfaction with opportunities to discuss self-monitoring.

A few studies have described day-to-day experiences following diagnosis of a cancer predisposition syndrome in terms of quality of life. Telenius-Berg, Ponder, Berg, Ponder, and Werner (1989) reported the results of a descriptive study of the quality of life of patients with MEN2a after adrenalectomy. Respondents described problems with long-lasting fatigue, intolerance to infection and stress, problems complying with medication schedules, worry, and fear. The most bothersome problem was the management of thyroid or adrenal hormone-replacement therapy, which requires periodic self-surveillance for signs of complications or feelings that herald a need for dosage adjustments. Those problems are experienced daily and are self-reported; they require patients to be sensitive to gross and subtle changes in physical and emotional feelings. Patients’ attention to those problems requires a degree of cognitive sophistication sufficient to discern differences over time. Telenius-Berg et al. provided an important view of the overall experiences of patients who had the less common of the two surgical treatments for tumors associated with MEN2a. The study pointed to the self-monitoring of medications as a major category of self-surveillance. Since then, healthcare providers have called for additional research (Giarelli, 2002).

Psychological threat of cancer: Living with cancer or living with the threat of cancer may cause psychological distress. People with cancer predisposition syndromes may be thinking, every day, about a threat of cancer, loss, or disability. A study by Bliss and Johnson (1995) described patients’ perceptions of living with a diagnosis of cancer. Thirty percent (n = 65) had colorectal cancer. Three dimensions of need were identified: quality of disease management, sympathetic communication, and regaining control of life. All of the dimensions are related to lifelong health monitoring and may, specifically, have a self-surveillance component. For patients with a cancer predisposition syndrome, the impact of a cancer diagnosis may be the evolving threat of disease (uncertainty when expression of an altered gene is variable) rather than a discrete threat (diagnosis). No studies to date elaborate on when expression of an altered gene is variable) rather than a discrete threat (diagnosis). No studies to date elaborate on the link between the evolving threat of cancer and emotions associated with it.

In summary, formal lifelong monitoring guidelines for MEN2a and FAP are complex and well defined but do not include recommendations for patient day-to-day self-monitoring. Yet clinicians regularly gather and make decisions based on important data from patients’ self-report of symptoms (physical feelings) and emotions. Some aspects of lifelong monitoring and adjustment to physical and lifestyle changes associated with medical and surgical treatments generate a range of emotions that include a desire to have some measure of control over chronic illness. Research describing the behaviors and emotions exhibited by patients involved in self-surveillance as a component of lifelong health monitoring for genetic predisposition to cancer is lacking.

**Conceptual Framework**

The conceptual model constructed by Giarelli (2003) in her study of families with MEN2a is the source of the guiding concept of incidental surveillance (see Figure 1). Giarelli’s model of participation in lifelong surveillance for genetic predisposition to cancer proposes that patients engage in two kinds of surveillance activities: planned and incidental. Incidental surveillance is defined as day-to-day self-monitoring for signs of disease and response to treatment (see Table 1). Each surveillance event can remind a person of a threat to health, such as cancer occurrence or recurrence, disability, or loss. Once reminded, the person attends to the meaning of the event and interprets how the information collected during self-surveillance is associated with the threat. The information is self-managed as needed to promote health or prevent illness. After each surveillance event, the patient must integrate the relevant event and outcome information to restore a sense of wholeness and well-being (Giarelli, 2003). Incidental surveillance is self-surveillance. Its purpose is to collect information about personal health in relation to a predisposition to cancer and the impact of therapeutic interventions. Giarelli’s (2003) theoretical construction and definition of incidental surveillance guided the coding process and thematic analysis of transcripts for the current study.

**Methods**

This thematic analysis of interview transcripts searched for descriptions of self-surveillance behaviors and for evidence of patient reports of emotions associated with
surveillance behaviors. Transcripts of interviews were data collected as part of two previous but related grounded theory investigations to describe the phenomenon of “participation in lifelong surveillance.” Self-surveillance is a component of the phenomenon of lifelong surveillance. Although patients and family members were interviewed, only patient transcripts were used as data for the study.

