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Development of the Huntington Disease Family Concerns and Strategies Survey From Focus Group Data

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Abstract

Health concerns and management strategies among families of young and middle-age adults with Huntington's disease (HD) are unknown. This study developed and tested psychometric properties of the Huntington Disease Family Concerns and Strategies Survey (HDFCSS). Focus group data from 91 adult family members were used to develop content. Content analysis yielded four domains that were transferred into Personal, Person With HD, Community Health Care Services, and Strategies scales. Focus group data, expert validation, and cognitive interviews demonstrated survey content validity. Cronbach's alpha internal consistency coefficients for the scales were 0.83 or above. The measure can be used to generate reliable and valid data to identify adult family members' health-related concerns and management strategies for themselves and persons with HD.

Keywords

family; Huntington's disease; health management; focus group

Family members of persons with conditions that include cognitive and behavioral impairments face multiple challenges, and care giving by family may have beneficial as well as harmful effects (Aubeeluck & Buchanan, 2006; LoGiudice & Hassett, 2005). Despite the developing body of knowledge on informal caregivers, much of this knowledge reflects care giving by and for persons in their later adult years and has less often included populations with an onset earlier in life. The timing of the onset of a disorder can have profound implications on the lives of people in the family, depending on the life stage of each individual and the family (Rolland & Williams, 2006). Management of chronic conditions by family members often extends beyond care giving tasks for the person with the disorder to include assisting other caregivers, general family management, and health care environment maintenance. Self- and family management of chronic conditions mandates that behaviors to manage a condition occur within the context of one's everyday life (Grey, Knafl, & McCorkle, 2006).

Family management of a condition such as Huntington's disease (HD) is additionally complicated by the heritability of disease risk for the care recipient's offspring and siblings and the availability of predictive testing for persons who have not yet developed clinical symptoms of the condition. HD is an autosomal dominant disorder in which the offspring of each person with this condition has a 50% chance of inheriting the gene mutation and developing the disease. Persons age 18 or over who are at risk for HD can learn the likelihood that they will develop the disease through predictive testing to identify the presence of an expansion in the HD gene (Rosenblatt, Ranen, Nance, & Paulsen, 1999). Although characteristic motor symptoms have not yet appeared in the so-called prodromal phase of HD, diminished ability to perceive one's own behavior creates health management challenges for family members of persons in the earliest stages of this condition (Hoth et al., 2007; Williams et al., 2007). Although approximately 30,000 persons in the United States have HD and another approximately 150,000 are at genetic risk (Dubinsky & Huntington's Disease Peer Workgroup Members, 2004), existing measures do not address the spectrum of issues facing families of persons with HD across the disease trajectory.

The Problem Behaviours Assessment for HD scale (Craufurd, Thompson, & Snowden, 2001) is a measure of behavioral abnormalities in HD for persons with a clinical diagnosis of HD and is completed by health care providers. No existing measures were located that address how family members manage their concerns during the prodromal phase or are intended for family members to complete across the disease trajectory.

Researchers have identified genetic aspects of other neurodegenerative conditions appearing during young to middle adulthood, including early-onset Alzheimer's disease (Ertekin-Taner, 2007), very early onset Alzheimer's disease (Filley et al., 2007), familial amyotrophic lateral sclerosis (Gros-Louis, Gaspar, & Rouleau, 2006), and frontotemporal dementia (Van Deerlin et al., 2007). The families of persons with these conditions may, therefore, face similar challenges as families of persons with HD. This article presents the initial development and testing of the Huntington Disease Family Concerns and Strategies Survey (HDFCSS). It is anticipated that the HDFCSS measure reported in the current article may be adapted for use with other early-onset neurodegenerative conditions for which family members are also at risk.

BACKGROUND AND CONCEPTUAL FRAMEWORK

First described in 1872 (Hayden, 1981), HD is characterized by a triad of symptoms involving loss of motor and cognitive function as well as psychiatric disorders (Rosenblatt et al., 1999). Cognitive impairment includes alterations in memory, psychomotor processing, and executive functioning. These symptoms frequently precede the clinical diagnosis that is

made when characteristic movements are present in persons with a family history of HD or a gene test produces positive results (Paulsen & Conybeare, 2005). Symptoms of HD are caused by the abnormal expansion of CAG repeats in the *IT15* gene (Huntington's Disease Research Collaborative Group, 1993). The average age of diagnosis of symptoms is 39 years, and the average survival rate is between 15 and 20 years after diagnosis (Rosenblatt et al., 1999). Treatment is symptomatic, and at this time there is no cure.

