THE EXPERIENCE OF
COPING WITH HUNTINGTON DISEASE
A DESCRIPTIVE STUDY

by

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CHAPTER I

INTRODUCTION

Huntington Disease (HD) is a degenerative neuromuscular disorder manifested first by loss of voluntary muscle control and later by loss of involuntary muscle control. After years of gradual debilitation, the disorder results in death. HD is a unique genetic disorder in that symptoms generally are not manifested until age 35, well after important life decisions have been made regarding career, marriage, and childbearing. Since each child born to a parent with HD has a 50:50 chance of inheriting the disease, the person at risk enters adulthood with the threat of uncertainty; either taking the "chance" of having children or failing to do so may result in feelings of intense personal loss.

Although there is no known cure or effective treatment for HD, advances in the technology of recombinant DNA have made presymptomatic identification of affected individuals possible. Thus, individuals who have already adapted to living with the stress of being "at risk" are now faced with the dilemma of deciding whether they want to know if they will in time develop the disease.

Surveys indicate that most persons at risk are in favor of the test and believe they would use it if questions of support and confidentiality were considered. An understanding of how such individuals appraise the situation of being at risk and how their

appraisal changes with time and circumstances would assist health care professionals in developing counseling programs in conjunction with carrier testing. This knowledge is necessary if nurses are to assist HD individuals and families in making informed, autonomous decisions about their lives, health, and reproduction, and provide support appropriate for helping these individuals to cope with their situation.

Literature Review

Medical, nursing, and psychological literature related to HD are reviewed in this paper. This review of the literature is organized into four major areas: a general overview on HD, being at risk for the disease, the impact of HD on the family, and issues of carrier testing. The review of the literature is followed by the conceptual framework and research questions for this investigator's study.

Huntington Disease

HD is a genetic disorder named for George Huntington, an American physician who, in 1872, described the three characteristics of the disease: "It is hereditary, often leads to insanity and suicide, and manifests only in adult life" (Huntington, 1872, p. 317). Today it is known that it is passed from one generation to another in an autosomally dominant manner, meaning that each offspring of those affected is at a 50% risk for developing the disease. Males and females are equally affected, and the gene for the disease is fully penetrant (100% of those with the gene will show the symptoms). The mean age of onset is 36.1 years, while fewer than 20% of cases are of the juvenile onset type (Conneally, 1984). Symptoms include involuntary (choreic) movements and

intellectual impairment, which progresses to severe personality and mood disturbances. Cardiovascular disease and pneumonia are the most common causes of death, with choking secondary to aspiration frequently reported. The average length of time a person lives after onset is 17.1 years (Conneally, 1984).

The rate of suicide in a study of nonhospitalized persons with the disease was reported by Reed and Chandler (1958) to be 7.8% in males and 6.4% in females. The rate of 5.7 suicides per 100 deaths for the HD population was nearly 4 times greater than the reported rate of 1.5 for the United States Caucasian population in 1979 (National Center for Health Statistics, 1984).

Incidence, or the number of new cases of the disease in a year, cannot be determined directly due to late onset and the fact that only one third of the persons with the disease may show up as affected at any one time. The problem of determining incidence occurs in this and other diseases with late onset where an individual may die from other causes before exhibiting the disease. Prevalence, the total number of cases existing in a population for a given time and place, can be determined from mortality and morbidity data. A study by Hogg, Massey, and Schoenberg (1979) of death rates due to HD in the United States for 1968-1974 reported that overall rates for Caucasians were 1.2 per million per year and for Blacks 0.4. Geographical data showed the highest mortality rate in the North Central and Western regions and the lowest in the South. Globally, the greatest prevalence is found in populations of Western European ancestry, including those in North

America, Australia and South Africa. The estimated prevalence in the United States of 10,000 to 25,000 persons means there are 20,000 to 50,000 at-risk individuals (Bird, 1985).

There is no cure or preventive therapy for HD. Available treatment is limited to symptom control, and it is only marginally effective. HD has been shown by family studies to be linked to a deoxyribonucleic acid (DNA) marker on chromosome 4 (Gusella et al., 1983). Prior to 1983 there was no predictive test to determine carriers presymptomatically. The predictive test raises the ethical issues of discrimination in employment and insurance coverage. A negative test result may significantly reduce the uncertainty associated with being at risk for HD. However, positive results in persons inadequately prepared to cope with the probability of manifesting the disease may cause chronic depression, anxiety related to the onset of symptoms, guilt about risk to their children, and suicide. The psychosocial implications of risk status for HD were explored prior to the development of preclinical diagnosis; however, it is imperative to consider the additional issues raised by the availability of predictive testing.

At Risk

The at-risk individual enters adulthood and middle age with an ever-increasing threat of developing symptoms of the disease. It is conceivable that an at-risk individual who did not inherit the gene could suffer this uncertainty longer than one affected, since one is not considered free of the disease until remaining symptomless to age 70. This state of uncertainty has been described by one at-risk individual

as "playing Russian Roulette with a two-barreled gun and somebody else's hand on the trigger" (Wexler, 1979, p. 209). Wexler characterizes the state of being at risk as having no control and no means of escape. Other descriptions by at-risk individuals illustrate a similar theme of lack of control (Hayden, 1981).

Elash (1977) studied 3 at-risk individuals who volunteered to participate in structured interviews, undergo psychological tests, and keep a diary for 1 month. His results suggest that those at risk do not deny the existence of the disease so much as they avoid focusing on it. These individuals felt a strong motivation to resolve the uncertainty of the at-risk status, and there was a significant interaction between at-risk siblings--an apparently self-protective selection of a sibling to be "more at risk" than oneself, thereby diminishing one's own fears. Elash noted that intensive study of individuals provides the most effective avenue for collecting and considering significant data which could lead to the development of more general hypotheses. The instruments he used provided data on the individual's conceptualization of his world and on his behavior in order to get at the individual's interpretation of events and expectation of outcomes.

Wexler (1979), a clinical psychologist and herself at risk for HD, interviewed 12 men and 23 woman who were at risk. The subjects were between the ages of 20 and 36, were predominantly middle to lower middle class, and had a mean education level of 14 years. Results are discussed in a thematic style. Key observations were identified. The quality of childhood exposure to HD was found to be a critical

determinant of adult adjustment to genetic risk, that is, at-risk adults retained the image of the illness which they had conceptualized as children. Further, the most frightening aspect of the disease was the loss of intellectual capacities. The terminal aspect of the disease was perceived at the time of diagnosis, not with impending death. One half of the sample would consider suicide if and when they showed signs of deterioration; of those, most had a parent who had attempted suicide. All believed a predictive test should be developed, and two thirds would take it. In addition, all believed counseling should be available with the test. From these observations, the investigator developed counseling suggestions which are applicable to HD families as well as other chronic or terminal diseases. These suggestions include listening, offering hope, anticipatory guidance and relieving guilt.

Impact on Family

The progressive physical and mental deterioration in HD imposes great stress on both the patient and the family. Anxiety, regarding the mode of inheritance and anticipation of the disease in offspring, contributes to the distress experienced by all family members.