In the original studies, the patient participants were selected purposively, using the following criteria: (a) diagnosed with MEN2a or FAP by molecular genetic testing or clinical presentation and family history, (b) aged 14 years or older, (c) able to speak English, (d) participating in follow-up care, and (d) able to communicate abstract ideas. Approximately 25% of patients were recruited from a large, eastern U.S. research university and a regional children’s hospital. Seventy-five percent of participants were recruited by referral from patients and family members. Written informed consent and assent were secured from adults and minors, respectively, to engage in interviews that were tape recorded for analysis. Before thematic analysis, identifying information was removed from all transcripts, and participants were offered the opportunity to review and comment on the transcripts before analysis. Both studies were approved by the scientific review committee of the regional cancer center and by the institutional review boards of each recruitment site.

### Procedure

The original interviews were conducted by the author, who was the principal investigator for the original grounded theory studies. The first open-ended interview focused on eliciting patients’ descriptions of experiences participating in lifelong surveillance for a genetic predisposition to cancer. Each interview used identical open-ended guiding questions. The second interview was conducted for theoretical sampling (i.e., clarifying and expanding concepts and ideas raised in the first interview). Interviews were conducted by telephone or in person. Some researchers have suggested that telephone interviews may elicit truncated answers (Fry, 1983; Groves & Magilavy, 1981; Rogers, 1976). However, the quality of the telephone interviews was comparable to those that were conducted face-to-face by virtue of their equivalent length and richness of description.

### Data Analysis

The data consisted of 58 transcripts collected from hour-long, in-depth interviews conducted with 29 patients. Transcripts ranged in length from 10–18 single-spaced pages.

The author conducted two types of analyses. Sociodemographic data about the patients who were interviewed during the original studies were summarized descriptively to illustrate characteristics of the participants. NVivo qualitative data management software (QSR International, Melbourne, Australia) was used to store, retrieve, index, and search data and to facilitate data management and thematic analysis (Richards, 1999; Richards & Richards, 1998).

Transcripts of interviews were studied using thematic analysis, a technique that searches text for units of meaning, sometimes called critical incidents or codable moments, that match a conceptual construct (Boyatzis, 1998). As opposed to grounded theory, thematic analysis develops themes using theory-driven or prior-research–driven methods and then applies a code to a critical incident. Similar to other kinds of qualitative data analysis, such as grounded theory (Strauss & Corbin, 1998) or narrative analysis (Manning & Cullan-Swan, 1998), in thematic analysis units of text must be microanalyzed (coded) to search for and identify behaviors and emotions in the recorded descriptions.

The coding of interview data was performed at two levels. First-level (line-by-line) analysis was used to search the text for reports of behaviors and associated emotions. The coding scheme comprised two steps. The first step was an initial search for descriptions of thoughts a patient expressed about his or her disorder followed by actions taken on that thought. The behaviors were labeled as incidental surveillance (self-surveillance), according to the conceptual construct defined by Giarelli (2003). Incidental surveillance activities were distinguished from planned surveillance (e.g., follow-up visits) by definition. Once an incidental surveillance event was coded, the second step consisted of examining the surrounding text for descriptions of feelings associated with the behavior (self-surveillance activity). The second level of coding was used to group the self-surveillance behaviors into categories (themes) of behavior along with the associated feelings.

The following quotation illustrates how, during thematic analysis, critical incidents are identified and coded as a behavior and an associated emotion. In the following example, a 48-year-old female patient with MEN2a stated,

If I haven’t been to the doctor for a while, I feel kind of on my own and I become much more conscious [feeling before: mild worry] of the way I take care of myself ... you know ... sort of making sure I have enough [levothyroxine sodium tablets] [self-surveillance: monitoring medication] or making sure I take my calcium [self-surveillance: monitoring medication]. . . . I guess I become more nervous [feeling before: anxiety] about my health and start to pay attention. . . . I feel kinda guilty, especially since I insist my kids do healthy things [feeling before: guilty]. . . . It helps [feeling after: relieved].

All transcripts were coded in the same manner, and emotions were grouped as either before or after self-surveillance.