Self- and family management of the illness can reflect multiple factors, including the condition, individual factors, family factors, and the environment, that encompasses social, community, and the health care system (Grey et al., 2006). Despite these far-reaching implications, little is known about how families manage HD, especially within the context of changes in symptoms from the prodromal phase through end of life. Researchers document distress in partners of persons receiving a positive predictive HD test (Quaid & Wesson, 1995; Sobel & Cowan, 2000), and Decruyenaere et al. (2005) suggest that this distress may reflect not only feelings of loss and grief but also thoughts regarding a future role as a caregiver. Family members assume substantive care giving responsibilities for persons with HD and experience personal distress, yet the authors found no measures that would provide estimates of the nature, frequency, and intensity of responsibilities or of the extent of distress experienced by family of persons with HD. Furthermore, no instruments were located that measured strategies used by family to manage their responsibilities and concerns. The purpose of this study was to identify the health management concerns and strategies of family members of persons with a mutation in the HD gene. In order to accomplish this purpose, the authors developed a quantitative measure of family member concerns for themselves, their families, and the person with HD, including concerns regarding community health care services, and strategies that family members used to manage these concerns.

PROCEDURES FOR INSTRUMENT DEVELOPMENT

Development of an instrument for research includes domain identification, item generation, and instrument formation (Lynn, 1986). Although literature review and expert advice are sources of information for survey development, they may not provide insights into the points of view of persons who are members of the population under study (Willgerodt, 2003). When developing measures of poorly understood phenomena, qualitative methods to identify relevant and appropriately worded items may include observations, interviews and focus groups (Wackerbarth, Streams, & Smith, 2002). There are several advantages to using focus groups for instrument development: the resulting measure is more likely to be grounded in the experiences of the population, the language and terminology familiar to the group can be used, and focus groups may identify issues of importance to a group that may not be identified through literature reviews or through experts (Willgerodt, 2003). In addition to identification of topics, subscales, and survey items, instrument development includes review of items, pilot and field testing, and conducting psychometric assessment (Gilgun, 2004; Weisberg, Krosnick, & Bowen, 1996). Researchers collected and analyzed data from focus groups during the first phase of development of the HDFCSS.

Focus Group Data Collection and Data Analysis

Focus groups at HD medical and research centers in the United States and Canada were conducted to identify content for the survey. Although prior data from family members of persons with HD from one midwestern state were available (Williams, Schutte, Holkup, Evers, & Muilenburg, 2000), perspectives of family members across geographic settings, ethnicity, and proximity to HD health care expert services were needed to better represent the HD population concerns and strategies.

Descriptions of portions of the sample and focus group procedures are described more fully elsewhere (Sousa, Williams, Barnette, & Reed, 2010; Williams & Ayres, 2007; Williams et al., 2007, 2009;). Briefly, adult family members of persons with HD from prodromal through end of life stages were recruited by site coordinators from HD clinical and research centers. The sample consisted of adult family members of persons with HD across the disease trajectory (Table 1). Human subjects committees at each site approved the study. Focus groups for three categories of adult family members were held at six sites, and a second focus group for additional group of participants was held at the sixth site. The three categories were (a) adults whose family members with HD had a positive DNA test but had not received a clinical diagnosis (Prodromal), (b) adults whose family members with HD had been diagnosed and lived at home (At Home), and (c) adults whose family members with HD lived in health care facilities (Long Term Care Facility). Researchers asked participants to describe their own concerns, concerns regarding the health of the family member with HD, strategies used to manage these concerns, and what could help them. Ninety-one adults ages 25 to 79 enrolled in the focus groups. The majority (74%) were female. Participants were spouses (65%), parents (19%), children (5%), siblings (7%), or other family members (aunt, daughter-in-law) (2%) of persons with HD.

Audiotaped data were transcribed and validated with the audiotapes. Data analysis commenced following completion of the first focus group and continued throughout the remainder of data collection using descriptive qualitative analysis procedures (Sandelowski, 2000). Six members of the research team conducted the initial coding and agreed-on code definitions. All members coded focus group data from the first and second data collection sites to establish consistency of coding. Differences in coding were discussed until 100% agreement was reached. Two or three members of the research team coded data from each subsequent focus group and reviewed findings with the rest of the team at weekly meetings. The research team created a coding scheme at the start of data analysis to capture broad domains of personal concerns, concern about the person with HD including access to health care, and management strategies. Other domains were added that reflected the dimension of time, sources of assistance for problems, and focus group processes. The research team created descriptive codes and placed them under the broad domain that best fit with the team's conceptual definition of the code. Categories within the domains were created when several codes illustrated a concept within a domain. As the team completed analysis of each transcript, one member summarized the results of the coding, which were reviewed by the entire team. When applicable, summaries included comparative remarks to illustrate the differences and similarities between focus groups and between the research sites. Observation notes recorded by each focus group assistant were also entered into analysis. A research team member not involved in the coding scheme development validated the coding scheme.