Tyler, Harper, Walker, Davies, and Newcombe (1982) examined the effects of HD on employment, hospitalization needs, and financial burden to the state among 92 patients with HD in South Wales. This survey of the socioeconomic effects was part of a long-term prospective study of the disease for the purpose of examining a program of systematic genetic counseling. Results were compared with a matched control group. Although a rare disease, the impact of HD was found to be great,

particularly in hospital bed usage. A later study by the same authors (Tyler, Harper, Davies, & Newcombe, 1983) using the same sample, examined the relationship between family breakdown and stress in families with a parent diagnosed with HD. Interviews with reliable family members revealed that marital breakdown for persons with HD was higher with onset under 40 years of age. Reasons for divorce and separation were related to the affected person's behavior, violence, and promiscuity. The implications to children were of two types: (a) If ties with the affected parent were broken before a diagnosis was made, the child grew up unaware of the personal risk; or (b) custody could be awarded to the affected parent leading to possible neglect, abuse and isolation.

When the families described their stress, they identified the following behavioral traits of affected persons: easily upset, confused, demanding, and bad-tempered. Unmet needs for medical supervision and counseling, and in practical and financial crises were high because of the lack of knowledge regarding benefit entitlements, lack of facilities, and lack of understanding by medical and social agencies of the unique problems of these families. The authors concluded that improved management of these families might be enhanced by "reaching out," by improving education and training, and through better access to specialist-advisors for those engaged in their care.

Noting that existing literature on child abuse fails to recognize HD as an important predisposing factor, Pearlstein, Brill, and Mancall (1982) presented a case report of a 9-year-old girl living with her

affected mother in which there was reported neglect, abuse and threatened safety. The authors outlined three interactive factors--personality of parent, vulnerability of child, and environmental stresses--that lead to maltreatment of children, and suggest that the family affected by HD provides a high-risk setting for the pattern of child abuse to develop.

Citing a lack of literature examining the direct effect on the spouse of an affected person, Hans and Koeppen (1980) interviewed 15 wives of men with HD. The two major psychological responses of the wife to her husband's illness were disbelief/denial and resentment/hostility. Needs of the spouse were long term and focused on her individual reaction to stress, responsibility, loss, and single parenthood. The impact of the disease on the spouse is permanent if there are children, because of the repetition of HD in successive generations.

Issues in Carrier Testing

Swavely, Silverman, and Falek (1987) evaluated a group conference format as a means of providing information regarding the presymptomatic test for HD to at-risk families. The Spielberger State-Trait Personality Inventory (STPI), which measures anxiety and anger, was administered to at-risk individuals and their families attending the lecture. The scores for the HD sample were not different from the available normative data.

Attitudinal surveys of those at risk conducted before the availability of the presymptomatic test indicated that 56% to 84% would like such a test if one became available (Schoenfeld, Berkman, Myers, &

Clark, 1984). Attitudes of the impending availability of the test were surveyed from four different parts of the United States, confirmed that most at-risk persons would use the test (Kessler, Field, Worth, & Mosbarger, 1987; Markel, Young, & Penney, 1987; Mastromauro, Myers & Berkman, 1987; Meissen & Berchek, 1987). In three of the four studies, fewer than half of the subjects would use the test for prenatal diagnosis or would terminate the pregnancy if the fetus was found to carry the gene. These surveys also revealed that few at-risk persons are adequately informed about test procedures and limitations.

The focus of this research was on the experiences described by individuals at risk for HD and how these experiences contribute to the coping process. Based on this review of the literature, the following concepts are believed to affect how these individuals appraise and cope with the situation of being at risk for HD: (a) history, (b) impact of disorder on the family, (c) perception of the disorder, and (d) availability of presymptomatic testing.

Conceptual Framework

The conceptual framework for this study is Lazarus and Folkman's Theory of Stress and Coping (Lazarus & Folkman, 1984). The theory contains three major concepts: stress, appraisal, and coping. A schematic representation of Lazarus and Folkman's theory of stress and coping is depicted in Figure 1. Stress is defined as a relationship between the person and the environment that is appraised by the person as taxing their resources and as endangering their well-being. Coping is defined as "constantly changing cognitive and behavioral efforts to

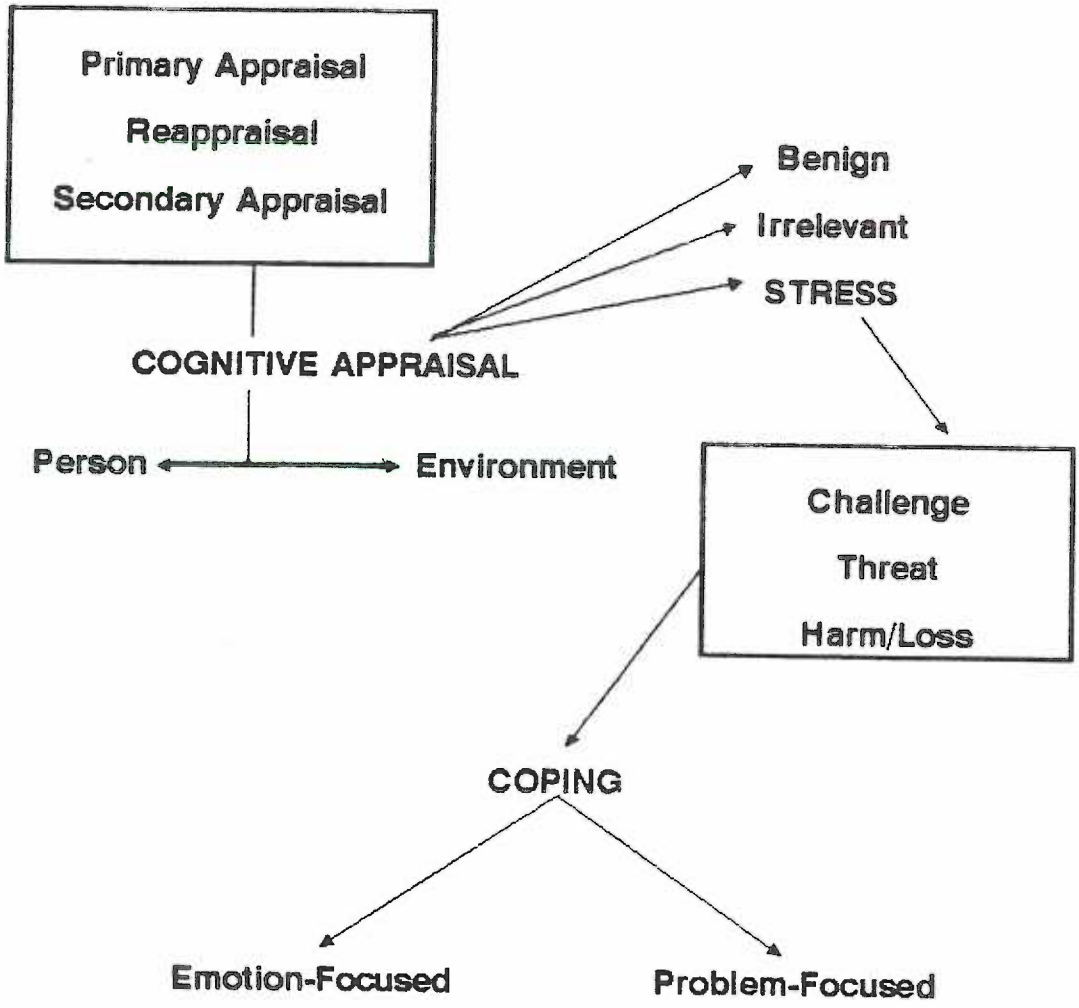

Figure 1. Lazarus and Folkman's Theory of Stress and Coping

manage demands that are appraised as exceeding the resources of the person" (p. 141). Through cognitive processes the individual evaluates the significance of what is happening for their well-being, thereby mediating the quality and intensity of emotional responses. The three kinds of cognitive appraisal are primary appraisal, secondary appraisal, and reappraisal.