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### Table 1. Definitions of Surveillance Events

<table>
<thead>
<tr>
<th>Type of Surveillance</th>
<th>Definition</th>
<th>Examples</th>
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<tbody>
<tr>
<td>Planned</td>
<td>Events are scheduled in advance, involving the participation of a professional healthcare provider as an observer. They occur according to recommended guidelines for follow-up care and are proposed for specific intervals. The events may require hours or days to complete.</td>
<td>Serum and urine biochemical analysis, Diagnostic tests, such as colonoscopy, sigmoidoscopy, and magnetic resonance imaging, Physical examination, Consultation</td>
</tr>
<tr>
<td>Incidental</td>
<td>Events do not involve a professional healthcare provider as the observer. They are performed by the patient or by a family member for an affected person. They occur at irregular or regular intervals and may occur many times per day, daily, or less frequently.</td>
<td>Attention to compliance with medication schedule, Assessment of side effects or adverse effects of pharmaco-therapeutics, Assessment of physical symptoms</td>
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ONCOLOGY NURSING FORUM – VOL 33, NO 2, 2006
Results

Participant Characteristics

The 58 transcripts were collected from 29 patients who participated in the original studies. Seventeen patients were diagnosed with FAP or the variants of Gardner syndrome and attenuated FAP and ranged in age from 15–80 years (X age = 40 years). Twelve patients were diagnosed with MEN2a and ranged in age from 19–74 years (X age = 41 years). Participants comprised 18 kindreds from eight states. Participants in the FAP group were less likely to be employed full-time and to report excellent health. Other sociodemographic characteristics were comparable across cancer syndromes (see Table 2).

Participation in Planned Surveillance

Twenty-eight patients (97%) reported regular visits during the previous five years and attending at least one follow-up visit with a primary care provider or specialist (endocrinologist or gastroenterologist) in the past year. This reflects appropriate adherence to recommended screening and monitoring guidelines for MEN2a and FAP. One patient with FAP reported not attending a medical follow-up evaluation in more than five years. He did not have health insurance and was intermittently unemployed. His participation in self-surveillance activities was equivalent in frequency and category to the others. Twenty-seven participants reported consistently taking one or more medications as prescribed by their physicians. Two patients with FAP reported not taking chemopreventive medication (celecoxib) because they could not afford the drug. Those two patients reported participating in clinical trials and receiving medical advice to continue the medication. Once the trials concluded, the drugs were no longer free.

Self-Surveillance Behaviors

From patients’ descriptions of day-to-day living with their genetic disorders, all patients apparently developed an elaborate set of self-surveillance behaviors. Self-surveillance behaviors are grouped into five thematic categories that seem to occur independent of the type of syndrome, disease penetrance, and gender of the patient: Medication Appraisal, Phenotype Tracking, Intake and Output Monitoring, Laboratory and Treatment Recording, and Tracking of Visits. Each category comprises behaviors that correspond to treatment recommendations and to the patient’s understanding of the natural history and genetics of the disorder. As expected, the frequency and sophistication of self-surveillance behaviors were dependent on age. All patients reported behaviors in each category, but older patients described more elaborate and sophisticated self-surveillance behaviors. Intuitively, that finding seems correct because a patient who has lived with a disorder for many years has had the opportunity to try to refine various self-monitoring behaviors. Self-surveillance behaviors were found in all transcripts, and no category of behavior was unique to one kind of patient (i.e., MEN2a or FAP). Self-surveillance categories and definitions are found in Table 3.

Categories of Self-Surveillance

Medication Appraisal

Medication Appraisal is defined as the evaluation of the use and impact of the pharmacologic management of a health problem or health risk associated with the disorder. Patients attended to the details of their medication schedules, including signs of adverse effects, the cost and availability of medicines, and whether doses were missed. For example, a 49-year-old man with FAP said,

I have a very good system for controlling my bowel problems. At least it works for me, and I’ve told my niece about it, and she seems to think it works well, too. It’s basically a juggling of laxatives and fiber pills. I have learned to be very sensitive to my body, and I take what I need when I need it.

Phenotype Tracking

Phenotype Tracking is defined as the temporal monitoring of physical manifestations of a genetic disorder and the effects of treatment. In that category of behavior, patients tracked changes in their physical appearances. They looked for evidence of symptoms that meant the expression of their genotype (their specific genetic makeup) or represented evidence of recurrent cancer. For example, a 33-year-old woman with MEN2a said,

They told me . . . and I read in an article from my doctor . . . that people with MEN2a can get adrenal [adrenal gland] tumors, too. I have had thyroid cancer so far . . . but lately I’ve been feeling really uptight, really stressed and nervous. These are some signs of adrena problems.
Adherence: monitoring adherence to follow-up visit recommendations