Development of Survey Items

Data redundancy was apparent after completion of data analysis from the first four focus groups. Research team members identified major conceptual scales for the survey: Personal, Person With HD, and Strategies. After completion of validity procedures, the team created a fourth scale: Community Health Care Services. The Personal scale reflected the family member's own health, including feelings about one's own well-being, and family health, including feelings about other family members at risk for HD. Items pertaining to clinical aspects of HD were placed in the Person With HD scale and included topics pertaining to cognition, behavior, psychiatric symptoms, motor symptoms, interpersonal relationships, functional activities, and health habits. At this time, items referring to interactions with health care providers, health-related services, and the public regarding HD were still in the Person With HD scale. The Strategies scale included the family member's efforts to manage

concerns, whether for self, family, the person with HD, and/or community health care services.

Team members converted coded data addressing the scale topics into declarative survey statements. Team members wrote sample items for each statement using the participants' own words whenever possible. The team reviewed statements at weekly meetings, revising them to improve clarity. Items were compared and condensed by the team to remove redundant statements. Data from the last two focus groups were coded and compared to each scale's content. Research team members revised wording on individual survey items as a result of analysis of the last two focus groups' data, but no new content areas were identified.

Review of Similar Surveys

Following the first draft of survey items, the research team compared the items with content from clinical descriptions of HD (Rosenblatt et al., 1999), research reports on HD, and other HD-related measures. Specifically examined was content describing behavioral symptoms in persons with HD (Craufurd et al., 2001); measures that address care giving for persons with dementia, memory and behavior problems, health care services, and assessment of memory and behavior of persons with dementia (Bakas & Champion, 1999; Davis et al., 1997; Kinney & Stephens, 1989; Mahoney et al., 2003; Montgomery, Gonyea, & Hooymann, 1985; Nijboer, Triemstra, Tempelaar, Sanderma, & vandenBos, 1999; Novak & Guest, 1989; Picot, Youngblut, & Zeller, 1997; Robinson, 1983; Schweibert, Gieordano, Zhang, & Sealander, 1998; Shyu, Lee, & Chen, 1999; Vitaliano, Russo, Young, Becker, & Maiuro, 1991); and family coping with health services (Cummings, Kelly, Holland, & Peterson-Hazen, 1998). Although existing measures contained components that overlapped with the HDFCSS components on personal feelings, personal health, relationships, care giving, the care recipient's health, and the health care system, none addressed strategies, resources, and concerns about risk for disease in other family members. Furthermore, specific topics of concern for family members of persons with HD, such as health habits and function of the person with HD, were unique to the HDFCSS, as they were not included in existing measures.

One measure was located that included needs specifically related to care giving management in a health care facility. This measure (Cummings et al., 1998) contains items to assess perceived psycho educational needs of family caregivers of hospitalized elderly patients. Although not identical to issues faced by family of persons with HD, the four conceptual categories of information, coping, the person, and the hospital in this instrument were reflected in items created in the HDFCSS.

Demographic items were developed through review of existing measures and published research with persons with HD. Questions regarding whether the person with HD had received a clinical diagnosis or was in the prodromal period but had received a positive predictive genetic test for HD were written after comparison of draft items with published studies of persons across the HD illness trajectory extending from the time of a positive predictive test through end of life (Bachaoud-Levi et al., 2001; Benjamin & Lashwood, 2000; Codori & Brandt, 1994; Craufurd et al., 2001). The demographic section contains 26 items regarding the respondent and person with HD. Items included the number of hours spent on care giving, age at diagnosis of the person with HD or age when received positive predictive test result, and the health care provider who made the diagnosis.