Initial judgment of an encounter as irrelevant, benign-positive, or stressful constitutes primary appraisal. Secondary appraisal involves a judgment about what can be done. A changed appraisal based on new information from the environment and/or person is termed reappraisal.

Person factors (e.g., commitments and beliefs) work interdependently with situation factors (novelty, predictability, and event uncertainty) to determine the extent to which harm/loss, threat, or challenge will be experienced. The significance of situation and person factors for stress and coping is in their interdependence; cognitive processes operate to give weight to one in the context of the other. Commitments and beliefs are the most important person factors affecting cognitive appraisal. Commitments are expressions of what is important to the person, and they influence the choices people make. Beliefs about personal control can be both general (i.e., concerning the extent to which the person believes outcomes can be controlled) or situational. Situational appraisals of control can refer to environmental expectations or to expectations for control of one's response to the transaction.

Situation factors creating the potential for threat, harm, or challenge are novelty, predictability and event uncertainty. A novel situation is stressful only if there has been previous experience with or general knowledge of the situation as having been associated with harm, danger, or mastery. Predictability is related to event uncertainty in this cognitive model. Lazarus and Folkman (1984) believe that event uncertainty is stressful in that it immobilizes anticipatory coping processes. In addition, mental confusion can result as one considers first one possible outcome and then another.

Three temporal situational factors are imminence, duration, and temporal uncertainty. Imminence refers to the period before an event occurs, and duration to the period of time during which the stressful event is occurring. Temporal uncertainty refers to not knowing when an event is going to happen.

Coping in this conceptualization is process-oriented and has two functions: managing or altering the problem with the environment (problem-focused behaviors) and regulating or controlling the emotional response to the problem (affective-oriented behaviors). Problem- and emotion-focused coping influence each other during a stressful encounter and can thereby facilitate or impede each other (Lazarus & Folkman, 1984).

Subjective self-report is the primary source of data about stress, appraisal, and coping (Lazarus & Folkman, 1984). Intra-individual analysis enables the investigation of person and situation antecedents

of appraisal and coping which can also be used for interindividual comparison (Lazarus & Folkman, 1984, p. 300).

To the individual at risk for HD, the period of time in anticipation of symptom onset can be "filled with dread, quiet apprehension, noisy emotional disarray or intense productivity" (Wexler, 1979, p. 200). In the psychosocial context, stress and threat are equivalent. If the person appraises the uncertain situation as threatening, strategies are developed to remove or minimize the threat. The individual's appraisal of a situation may help explain the variability in coping in the context of the HD family environment.

Purpose of the Study

The purpose of this study was twofold: (a) to identify concepts affecting appraisal and the process of coping among individuals at risk for HD, and (b) to explore the difficulties these individuals experience with understanding and coping with the new diagnostic technology (i.e., the availability of the presymptomatic test). Two specific questions were addressed in this study:

1. What are the relationships among the following concepts: (a) history, (b) impact on family, (c) perception of the diagnosis, (d) issues (concerns for future), and (e) emerging concepts?

2. What are the subconcepts and how do they influence the individual's appraisal and coping concerning the availability of preclinical diagnosis?

CHAPTER II

METHODS

Design

A descriptive design using qualitative methods was used for this research. An intensive interview format was used to collect data from 5 individuals at risk for HD to elicit the subjects' perceptions and behaviors concerning the at-risk state. This method yielded a rich description of the experience, because data collection was more in-depth than if the data had been collected using a quantitative approach. The goal of intensive interviewing is to elicit from the interviewee rich, detailed descriptions of experiences that can be used in qualitative analysis (Lofland & Lofland, 1984). It is one of the approaches for field research in natural settings.

The research proposal was submitted to the Oregon Health Sciences University (OHSU) Human Subjects Committee. The study was considered to be exempt from review by the Committee.

Procedure to Secure Subjects

Subjects were selected from a population of individuals for whom confirmation of HD in their families had been established. There were four steps in the process of securing subjects.

Step 1. An alphabetized file identifying patients seen with a diagnosis of HD in their families was reviewed. From this file a list of potential subjects was made. Numbers were assigned to each patient by the investigator.

Step 2. Using a table of random numbers, 20 of these potential subjects were selected, and their family folders were reviewed by the investigator. Persons were removed from the potential subject list if they had been diagnosed as affected by HD, were less than 18 years of age, had been identified as severely depressed, had requested no further contact, or were otherwise considered inappropriate to contact, or lived more than 50 miles from the investigator. The investigator then met a designated staff member who reviewed and approved the list of potential subjects. An address and telephone number for each potential subject was then obtained from their registration forms.

Step 3. Nine persons on the list were sent a letter of introduction inviting them to participate in the study (see Appendix A). This letter included a self-addressed, stamped postcard with an identifying code number. If the individual declined to participate, they were instructed to return the postcard to the investigator, and no further contact was made.

Step 4. Of the nine persons sent information, one returned the postcard declining to participate. One letter was returned as undeliverable, and unable to be forwarded. After 10 days, those individuals who had been mailed letters and from whom postcards had not been received were contacted by telephone. If the person agreed to participate, a time and place for the interview was selected. Two potential subjects could not be contacted using the home phone number listed in their folder. Since five people had already agreed to participate, a follow-up letter was sent to the two who could not be

reached by phone, indicating that data collection was complete (see Appendix B). Both of those letters was returned as undeliverable, and unable to be forwarded.

Procedure for Data Collection

The investigator called the subject to confirm their willingness to participate and to set up the time and place for the interview. Prior to beginning the interview, each subject read and signed an informed consent form (see Appendix C).

Information identifying the subject or setting was excluded from the interview data to protect confidentiality. Data from each interview were identified by the code number assigned during the process of securing subjects. A log indicating the code number and the identity of the subject was kept separate from the interview data. This log contained the subject's name, date of birth, address, phone number, and family number. The log was destroyed once the project was completed.

The data were collected through audio-taped interviews with the subjects. All interviews were conducted by the investigator. Interviews ranged in length from 1 hour and 15 minutes to 1 hour and 45 minutes.

All five audio-taped interviews were transcribed using a word processor. Four subjects requested the return of the audio tapes from their interviews. The investigator honored this request. The remaining tape was erased at the completion of the project. The original transcripts and computer disk have been stored in a locked cabinet. Identifying information is not stored in this file cabinet.