<table>
<thead>
<tr>
<th>Category</th>
<th>Definition</th>
<th>Dimensions</th>
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<tbody>
<tr>
<td>Medication Appraisal</td>
<td>The evaluation of the use and impact of the pharmacologic management of a health problem or health risk associated with a disorder</td>
<td>Prescriptions and refills: evaluating accuracy and timing of refills</td>
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<td>Cost of medication: monitoring changes in costs of prescriptions</td>
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<td>Availability of medication: ensuring the availability of sufficient quantities</td>
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<td>Adherence: monitoring the scheduling and dosage of medications</td>
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<td></td>
<td></td>
<td>Effects of medication: monitoring desired effects and adverse effects</td>
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<tr>
<td>Phenotype Tracking</td>
<td>The monitoring and tracking of the physical manifestations of a genetic disorder and the physical manifestations as a consequence of treatment</td>
<td>Physical appearance as a consequence of genotype: tracking the physical appearance for changes in skin, hair, body shape, lymph nodes, and weight</td>
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<td>Physical appearance as a consequence of treatment: tracking the physical appearance for changes in the visibility of scar tissue and incision lines and changes in scar tissue</td>
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<td></td>
<td>Signs of changes in the already expressed genotype: tracking physical feeling, nonvisible signs, and well-being for evidence of recurrent disease</td>
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<td></td>
<td>Signs of expression of another aspect of the genotype: tracking physical feelings, nonvisible signs, and well-being for evidence of the expression of a previously unexpressed health risk associated with the genotype</td>
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<td></td>
<td></td>
<td>Keeping family history: making and updating family pedigree</td>
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<tr>
<td>Intake and Output Monitoring</td>
<td>The monitoring of dietary and fluid intake and elimination and the impact on the body</td>
<td>Quantity of dietary intake: observing the amount of foods taken at each meal and daily</td>
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<td>Type of food intake: observing the kinds of foods consumed</td>
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<td>Weight changes: monitoring fluctuations in body weight and dimensions</td>
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<td>Food and fluid tolerance patterns: observing the physical and functional impact of kinds and quantities of foods and fluids</td>
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<td>Elimination patterns: observing the patterns of elimination</td>
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<td>Effect of intake and output disturbances on other activities: monitoring the effect of symptoms on rest and sleep</td>
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<tr>
<td>Laboratory and Treatment Recording</td>
<td>The keeping of records on laboratory test results and treatments</td>
<td>Laboratory test results: storing and listing results from diagnostic and follow-up tests</td>
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<td>Surgical or medical treatment: storing and listing results from surgical and medical treatments</td>
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<td>Matching diagnostics to feelings: comparing physical feelings and well-being with laboratory findings and medical treatments</td>
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<tr>
<td>Tracking of Visits</td>
<td>The tracking of frequency and adherence to scheduled follow-up visits with medical professionals</td>
<td>Transfer of laboratory findings: ensuring that laboratory test results are delivered to the respective medical professionals</td>
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<td>Kinds of visits: tracking the kinds of professionals seen and reasons for follow-up visits</td>
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<td>Adherence: monitoring adherence to follow-up visit recommendations</td>
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</table>

Right? I’ve had this MEN2a thyroid cancer for about six years, so it’s just a matter of time before I get the other stuff.

**Intake and Output Monitoring** is defined as the monitoring of dietary and fluid intake and elimination and the impact on the body. This category included tracking how much patients were consuming and whether the consumption of certain kinds and quantities of foods and fluids had an impact on their health and well-being. The category included monitoring changes in their weights and body dimensions and evidence of food intolerance and elimination patterns. It also included the assessment of sleep disturbances because frequency of bowel movements was the most often-cited reason for lost sleep for patients with either MEN2a or FAP. An example of Intake and Output Monitoring is seen in the statement made by a 57-year-old man with FAP.

Since I had the Whipple procedure, I’ve had trouble gaining weight, which my doctor told me to try to do. I can’t seem to keep any weight on. Foods . . . especially some kinds of foods . . . go right through me. . . . It’s really discouraging to be this thin and have no energy.

**Laboratory and Treatment Recording** is defined as the keeping of records on laboratory test results and treatments.

The category included the specific behaviors of collecting and recording the dates and results of laboratory tests, diagnostic evaluations, and treatments. In that category, patients attempted to match test results with physical feelings (e.g., symptoms). For example, one man with MEN2a was continually surprised by the incongruence between how he felt and what the laboratory tests were reporting.