Description, Administration, and Scoring of the Instrument

Each scale has a closed-ended question format (Table 2) and yields scores for that scale. In the Personal and Person With HD scales, the respondent indicates frequency from never to several times a week (0–4) and intensity of reaction from no bother at all to extreme bother (0–4) for each concern. After completion of validity procedures, the final number of items in each scale was identified. The Personal scale contains 31 items, and the Person With HD scale contains 36 items. Scores can be calculated for frequency and intensity of reaction to items on the Personal and the Person With HD scales. Higher values indicate higher frequency or higher intensity of reaction. Since it was likely that some items would not be answered, scoring was based on the mean item response for the scales rather than the sum of item scores. For the Personal scale, scores can range from 0, which would represent absence of frequency of any personal concerns or absence of personal reaction, to 4, which would represent maximum possible frequency of personal concern or maximum possible reaction. For the Person With HD scale, scores can range from 0, which would represent absence of frequency of any personal concern about the person with HD or absence of personal reaction, to 4, which would represent maximum possible frequency of personal concern about the person with HD or maximum possible reaction. Each scale ends with an open-ended response opportunity to list up to two items not covered in the scale and to identify the top two concerns from the entire list. This section provides an opportunity to provide the context for an item, to provide more details about an item, and/or to raise a new issue.

In the Community Health Care Services scale, respondents indicate extent of agreement or disagreement and intensity of reaction to each of 14 statements on a 4-point agree-or-disagree scale. The range of possible scores on this scale is 1, which would represent „strong disagreement% with all 14 items, to 4, where the respondent indicated „strong agreement% on every item. This scale ends with the open-ended response section. Scores can be calculated for extent of endorsement and intensity of reaction to statements. On the extent of endorsement and reaction intensity scale, the range of possible scores on this scale is 0, which would represent no intensity of reaction on all 14 items, to 4, which represents the maximum possible intensity of reaction for these 14 items.

On the Strategies survey, respondents indicate whether they use each of 82 strategies and, if so, how helpful that strategy is on a 3-point ordinal scale. Respondents can list up to two additional strategies if not on the list. Scores can be calculated for the frequency and helpfulness of each strategy. This scale was scored in two ways. On the frequency-of-use scale, the scores can range from 0, where none of the 82 strategies were used, to 82, where all 82 strategies were used. On the effectiveness scales, scores could range from 1, where none of the used strategies were considered helpful, to 3, where all 82 strategies that were done were considered very helpful.

METHODS

Expert Validation

Prior to external review, members of the research team conducted a final review of the survey. A panel of content experts representing a range of expertise with families and persons with HD reviewed the survey. In addition, experts from different geographic regions reviewed the survey in order to identify terminology that may be regional in usage. Nine experts conducted the content validity review, which is within the suggested range of 2 to 20 (Grant & Davis, 1997). Experts were clinicians or researchers with expertise in medicine, neuropsychology, families of persons with HD, home care of persons with HD, nursing home care of persons with dementia, and qualitative research with families.

A cover letter provided instructions to each expert, including the purpose of the surveys, the population for whom the survey was intended, and the request for experts to assess appropriateness, accuracy, and representativeness of each item. For the purpose of quantitative evaluation of the survey's validity, experts were asked, following conventional practice (Polit & Beck, 2005), to rate each survey item on a 4-point ordinal scale, with 4 indicating the strongest endorsement of the item by the expert. Ratings were requested for both clarity of the item and relevance of the item to its intended target. Clarity was rated on a scale from 1 (very confusing) to 4 (very clear). Relevance was rated on a scale from 1 (not relevant) to 4 (highly relevant). Surveys and definitions of the concept/topic for each survey were given to each expert. Experts were also asked to suggest rewording of confusing items, identify any items to be added, and comment on the comprehensiveness of the surveys (Grant & Davis, 1997).

For each item, a content validity index (CVI; Lynn, 1986; Martuza, 1977) was calculated for both clarity and relevance by determining the proportion of the experts who chose a rating of 4. Thus, the CVI was based solely on expert agreement on the strongest endorsement, and agreement on a lower level of approval was not counted. After the values for individual items were calculated, a scale-level CVI was obtained by averaging across all items within a content area to obtain an overall value for that content area. The three content areas were Personal, Person With HD, and Strategies. Scale-level CVIs were calculated for both clarity and relevance ratings. For example, an average of the proportion giving a rating of 4 for relevance was calculated over all the items pertaining to the Personal scale. Items for the Community Health Care Services scale were included in the questions pertaining to the Person With HD scale.

Cognitive Interviewing Participant Validation

The research team also validated items in each scale through cognitive interviews. The purpose of the cognitive interviews was to understand how respondents perceived questions and to identify potential problems that may lead to response error (Drennan, 2003; Willis, 2004). Verbal probes were used to ask the respondents to comment on the wording of the items and elicit perceptions of meaning of items (Knafl et al., 2007). A research team member asked 15 adult family members from one state who had participated in the focus groups about the clarity of each item as well as suggestions for improving unclear items.