Instrument

The instrument used for data collection was a semi-structured interview guide developed by the investigator (see Appendix D). It consisted primarily of open-ended questions designed to obtain information about the individual's past and present experiences in a family affected by HD. The interview questions began broadly focusing on historical events: "Would you share with me how and when you first learned that you were at risk for HD"? After asking what they wish had been different about anything shared so far, the interviewer moved to questions to help them recall what changes resulted from their parent's diagnosis (impact), and who or what helped most in providing strength, support, and encouragement. Then subjects were asked to explain HD in general, and what it means for them (perception). They were questioned regarding their knowledge of the presymptomatic test, where they acquired information, and asked if they wanted more information. Probes were then used to elicit the subject's attitudes about presymptomatic testing. They were asked to describe their concerns for the future regarding employment and resources available for their care (issues). They were then asked to describe what it is like to be at risk for HD and to offer suggestions they had for those just learning of the diagnosis in their families. Finally, the subjects were asked to suggest what things they think nurses should understand when managing care for families having this disorder.

As a result of the first interview, the leading question eliciting information about the subject's perception of the diagnosis was

clarified, that is, more probes were used by the investigator. Probes listed under family impact were sometimes not applicable if the individual was not aware of changes resulting from the parent's diagnosis or if they were not involved in the care of the parent.

The subject's family was sketched during the interview using the conventional symbols commonly found in descriptions of genetic disorders. This method summarizes family data providing a shorthand method of classifying the data for ready reference. Demographic data not revealed earlier was ascertained by questions at the end of the interview.

Data Analysis

Data analysis began after all interviews were completed. After transcription of the audio tapes, the investigator began by identifying comments that she made. Next, a search for themes, recurring concepts, or significant pieces of data was made. Data were identified as significant if they described a problem, a positive or negative event, or a recurring concept or theme relating to any of the four areas of inquiry: (a) history, (b) impact on family, (c) perception of diagnosis, and (d) issues.

Significant additional information that emerged was highlighted for later analysis. The pieces of data identified as significant by the researcher were highlighted by a symbol on a photocopy of the transcripts and then coded (refer to Appendix E). Coding refers to the process whereby a researcher develops a one- or two-word description or code to describe recurring concepts or themes or data identified as

significant by the researcher (Polit & Hungler, 1987). A classification system for coding enabled the investigator to track any given subject's response back to the original transcription. Identification numbers (subjects 1 through 5) and page numbers were assigned to the working copy beside each excerpt from the interview corresponding to a coded concept. This copy was then cut up and filed into the four major categories: history, impact, perception, and issues.

A significant amount of data focused on history and impact, categories seeming to have great relevance. Eight areas of primary interest to the interviewer were extracted from the historical data, 13 areas from the impact, 5 from perception, and 6 from the issues data. Data from each concept file was subcategorized by use of colored marking pens, yielding a color-coded scheme (see Appendix E). Next, data that had not been categorized according to the subconcepts were examined. Subconcepts added were systematically labeled for tracking frequency of occurrence within a subject and across subjects. Each subconcept was abstracted, revisited and refined. Nurses with expertise in qualitative data analysis assisted the investigator in summarizing the results of the study.

CHAPTER III

RESULTS AND DISCUSSION

In this chapter, the results of the study are presented, followed by a discussion of the major findings. Following a description of the demographic information concerning the subjects, results are discussed according to the two research questions, which included:

1. What are the relationships among the following concepts: (a) history, (b) impact on family, (c) perception of the diagnosis, (d) issues (concerns for future), and (e) emerging concepts?

2. What are the subconcepts and how do they influence the individual's appraisal and coping concerning the availability of preclinical diagnosis?

The chapter concludes with additional findings and a discussion of the appropriateness of the conceptualization.

Sample Description

All 5 subjects were Caucasian and had a parent who had Huntington Disease (HD). They ranged in age from 35-46. There were 4 females and 1 male; 4 of the 5 were currently married, and 1 was divorced. Two of the subjects had no children, 2 had biological children, and another subject had adopted children. Four subjects identified a religious faith which they currently practice. They had all completed high school with the majority having at least 2 years of college education. All were employed, and their family incomes ranged from $25,000 to $60,000.

Research Question 1:

Interrelationships Among Concepts and Subconcepts

For research question 1, data on each concept and subconcept is described, followed by a description of the interrelationship among the concepts. Selected quotations from subjects are used to illustrate the findings.

The questions were open-ended and were organized in sequence beginning with history, moving to impact on the family, then perception of the disorder and finally issues and additional categories that emerged. The open-ended questions and probes are listed preceding the findings.

History

Questions were asked to obtain information about the individual's past and present experiences in a family affected by HD.

1. Age subject learned of disorder: The subjects learned HD was hereditary anywhere from when they were 7 years old to in their mid-20s.

2. Person relating information: Two subjects received information from their unaffected parents, 1 from a sibling, 1 from a physician, and 1, although unaware of HD, put it together after their own research.

3. Information content: Three subjects were told they were at risk (i.e., they could get the disease), 1 had no recollection of what they were told, and 1 was told only of the relationship between the affected parent's suicide and HD.

4. Who/what was most helpful: One subject recalled that a variety of people had kept them informed about HD and stated that what was

helpful was knowing who to go to with their questions. The other subjects went out on their own for information or were vague about who or what was helpful. Additional questioning in this area proved difficult for subjects to answer and did not provide specific information on what was helpful.

5. Who/what was least helpful: Overall, subjects had difficulty identifying things that were least helpful; however, 3 identified issues that revolved around the availability of information. Of these, 1 person's search for a common disease manifestation (i.e., the variability of expression of symptoms) clouded their understanding; 1 identified the lack of anyone to sit down and provide them with the information they needed; and 1 stated the lack of information was least helpful.

6. Information withheld: Three subjects reported that information was withheld from them as a means of protection. Two said information was not withheld, that there was a lack of education or lack of information to explain it.

7. Wish had been different: Two subjects could not identify what they wish had been different, 2 wanted honest information, and 1 reflected on the time when the parent had lived through the disease and for whom drugs might have helped:

> I'm sure this was a difficult thing to deal with. Today where at least there's a little information out, there's some medication for people, and there's a little bit that can be done to help people.

But I think at that time there was nothing, and it must have been a discouraging time for him.

8. Perception change: Responses to the question of how perception of the disorder had changed revolved around (a) the reality of providing care to an affected relative, and (b) their conscious decision not to have children (n = 3). Two stated their negative perception had changed over time to a more positive one. One subject had elected to reverse an early sterilization procedure. In the case where a sibling was affected (n = 4), the reality of the disease was ever present.

I didn't know what was involved in it, what would happen to a person and how their personality would change. Now having been through it [with parent] and going through it now again with my [sibling], it's a real reality of dealing with things.

Impact on Family

Questions were asked to assist subject's recall of what changes resulted from their parent's diagnosis, and who or what helped most in providing strength, support, and encouragement.

1. Family structure: No changes were reported in the family structure (n = 3), 1 did not experience change until their unaffected parent's death, and 1 had been adopted into another family.

2. Family activities: Family activities did not change remarkably (n = 3), and 1 subject took on the adopted family's activities. Isolation was reported by 1 subject and is illustrated in the following example: "[Affected parent] withdrew from outside contact and from the

public . . . [parent] was usually withdrawn, and so all the shades had to be drawn and the doors locked and we didn't invite people in."