I really don’t get it. . . . My calcitonin level was, again, through the roof [approximately 6,000 with a reference range of less than 20] . . . even higher than the last time. . . . Shouldn’t I be feeling worse than this? Shouldn’t they be able to find out why? . . . As long as I’m feeling good, these tests don’t make much sense. . . . So why do they keep taking them and doing nothing?

This interesting behavior occurred regularly in patients and reflects their need to match empirical and intuitive data with theoretical or scientific data.

**Tracking of Visits** is defined as the tracking of frequency of adherence to scheduled follow-up visits with medical professionals. The category included making and keeping appointments, ensuring that physicians had received laboratory results, and recording the kinds of visits and reasons for them. For example, a 30-year-old woman with FAP shared the following experience.
If I don’t keep track of who I should see and when, no one will. . . . I have to. . . . I mean, I see so many doctors every year. There’s my oncologist, my gastroenterologist, my general doctor [who takes care of my regular issues, like blood pressure checking], and my chiropractor . . . and I always, without fail, bring my latest lab tests. I really hate when they say [the test results] weren’t sent to them by the lab.

As the patients recalled their participation in lifelong surveillance, they linked surveillance with an emotional response. Informants offered discreet statements of their feelings before and after the behaviors.

Feelings Associated With Self-Surveillance Behaviors

Self-surveillance behaviors are associated with a range of emotions. Each category of surveillance behavior was associated with negative and positive emotions before and after the surveillance behavior (see Table 4). The threat of cancer, loss, or disability dominated the emotions expressed before self-surveillance behaviors. The emotion of feeling vulnerable spanned a range of severity from mild concern to anxiety. One man with FAP reported, “I worry about [bowel] obstruction every time I feel even a little pain in my stomach.” Often, patients offered only clues to their feelings rather than stating them explicitly. For example, a woman with FAP stated, “I look at my stool every time I [have a bowel movement]. I never see blood, but I don’t feel right unless I check.” The statement suggests a prebehavior feeling of anxiety because of an atypical need to check her stool on every occasion.

Some behaviors were more likely to be precipitated by negative emotions than others. For example, the behavior of “observing the physical or functional impact of kinds and quantities of foods and fluids” was preceded by patients’ reports of uncertainty, concern, irritation, vulnerability, isolation, and feeling alone. The initiation of that behavior also was linked with patients feeling the need to better understand and the need to control some aspect of their care. Emotions that were dominant before self-surveillance were the need to control and understand fear, vulnerability, and worry.

Self-surveillance behaviors concluded with emotional reactions that, similar to prebehavior emotions, were variably positive, negative, or a mixture of both. After self-surveillance behaviors, patients reported feeling less worried, relieved, in control, and satisfied. Also, the emotions that were reported after surveillance events depended on the observations made by patients. In other words, when a patient was monitoring the desired or adverse effects of medications, he or she might note a negative effect and feel more anxious or worried. For example, several patients with MEN2a self-evaluated for hyperparathyroidism. The emotional reaction to hyperparathyroidism was the same as for any other disease; patients reported feeling worry, uncertainty, and relief. One man with MEN2a who had his parathyroid glands removed commented, “I don’t keep track of who I should see and when, no one will. . . . I have to. . . . I mean, I see so many doctors every year. There’s my oncologist, my gastroenterologist, my general doctor [who takes care of my regular issues, like blood pressure checking], and my chiropractor . . . and I always, without fail, bring my latest lab tests. I really hate when they say [the test results] weren’t sent to them by the lab.

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When I wake up in the morning and feel pins and needles in my arms . . . I know [before: certainty] that I need some calcium. This usually makes me feel better [after: certainty, relief]. But sometimes it doesn’t help, and then I start to think there is something more going on [after: worry, uncertainty].
Table 4. Self-Surveillance Categories, Behaviors, and Associated Emotions