Procedures for contacting participants from prior focus groups were approved by the institution's institutional review board. Participants were sent an information postcard inviting their participation, and those who were interested received a survey by postal mail. A member of the team contacted each participant to arrange an hour-long appointment for the phone interview. Participants were asked to review the survey prior to the interview. The interviews were audiotaped. During the interview, the interviewer read aloud each item on all scales. Participants indicated whether each item was clear or unclear. If unclear, they were asked (a) to describe what they thought the item was asking, (b) what specifically made it unclear, and (c) for suggestions for improving clarity. The interviewers did not ask the participants to give a response to the survey items. Some participants commented on each item, while others commented specifically on items that were not worded clearly or completely, on items the participants thought should be removed or added to the survey, and on items needing improvements. When a participant made a response, the interviewer recorded the response on a grid that portrayed columns for each participant and rows for each item. For each mark on the grid, the interviewer created a narrative statement indicating the nature of the comment. The team then discussed each participant's comment and/or suggestion and made a decision regarding whether an item was retained, added, or edited. On completing the item revisions, the team separated items referring to health care services from the Person With HD scale, creating the fourth scale, Community Health Care

Services. The team conducted an analysis to determine what changes were made for each item as a result of these cognitive interviews, including the nature of these changes as well as the impetus for changes made.

The first step of the analysis was to compare the first version of the scales with the final version. An item-by-item examination between the first and final versions revealed one or more of the following: (a) the item remained unchanged; (b) the item number changed; (c) a portion of the item wording was added, deleted, or changed; (d) the entire item changed while retaining same/similar meaning; (e) an item was entirely deleted or added; or (f) the item was changed only with minor editorial revisions. For example, the first version of one of the scales included the item „I feel a loss of companionship.% On the final version, this item was „I miss the relationship I had with him/her.% This change constituted a change in the entire item while retaining same/similar meaning and was coded as a 4 using the rating scale established.

Once the types of changes were determined, the next step of the analysis was to link each item from the first version with participant comments made during the interview, regardless of whether a change was made between the two versions or the nature of the change. All participant comments were matched with respective items. The total number of comments made for each item was recorded. This step of the analysis revealed, for example, that although two comments were made on the item „I hold in my emotions,% the team decided to retain this item as originally written.

The next step in the analysis was to examine all participant comments in order to make a conclusive statement for each item in terms of the nature of and impetus for any changes made following the cognitive interviews. Several categories emerged on the basis of this evaluation. The first included all items for which no substantive changes and no participant comments were made. Substantive changes were defined as any changes in an item reflecting additions, deletions, or wording changes; changing an entire item while retaining same/similar meaning; and entire item additions or deletions. Minor editorial changes and changes in item numbers made between the first and final versions were not categorized as substantive changes. Out of a total of 125 items, 33 items were neither commented on nor substantively changed.

The second category included items that remained unchanged between the first and final versions despite participant comments; 34 items were in this category. Participant comments associated with these items were categorized as substantive or nonsubstantive to represent comments that were meaningful or inconsequential, respectively, in terms of potential to contribute to any type of item change. For example, no changes were made between versions on the item „I make changes in our home to keep him/her safe (e.g., removing rugs & other things that can be tripped on).% Two participants commented on this item. The first participant liked the examples listed in the statement (comment deemed nonsubstantive), while the second participant suggested adding „facility% or changing the statement to „I make changes in the home environment% to make the statement more applicable for those with family living in a nursing facility (comment deemed substantive).

A third category included items for which substantive changes were made on the basis of participants' comments. For example, the first version item „I spend time with him/her% changed entirely on the basis of one participant's comment about whether the statement meant quality time or being with him or her because one has to spend time. The final version item was „I make an effort to spend quality time with him/her.% There were 27 such items in this third category. A fourth category included items for which substantive changes were made despite a lack of participant comments. For these such items, expert review, team

discussion, or a combination of these provided the impetus for the change. There were 20 items in this category.

Survey Sample and Procedures

Components of Dillman's (1978) method for mailed surveys were used to distribute the revised HDFCSS. These included an announcement with a reply form and postage-paid envelope from the research team. Recruitment sites included the Iowa HD Registry, the HD Lighthouse (an Internet site), the Huntington Disease Society of America's annual meeting, and the University of Connecticut Health Center, which served as one of the focus group sites. On receipt of the reply form, a questionnaire with a detailed cover letter was sent to participants. If the survey was not returned, the team sent a reminder letter after 2 weeks and a second survey mailing after 3 more weeks. Persons returning surveys received a thank-you letter and a telephone card as a token of appreciation. This method yielded 269 returned surveys out of 390 requests (69%), with 245 usable surveys. This method provided 56% (24/43) from the Internet support group, 69% (25/36) from the clinical site, and 70% (219/311) from the research registry.