3. Friends (confidant): When asked about the availability of a confidant to talk with about HD, the subjects identified friends, siblings, and a babysitter. In 2 cases, no one was identified as a confidant.

4. Lifestyle: No change was reported in lifestyle by 3 subjects. Change in lifestyle was apparent in the subject who combined a caregiver role with their homemaking responsibilities and in the subject who was adopted.

5. Role changes, household chores: Two subjects became caregivers to affected parents, while 1 became more self-reliant and assumed household chores. One subject became a step-sibling and assumed farm chores when the unaffected parent remarried and moved to a farm. One assumed chores in the adoptive family; however, this subject recalled a situation occurring earlier in the biologic family that demonstrates a sibling's caregiver role: "[Sibling] would rush home from school to change my diapers. Because of [affected parent's] loss of coordination, [affected parent] was afraid to change me."

6. Subject's age when parent became symptomatic: One subject did not know when signs of the disease first manifested themselves; however, the other subjects' responses ranged from their parent already being symptomatic when they were born to the subject being 12-years-old.

7. Age of parent at diagnosis: The age of the parent at the time of diagnosis ranged from mid-30s to late 40s ($\underline{n}$ = 4).

8. Lived with parent: Most subjects lived with the affected parent after symptoms appeared (n = 4), the length of time ranging between 4 and 10 years.

9. Cared for parent: Of the 4 subjects living with the parent, 2 were involved in their care.

10. Siblings disagreements about care management: Most subjects did not report disagreements with siblings about their parent's care management (n = 3). However, 1 reported disagreements between the unaffected parent and the subject's spouse surrounding care management, and 1 subject reported their sibling felt the subject was too protective.

11. Wished-for changes in care management: Responses varied as to what would have facilitated care. Assistance and a sense of humor were identified as lacking. Regret was expressed over a long hospital stay which resulted in financial ruin, or the manner in which the parent was "picked up in the park and taken to the (state) hospital," or the unavailability of medications for their affected parent.

12. Support available: Friends and faith were reported to be a source of strength and support (n = 3). Family members were important (n = 2), 1 identifying the affected parent's remaining functional as encouraging and support groups as helpful later on.

13. Changes in attitudes toward life: Appreciation for life had increased in all 5 subjects. One further explained by stating that "part of being alive is to have children of your own . . . it's what we're here for," and another wanting their children to be independent

and self-sufficient. Religious faith strengthened in some (n = 2), while self-determination and inner strength had emerged in others as demonstrated in the following statement: "Make the best of things, be a happy person even though bad things happen to us." One subject had openly rejected religion.

With regard to scientific knowledge, most were encouraged by research (n = 3) and 2 claimed an increased awareness of scientific information about HD. Hope for the future was expressed by the following: "Research is on the verge . . . all they need is one breakthrough, and they're going to find what it is and they're going to find how to treat it."

When asked to describe changes in their attitudes regarding the order and meaning of life, 2 subjects took fewer things for granted, 1 of them stating: "I can't imagine what it would be like to have your parents growing up--I'm envious of friends who have parents." One subject felt things happen for a reason, and 1 stated that "[life] is not always nice," that after their experience with the affected parent came the realization that "there are things in life that were not easy to deal with."

Perception of Diagnosis

Questions were asked to elicit perception of the reason for HD.

1. In general: Subjects found it difficult to differentiate the meaning of HD in general from the personal meaning the disorder had for them. However, HD was identified as a genetic disorder, or "part of our

makeup" by most (n = 3), while 1 simply said "there was a reason for it."

2. Personal: Two who had affected siblings described the difficulty in being asymptomatic, 1 of these expressing "sorrow that we both [subject and sibling] could not have escaped." One felt it was a livable disease until the advanced stages, while another expects to be here a short time and expects the worst.

3. Chance/act of God: When asked if they viewed HD in their family as chance or an act of God, most subjects felt HD occurs by chance (n = 4), while 2 indicated it occurred by an act of God.

4. Knowledge of the presymptomatic test: All had heard about the presymptomatic test, although 1 subject mistook presymptomatic testing to be the same as the testing done at their regularly scheduled evaluation. The sources of information varied from the media to professional. Most stated they kept current (n = 3) while 2 admitted they had not actively sought information. Most subjects wanted more information (n = 3), but were specific in what they wanted and in the source they preferred.

5. Effect of removal of uncertainty: All subjects felt the presymptomatic test could help in family planning, if not for them, for their children's children. Two subjects wanted the test even though there was no treatment, 1 of whom felt there would be less fear of testing if treatment was available and used breast cancer screening as an analogy. One subject was ambivalent about testing for themselves while another would not have the test without an available cure. Two

subjects wanted the test to help in decisions about job and retirement; however, most did not indicate a great desire to end the uncertainty (n = 3). One subject's response was paraphrased to say: by removing the uncertainty about whether one got the gene, one would add the uncertainty about how one would deal with that information. All subjects denied feeling pressure to take the test. Most subjects stated that the age to offer the test varies with circumstances; however, 1 of those felt strongly that the test should be offered before marriage and family, perhaps during the late teens.

Issues

Questions were asked to elicit the subjects' concerns for the future.

1. Employment: When asked to relate their concerns for the future, most subjects did not identify maintaining a job as a major concern (n = 3). One feared losing control, while another wanted to remain productive. Another described their need not to simply maintain a job, but to be able to do their very best.

2. Maintaining health insurance: Although having health insurance was important (n = 4), maintaining it was not a great concern to most subjects (n = 3) (i.e., they were covered on their spouse's plan). One subject questioned the value of health insurance.

3. Who to assist with own care: Two subjects named spouses and family as being available to them to assist with their care and 3 did not know. However, 1 felt they "would be cared for somehow . . . there

are enough people in my life that care enough about me . . . can't imagine being put out on the street."

4. What it is like to be at risk: Fear of the disease, watching self for symptoms, and dread with each evaluation were identified by most (n = 3). Another described guilt about having children, while another felt resentment at the tag "at risk for HD." One subject stated they did not dwell on the disease while another said they "could not put it out of their mind . . . deal with it everyday."

5. Suggestions/advice for HD families: Knowing who to call was suggested as helpful by 2 subjects; one suggested a support group while another suggested family counseling. One subject stated, "be as open as you can with family members . . . keep a sense of humor."

6. Information for nurses: Two subjects did not have any suggestions for nurses or genetic counselors. The remainder (n = 3) stated that nurses or genetic counselors should be supportive but not directive, should know the dilemmas faced by these families, especially around family planning, and should understand the illness and the ramifications on the family. One subject elaborated:

> Families are not happy about the need for care. . . . [they] want to know that their family member is being taken care of--that the person treating them understands that they [affected parent] may do something outrageous, but that that person behaved differently before they were ill. It is very challenging care.