<table>
<thead>
<tr>
<th>Category</th>
<th>Behavior</th>
<th>Emotions Before Behavior</th>
<th>Emotions After Behavior</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Medication Appraisal</strong></td>
<td>Evaluating the timing of refills and the accuracy of the pharmacy</td>
<td>Concerned, worried, angry, irritated, bothered, out of control, need to control</td>
<td>Irritated, relieved, eased, in control</td>
</tr>
<tr>
<td></td>
<td>Monitoring changes in the cost of prescriptions</td>
<td>Concerned, worried, afraid, angry</td>
<td>Concerned, worried, afraid, angry</td>
</tr>
<tr>
<td></td>
<td>Ensuring the availability of sufficient quantities or availability of medications (market trends and supply and demand)</td>
<td>Concerned, worried, fear, annoyed, at risk (vulnerable), out of control, need to control</td>
<td>Concerned, worried, annoyed, out of control, at risk</td>
</tr>
<tr>
<td></td>
<td>Monitoring self-administration of medications</td>
<td>Worried, guilty, need to control</td>
<td>Worried, in control, less worried</td>
</tr>
<tr>
<td></td>
<td>Monitoring desired effects and adverse effects of medications</td>
<td>Worried, at risk, interested, need to control, need to understand, certainty</td>
<td>Worried, more ill, uncertain, less ill, understanding, certainty, relieved, less worried</td>
</tr>
<tr>
<td><strong>Phenotype Tracking</strong></td>
<td>Tracking physical appearance for changes in skin, hair, body shape, lymph nodes, and weight</td>
<td>Concerned, worried, interested, need to understand</td>
<td>Concerned, satisfied, relieved</td>
</tr>
<tr>
<td></td>
<td>Tracking physical appearance for changes in visibility of scar tissue and incision lines and changes in scar tissue</td>
<td>Concerned, worried, fear, interested, need to understand, hope, need to control</td>
<td>Worried, satisfied, relieved</td>
</tr>
<tr>
<td></td>
<td>Tracking physical feelings and general well-being for evidence of a previously unexpressed (new) health problem associated with the genotype</td>
<td>Concerned, worried, anxious, apprehensive, at risk, need to understand, need to control</td>
<td>Apprehensive, worried, temporarily free, spared or hopeful, relieved, less apprehensive</td>
</tr>
<tr>
<td></td>
<td>Tracking physical feelings, nonvisible signs and well-being for evidence of recurrent disease</td>
<td>Worried, dreading, anxious, afraid, need to understand, need to control</td>
<td>Worried, afraid, temporarily free, hoped, relieved</td>
</tr>
<tr>
<td></td>
<td>Making and updating family pedigree</td>
<td>Need to understand, need to control, interested</td>
<td>Understanding, in control, satisfied</td>
</tr>
<tr>
<td><strong>Intake and Output Monitoring</strong></td>
<td>Observing the amount of food taken at each meal, daily, or weekly</td>
<td>Concerned, interested, need to control</td>
<td>Dissatisfied, concerned, pleased, satisfied</td>
</tr>
<tr>
<td></td>
<td>Observing the kinds of foods consumed</td>
<td>Concerned, worried, guilty, need to control</td>
<td>Certain, self-disappointment, resigned, in control</td>
</tr>
<tr>
<td></td>
<td>Monitoring fluctuations in body weight and dimensions</td>
<td>Concerned, certain, interested, need to control</td>
<td>Self-disappointment, discouraged, concerned, satisfied</td>
</tr>
<tr>
<td></td>
<td>Observing the physical and functional impact of kinds and quantities of foods and fluids</td>
<td>Uncertain, concerned, indifferent, bothered, at risk, isolated (alone), need to understand, need to control</td>
<td>Discouraged, afraid, worried, isolated, alone, encouraged, in control</td>
</tr>
<tr>
<td></td>
<td>Observing patterns of elimination</td>
<td>Concerned, worried, need to control</td>
<td>Concerned, discouraged, in control</td>
</tr>
<tr>
<td></td>
<td>Monitoring associated sleep disturbances</td>
<td>Concerned, isolated (alone), disturbed, sad, discouraged, need to control</td>
<td>Isolated or alone, discouraged, sad</td>
</tr>
<tr>
<td><strong>Laboratory and Treatment Recording</strong></td>
<td>Storing and listing results from diagnostic and follow-up laboratory tests</td>
<td>Interested, concerned, isolated (alone), need to control, certain</td>
<td>In control, certain</td>
</tr>
<tr>
<td></td>
<td>Comparing physical feelings and well-being with laboratory findings and medical treatments</td>
<td>Concerned, worried, confused, suspicious (doubting), uncertain, curious, need to understand, need to control</td>
<td>Confused, suspicious or doubting, relieved, certain</td>
</tr>
<tr>
<td><strong>Tracking of Visits</strong></td>
<td>Ensuring that laboratory test results are delivered to the respective medical professionals</td>
<td>Concerned, angry, irritated or annoyed, need to control</td>
<td>Angry, irritated, relieved, less irritated, in control</td>
</tr>
<tr>
<td></td>
<td>Tracking the types of professionals seen and reasons for follow-up visits</td>
<td>Fear, suspicious, interested, need to control, need to understand</td>
<td>Interested, satisfied, in control</td>
</tr>
<tr>
<td></td>
<td>Monitoring adherence to follow-up visits and treatment recommendations</td>
<td>Concerned, guilty, at risk, uneasy, isolated or alone, need to control</td>
<td>Guilty, relieved, less guilty, less worried, pleased</td>
</tr>
</tbody>
</table>