Emerging Concepts

Two additional concepts emerged from the data: one focused on the expression of symptoms and the other dealt with characteristics of the disorder that may affect communication. The variability of expression of symptoms was a phenomenon expressed in themes from the data from 4 of the 5 subjects. One subject expressed their mystification at the variability of symptom expression in their family, some affected family members having more motor impairment while others having more verbal involvement. Another subject described their affected parent's fear that their response would be violent as their parent had shown. The subject felt that this added to the fear of the disease. A speculation the researcher had about the subject whose parent had committed suicide was perhaps that that individual (parent) was influenced by past experience. Could this fear also contribute to early decisions regarding not having children? These perceptions changed over time and with circumstances. The uncertainty of the gene inheritance was compounded by the uncertainty of the manifestation of symptoms.

The dread of the long illness and its progression and not necessarily of death was described by 1 subject at the end of the interview. After obtaining the demographic data about employment and income, the subject said: "There is one thing that I wanted to share . . . I have less of a fear of actually dying of HD than I do of the extended time in getting to death. . . . I fear the illness more than death." This fear of the "extended time in getting to death"

diagnosis rather than death was also observed by Wexler (1979) in her study of at-risk individuals.

The second concept to emerge focused on the communication, or lack of, between the affected parent and the young offspring. The researcher searched for reasons to explain why the affected parent did not communicate directly with the offspring about the disorder, the symptoms, or the family history. The reason could be due to the affected parent's lack of insight about the disease, which has been described as a manifestation of the cognitive impairment charcteristic in HD. Other reasons could be a genuine lack of information, fear, or denial.

Summary of Interrelationships Among Concepts

The quality of the individual's early experience with HD was related to the adult perception of the disorder. Results of interviews of 35 at-risk individuals by Wexler (1979) revealed similar observations. Although 1 subject was involved in their affected parent's care for most of their childhood, the resulting perception of the disorder was that "it's not so bad." This subject reported a supportive family environment, an affected parent who was functional until the later stages, and an extended support system involving friends, priests, and medical personnel which contributed to the coping.

The adults in Wexler's study retained an image of the illness which they had conceptualized as children. Further, the adult optimism or pessimism observed in at-risk persons was directly related to the kind of care they perceive their ill parent received, regardless of the

severity of the symptoms. There was a great difference in the perception of the disorder between the 2 subjects who had witnessed and could recall their affected parent's symptomatology: 1 retained the image of wellness and had an optimistic outlook, the other retained the image of illness and is pessimistic. The "power of positive thought" is a mechanism for relieving the helplessness of passivity for many at risk (Wexler, 1979). Conversely, many in Wexler's study expressed that if they expected and prepared for the worst, they could only be surprised by something positive. Viewing the disease positively or negatively could be identified as emotion-focused coping or cognitive reappraisal.

Wexler's study revealed that a large number of at-risk individuals felt emotionally convinced that one of their siblings must develop the disease and subsequently express relief when they do not develop symptoms. This was found in 1 subject, the youngest of a sibship of three, whose symptom-free siblings were certain the subject would develop the disease. In contrast, 1 individual who experienced being adopted out and subsequently feeling "different" stated that their experiencing of symptoms might be different as well. This subject did not express great relief that they had past the age when their parent or sibling had shown symptoms.

One quarter of the subjects in Wexler's study had children after learning of the nature of the disease. This was true for 1 subject in the current study. It was suggested that if those at risk forego having children or choose to adopt, that they are acting as if the illness were a certainty. Three subjects in the current study did not have children

or had adopted. Lazarus and Folkman (1984) suggested that event uncertainty is stressful because of the individual's struggle with coping strategies for an event's occurrence on the one hand, and competing strategies to anticipate the event's nonoccurrence on the other hand. Childless subjects feel that they are faced with the choice of guilt or self-deprivation (Wexler, 1979). Feelings of guilt were identified in 2 subjects in this study, 1 of whom had decided to have children despite their knowledge of the disease. Feelings of self-deprivation were expressed by 3 subjects.

Individuals cope with the uncertainty of the disease by living for the moment ("quality vs. quantity"), and feeling driven to accomplish much or something of value in a short period of time. Many in Wexler's study experienced an increased willingness to take risks and that the courage to change something in their lives came directly from the knowledge that they were at risk. This philosophy was present in most of the subjects interviewed in the present study. The association of death with symptom onset was expressed by the subject who "feared the illness more than death."

The strongest relationship found between the concepts studied related to the subjects' experiences with what they perceived were the needs of families newly diagnosed. They identified strengths and deficiencies that were significant to them and suggested how these experiences could be helpful to others. This process drew from their histories, their perceptions and their concerns for the future. All of

the subjects held great hope for research that will result in a cure for the disorder.

Research Question 2: Influencing Concepts Affecting Appraisal and Coping

The availability of presymptomatic testing was thought by the researcher to be an important concept. Subjects were asked what they knew about the presymptomatic test, about their source of information and if they had considered what it would be like to have the uncertainty removed. The instrument used for data collection was not designed to address this question, and the major concepts of history, impact on family, perception, and issues did not seem to have a direct relationship to the availability of presymptomatic testing. Results from the data show that although the subjects felt the information would be helpful in family planning, they did not feel pressure from any source to pursue it. Two subjects wanted the test and felt they could handle the results, 2 were ambivalent, and 1 showed little interest. The literature reveals that most persons at risk for HD feel a presymptomatic test should be available. Attitudes from this sample toward the test and its use were no different. None had actively pursued the testing for themselves, but most were interested and wanted to remain informed about the availability of the test.

Additional Findings

There was one additional finding that did not directly tie into the research questions. When asked to describe what it is like to be at

risk, 1 subject stated that they "could not put it out of their mind," that they "deal with it everyday." The other subjects' responses suggest that although they share the uncertainty, they do not dwell on it but deal with it in their own way. Another subject did not like the tag "at risk"; perhaps it served as a reminder. This tendency to avoid painful situations is true of most people. However, for this population it has been described as common. Elash (1977) cautioned that it should not be confused with denial. The need to avoid painful situations may contribute to the understanding of the passive information-gathering described by 2 subjects; the use of support groups for information, not support, reported by another; and the avoidance of counseling identified by 1.

Assessment of Appropriateness of Conceptual Framework

The transactional orientation to stress is consistent with nursing's view of human experience and permits the researcher to focus on individual differences in stress experiences (Lyon & Werner, 1987). This study has focused on the interactive concepts of the person and environment that influence how the individual at risk for HD appraises the situation of event uncertainty. An effort was made to identify the appraised meanings of situations (which the researcher called perceptions) which influence how a person copes with event uncertainty.

This framework of stress, appraisal and coping described by Lazarus and Folkman (1984) seemed appropriate because it is process-oriented, reflecting the constant change that requires a person to shift

strategies as the status of the person-environment relationship changes. This process could be used in observing the long-term outcome. Coping changes to meet new demands, and as illustrated in the data of the present study, is facilitated by resources or impeded by high levels of threat.

Mishel (1988) developed a theory of uncertainty in illness from knowledge derived from nursing and other disciplines. The model offers an interactionist perspective to display the relationship of concepts within three major themes: (a) antecedents of uncertainty, (b) process of uncertainty appraisal, and (c) coping with uncertainty. Conceptual relationships have been confirmed in repeated investigations; however, further study using different patient populations in various settings has been suggested to enhance generalizability in nursing practice. Individuals at risk for HD would seem an appropriate population in which to test this theory.

CHAPTER IV

SUMMARY AND RECOMMENDATIONS

Environments and the families of origin for individuals at risk for Huntington Disease (HD) vary widely. In addition, individuals differ in their reactions to threat. Further, individual's decisions may change over time as they confront life crises. Since at-risk individuals share one thing in common, event uncertainty, the researcher studied the interrelationship of concepts from the person and environment and how these concepts related to the individual's appraisal (perception) of HD and presymptomatic testing. This qualitative study obtained data about the process of appraisal and coping in 5 individuals at risk for HD. The researcher hoped to describe the fears and hopes that emerged from interviews with persons coping with this unusual life situation.

Relationships among the following concepts were examined: (a) history of experience with HD, (b) impact on family, (c) perception of diagnosis, and (d) issues (concerns for the future). Issues around presymptomatic testing were explored; however, very little information emerged due in part to the design of the study.

This chapter presents a summary of the results of this study. Research methods are evaluated with limitations of the study identified. The implications for nursing practice and potential utilization of the findings are then discussed. Finally, recommendations for future research are suggested.

Summary of Research Methods

The researcher used convenience sampling to select 5 subjects living in the Northwest for participation in a one-time interview. The purpose of the study was to learn more about the experience of coping with HD in at-risk individuals. Most recent research has been done with attitudinal surveys regarding the availability of presymptomatic testing. This study elicited information from the individuals' experiences and looked for relationships between past experiences with HD and their perception of the disorder. The generalizability of the results are limited due to the small sample size.

Soliciting subjects by mail for voluntary participation in the project had numerous advantages. It provided them with time to consider participation, telephone numbers of persons to contact for more information and a mechanism for declining without an interaction with the researcher.

Qualitative methods, in general, have been criticized because of the difficulty of analyzing the large amount of narrative material in an objective and replicable fashion. Taylor and Bogdan (1984) identified the drawbacks of interviewing as follows: (a) interviews are subject to the same fabrications, deceptions, exaggerations, and distortions that characterize conversation; (b) people say and do different things in different situations; and (c) interviewers are deprived of the context necessary to understand many of the perspectives in which they are interested. However, the open-ended questions and interview method used in this study allowed for the collection of more complete data of the

subject's experience than could have been obtained from a written questionnaire with close-ended questions. The interview and subsequent transcription was useful in identifying new concepts that affect the process of appraisal and coping. Intensive interviewing requires a great deal of time which limits the number of subjects. This method, combined with the small sample size, limits the certainty one can place in the results.

Implications for Nursing Practice

Families in which HD has been diagnosed continually interface with the health care system, whether it be for diagnosis, treatment, or counseling. The major task of those at risk is in learning to cope with what is not known. Treatment must center around assisting the at-risk person and their family by offering them means of controlling the anxiety generated by ambiguity (Wexler, 1985).

Data from this study support the importance of knowing who to contact for specific needs and services. Second, support groups may best serve the needs of all by alternating the focus between education and psychosocial support in order to meet the changing needs of individuals. Third, the results of this study should encourage genetic counselors to examine methods to maintain support services for this population which include options for group support as well as one-on-one support. The findings accent the variability of needs of persons at risk for HD.

Nurses must be educated about the impact of the disorder on families. It is not reasonable to expect all nurses to know about HD;

however, those working with families in genetic services are continually assessing family strengths and identifying needs. These nurses should utilize and participate in research in order to optimize the care provided to these families.

Suggestions have been made in this study by the subjects (Chapter III) and by researchers (cited in the review of literature) to assist those providing services to individuals at risk for HD and their families. The need for sensitivity in communication is essential. HD is a progressive disease to an individual affected and a pervasive disease in its impact on the family. HD can strain the family's coping skills resulting in a breakdown of close interpersonal relationships, job performance, and emotional stability.

Recommendations for Future Research

Research has not been done quantifying the variability of person and situation factors and measuring outcome. It has been suggested that a longitudinal design is the most appropriate method for studying the process of coping in individuals at risk for HD.

Presymptomatic testing is predicted to become more available in the near future. It is imperative that the needs of those at risk for HD be recognized when considering the issues raised by the availability of predictive testing. Research is needed to understand the factors contributing to the individual's appraisal of the situation of being at risk, how to assess their coping over time and to evaluate the counseling programs in place at testing centers. Another population of

persons sharing the uncertainty component and worthy of study are those individuals at risk for Acquired Immune Deficiency Syndrome (AIDS).

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APPENDIX A

INVITATION LETTER

Dear Respondent:

I am inviting you to participate in a study entitled "The Experience of Coping with Huntington Disease". This study is being done in partial fulfillment of the requirements for a Master's degree in Nursing for the principal investigator, Margaret Hale, R.N., B.S.N., under the direction of Marie Scott Brown, PhD. The purpose of this study is to provide knowledge for health professionals on the coping process of individuals at risk for Huntington Disease. It is also to provide a basis from which future studies could investigate the effectiveness of these coping methods. The Chair of Medical Genetics, Robert Koler, M.D., and Ellen Magenis, M.D., the Director of Clinical Genetics at OHSU have approved the project.

If you agree to participate in this study, you are asked to consent 1) to be interviewed in your home or at a place convenient for you and 2) to the use of a tape recorder during the interview. The interview will take between 1-2 hours. It is possible that the research may produce some anxiety as you recall painful significant life events. There will be no benefits to you personally, other than a chance to talk about your experiences. All information you provide will remain confidential to this research project.

If you do not want to participate please return the enclosed postcard. If you do not return this card you will receive a follow-up phone call verifying that you received the information about this research. If you have any additional questions, you may contact me at (503) 279-7577 or Dr. Brown of the Family Nursing Department, (503)279-8382.

I appreciate your willingness to consider participating in my research. If you want additional information on Huntington Disease, available support groups, or clinical services, you may contact Carol Christianson, M.S., Genetic Counselor at OHSU, (503)279-8344. There is a toll-free number that you can call in Oregon, 1-800-452-3563, asking for extension 7577 to reach me or 8344 to reach Carol.

Sincerely,

Margaret Hale
Family Nursing Graduate Student

APPENDIX B

FOLLOW-UP LETTER

April 6, 1989

Dear

I recently sent an invitation to you to participate in my Family Nursing research. I attempted to follow-up on that letter by calling you at home. The number I have is apparently incorrect.

I have finished with my data collection but my interest in the support of families with Huntington Disease continues. Please feel free to call me at 279-7577 or Carol Christianson, Genetic Counselor, at 279-8344 for more information.

Sincerely,

Margaret Hale

Margaret Hale, R.N.
Family Nursing Graduate Student

APPENDIX C

CONSENT FORM

CONSENT FORM

Oregon Health Sciences University

Respondent's Name:_____Date:_____

1. I hereby authorize Margaret Hale to perform the following study entitled: The Experience of Coping With Huntington Disease. I understand that this is a study of how individuals at risk for Huntington Disease describe the situation.

2. I consent to having a researcher interview me one time, for one to two hours, at my home or a place convenient to me.

3. I understand that participation in this study may cause some anxiety as I recall painful significant life events. I understand that while I am encouraged to answer all interview questions, I am under no obligation to do so.