Note: Bold print indicates a positive emotion.

Future research may reveal that fear of cancer, threat of loss, and disability are not just artifacts or consequences of participation in lifelong surveillance but rather are prerequisite and essential to effective lifelong management. Anticipation of a threat to health experienced as worry and concern ensures that patients attend to personal lifelong healthcare needs. Common wisdom and kindness might guide healthcare professionals to aim to reduce the negative emotions and increase the positive emotions. Paradoxically, healthcare professionals may need to explore ways to optimize a balance of realistic worry and pa-
tients’ need to control with realistic relief and measures of control. (Re)minding, or bringing threat to the fore (Giarelli, 2003), may be the essential component of lifelong self-surveillance.

However, no indication exists that patients received any professional guidance or structured feedback on the observations that they made during self-surveillance. Some informants revealed that they sometimes knew when symptoms or feelings should be reported to their physicians. Most were uncertain of what to do with their observations. Herein lies the most significant disconnect between professional health care and self-management.

Implications and Significance

Although the analysis focused on two specific cancer predisposition syndromes, the experiences of patients with MEN2a and FAP may be relevant for patients with other genetic disorders that predispose people to health problems that are variably expressed over a lifetime. Findings from the current study may be used by clinicians to evaluate the extent and appropriateness of patients’ involvement in lifelong self-management of chronic genetic disorders. Clinicians may use the categories as a guide to assess patients’ knowledge deficits as they relate to the relationship between genotype and phenotype, symptom identification, and reporting problems to physicians.

The experiences of patients with MEN2a and FAP may be relevant to patients with other chronic illnesses besides cancer. Healthcare providers may use the themes of self-surveillance behaviors to better understand barriers to adherence to treatment recommendations. With improved understanding of self-surveillance, healthcare professionals may explore ways to assist patients in adhering to recommended follow-up guidelines and validating self-surveillance behaviors and may explore ways to improve the collaboration and sharing of patient-collected data with healthcare professionals.

Oncology nurses may look to the conceptual model of participation in lifelong surveillance and use what is known of self-surveillance behaviors to establish a meeting of the minds of professionals and patients with genetic cancer syndromes and to understand patients’ self-monitoring styles. Oncology nurses can prepare educational programs, based on the list of self-surveillance behaviors and feelings, to better explain the relationship between phenotype and genotype as well as laboratory test results and physical feelings and to provide anticipatory guidance for the management of associated emotional feelings for patients.

Recommendations

The themes of self-surveillance behavior and associated emotions could be developed into an instrument to assess patient adjustment to living with a chronic illness and the perceived burden of self-surveillance in patients who have FAP or MEN2a. Such a tool could be used in research to test the conceptual model and evaluate the effect of a nursing intervention to guide and promote effective self-surveillance. It also might be used in clinical practice to assess ineffective self-management.

Participation in lifelong self-surveillance is social in nature and such that it is influenced by family members in the same household. The experiences of unaffected family members (e.g., informal caregivers) should be studied and compared to those of patients. Future research may explore the question of why patients continue to engage in rigorous self-surveillance despite limited returns and whether some surveillance activities can be adapted or adopted as planned surveillance needs decline.

Much may be learned by looking for a pattern of emotions before and after self-surveillance. Future studies should investigate whether an increased likelihood exists that a feeling of guilt occurs with a particular kind of surveillance event. That may provide information essential to developing a nursing intervention to assist patients to adapt or participate most effectively in lifelong self-care.

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References