4. I understand that although the purpose of this study is to assist other nurses in understanding the experience of being at risk for Huntington Disease, there will be no benefit to me personally.

5. I understand that my responses will be kept confidential at all times. Neither my name nor my identity will be used for publication or publicity purposes.

6. I understand that I have the right to information as to the results of the study in which I am participating.

7. I understand that I may refuse to participate, or withdraw from this study at any time without affecting my relationship with, or treatment at, the Oregon Health Sciences University (OHSU). Because of the researcher's affiliation with OHSU, I understand the following statement applies:

> "Please note that it is not the policy of the Department of Health and Human Services, or any other agency funding the research project in which you are participating to compensate or provide medical treatment for human subjects in the event the research results in physical injury. OHSU, as an agency of the State, is covered by the State Liability Fund. If you suffer any injury from the research project, compensation would be available to you only if you establish that the injury occurred through the fault of OHSU, its officers or employees. If you have further questions, please call Dr. Michael Baird, at (503) 279-8014."

8. I acknowledge that Margaret Hale (investigator) has explained the procedure and the need for the research. I freely and voluntarily consent to my participation in this project. I understand that I may keep a copy of this consent form for my own information.

9. I also acknowledge that Margaret Hale has offered to answer any questions I might have and that she can be reached at (503) 279-7577.

I have read the foregoing and agree to participate in this study.

Volunteer_____ Date_____

Witness_____ Date_____

APPENDIX D

INTERVIEW GUIDE

Code No._____ 53

Interview Guide

Date_____

Length of interview_____

I am interested in learning from you what it is like to be at-risk for HD.

As the interview proceeds, I will be sketching your family using a genogram format.

1. History. Can we start from the beginning? Would you share with me how and when you first learned that you were at-risk for HD?

Probes: How old were you when you learned HD was hereditary?

who told you?

what were you told?

who/what was most helpful in understanding?

least helpful?

did anyone ever withhold information?

What do you wish had been different about any of what you have shared with me so far?

How have perceptions of disorder changed from diagnosis to the present?

2. Impact on family. There has been a fair amount of literature describing the impact of HD on the family. Tell me what you remember about the period of time when you knew something was wrong but did not know what?

What changes resulted from your parent's diagnosis?

Probes: family structure?

activities?

friends (confidant)?

life-style?

role changes, household chores?

age parent first showed signs of HD?

age parent diagnosed with HD?

did you live with parent after the symptoms appeared? How long?

Were(are) you involved in the care of your parent?

Tell me what a day is like for you as a son/daughter of a parent affected with HD.

Probes: siblings disagreements about care management

what changes would you like to see in how

care was/is handled?

Who or what seemed to help most in providing you/your family with strength, support, encouragement?

in early months

later when things got rougher

Have your attitudes or value toward life changed?

Probes: about what is important

about religion or spiritual beliefs

about scientific knowledge

about the order and meaning of life in general

3. Individual's perception of the diagnosis. How would you explain the reason for HD?

Probes: in general

for you

chance/act of God

Several reports have been published of surveys of attitudes toward the presymptomatic test. Tell me what you know about the test.

Probes: source of information

do you keep current

do you want more information?

Surveys have shown that many people at-risk would take the test. Have you thought about what it would be like to have the uncertainty removed and to know while still unaffected if you have inherited the gene or not?

Probes: affect on family planning, marriage, children

effect of no treatment for disorder on choice

to know or not to know.

feeling pressure to take the test

at what age should the test be offered

4. Issues

There have been concerns raised about unanswered legal and ethical issues of identification of individuals with HD, i.e., job discrimination, denial health insurance, etc. What are your concerns about your future?

Probes: maintaining a job

maintaining health insurance

who(family) might be available to assist with your care.

Is there anything you would like to add about what it is like to be at-risk for HD?

Do you have any suggestion or advice for those who are just learning of the diagnosis in their family?

What would you tell them?

Is there anything else you feel would be good for nurses or genetic counselors to understand?

Do you have any questions of me?

(If the following information has not been revealed during the interview, the respondent will be asked:

highest level of education completed

religious preference

employment type

estimated household income

Your willingness to share your experiences with me is greatly appreciated. I will be calling you tomorrow to learn how you are feeling after this interview.

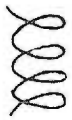
Genogram:

APPENDIX E

CODING SCHEME FOR CONCEPTS AND SUBCONCEPTS

CODING SCHEME

FOR CONCEPTS AND SUBCONCEPTS

	Symbol	Color
I. HISTORY		
1. Age subject learned of disorder		pink
2. Person relating information		orange
3. Information content		blue
4. Who/what was most helpful		green
5. Who/what was least helpful		black
6. Information withheld		red
7. Wish had been different		fuschia
8. Perception change		brown
II. IMPACT		
1. Family structure		pink
2. Family activities		orange
3. Friends (confidant)		blue
4. Lifestyle		green
5. Role changes, household chores		black
6. Subject's age when parent became symptomatic . . .		red
7. Age of parent at diagnosis		fuschia
8. Lived with parent		brown
9. Cared for parent		yellow
10. Siblings disagreements about care management . . .		purple

	Symbol	Color
II. IMPACT (Continued)		
11. Wished-for changes in care management		aqua
12. Support available		turquoise
13. Changes in attitudes toward life		lavender
III. PERCEPTION		
1. In general		purple
2. Personal		blue
3. Chance/act of God		green
4. Knowledge of presymptomatic test		orange
5. Effect of removal of uncertainty		pink
IV. ISSUES		
1. Employment		fuschia
2. Maintaining health insurance		lavender
3. Who to assist with own care		blue
4. What it is like at risk		green
5. Suggestions/advice for HD families		orange
6. Information for nurses		yellow

AN ABSTRACT OF THE THESIS OF
MARGARET HALE, B.S.N.

For the MASTER OF NURSING
Date of Receiving this Degree: May 15, 1989
Title: THE EXPERIENCE OF COPING WITH HUNTINGTON DISEASE

APPROVED: [redacted]
Marie Scott Brown, RN, PhD, Professor, Thesis Advisor

The purpose of this descriptive study was to identify factors affecting appraisal and coping among individuals at risk for Huntington Disease (HD). Specifically, the study examined relationships among the following concepts: (a) history, (b) impact of disorder on the family, (c) perception of the disorder, and (d) availability of presymptomatic testing.

The conceptual framework used was based on Lazarus and Folkman's (1984) theory of stress and coping. The study focused on the interactive concepts of the person and environment that influence how the individual at risk for HD appraises the situation of event uncertainty.

Data were collected through one-time intensive interviews with 5 subjects at risk for HD, to elicit the subjects' perceptions and behaviors concerning the at-risk state. Individuals were selected from a population among whom HD in their families had been established at a genetics clinics of a medical center in a Northwest state.

The quality of the individual's early experience with HD was related to adult perception of the disorder. Most subjects expressed a drive to accomplish much in a short period of time as a means of coping with the uncertainty of the disease. The individuals

identified strengths and deficiencies related to their experiences and suggested how these experiences could be helpful to others.

The study method, combined with the small sample size, limits ascertainment of significance of the interrelationships among the concepts. The findings accent the variability of needs of persons at risk for HD.