RESULTS

Table 3 presents a summary of the validity indices for questionnaire items within content areas. For both the content area of Strategies and the content area of Person With HD, the average validity indices for clarity and relevance were 90% or greater. For the Personal scale, the average validity indices for clarity and relevance were between 70% and 80%. Indices for individual items ranged from 33% to 100% for both clarity and relevance.

While there were not enough subjects to conduct a confirmatory factor analysis for the scales, one indication of validity would be that scales dealing with the same frequencies or reactions would be expected to have reasonably large correlations, while scales that measured frequencies and reactions on other aspects of dealing with HD would be expected to have lower correlations. As indicated in Table 4, this was observed. Correlation between frequency and reaction within the Personal Concerns was .87, within the Concerns Persons With HD it was .82, and within the Community Health Services it was .74. All the expected nonspecifically related variables ranged from $-.30$ to $.64$. The mean correlation for the within-scale variables was .81, and the mean correlation for the non-within-scale variables was .17, clearly a difference in these correlations in a direction that provides support for claiming that the scales are providing scores that validly measure frequency and reaction on different aspects of care giving for individuals with HD.

Estimates of reliability of the data were determined using Cronbach's alpha among items of each scale. Missing items were replaced using the item mean of the responders. Reliability statistics are presented in Table 5. For each scale, the Cronbach's alpha internal consistency coefficient is presented as well as the 95% confidence intervals determined using a program developed by Barnette (2005). The Cronbach's alpha values range from .83 to .94, well above the recommended criteria for determining internal consistency of new scales, set at least at .70 (Nunnally & Bernstein, 1994). Other reliability statistics included the minimum and maximum Cronbach's alpha values if one item was deleted from the scale and the mean and range of item-total correlations. The minimum and maximum values varied little from the overall alpha if an item was deleted. The mean item-total correlations for the eight scales ranged from .37 to .58. It was interesting to note the range of the item-total correlations. On four of the scales, the lower values were not unexpected at a value of .17 or greater. On the other four scales, there were items with relatively low item-total correlations. On further examination, these items tended to be items with low frequencies of occurrence or use. For example, on the Strategies Usefulness scale, six of the items that had the lowest item-total

correlations were also the least used strategies. The analysis of reliability statistics provides evidence that the scales are providing scores that possess the property of internal consistency reliability.

DISCUSSION

The HDFCSS is a self-report paper-and pencil measure that identifies Personal and Person With HD concerns, Community Health Care Services needs, and Strategies for managing concerns. The measure provides data with strong content validity and internal consistency. The survey scales include components of family member experiences that are not represented in existing measures. This measure provides the opportunity to examine the influence of being at risk for HD within the context of managing and/or providing care for a family member with this progressive, incurable illness. Validity and reliability analyses provide support for the claims that the scales provide scores that are internally consistent and differentiate between the specific aspects of concerns and strategies used in care giving for individuals with HD.

The survey aims to assess family concerns and strategies regarding chronic illness. It was designed to be applicable to adult family members of persons with HD across time of the course of the disorder. Findings from this survey may be useful in further application of theories of family management of genetic conditions across the life span. Furthermore, the survey may be modified by removing the term „HD% and inserting the name of other neurodegenerative conditions that share clinical characteristics, such as frontotemporal dementia or early-onset Alzheimer disease. Use of the survey will allow for comparisons of family management across these conditions as well as comparisons across health care systems. Many chronic conditions extend for a period of years, and some conditions, such as HD, may include the opportunity to predict the future likelihood of disease prior to the onset of symptoms in the preclinical phase of the disorder. For families of persons with illnesses that span many years, family issues will evolve during the time that the person knows that he or she has or will have the disorder. Family members who are responsible for disease management will also move through multiple family phases, for example, from being families with young children, to families with adolescents, to families launching children, and to families in later life (Carter & McGoldrick, 2005). The nature of concerns and the selection or availability of strategies to resolve these concerns are likely to vary not only across the progression of the disease but also across issues reflecting the family life cycle.

This survey allows clinicians and researchers to identify the frequency of concerns as well as the intensity of concerns for individual family members. In a clinical setting, the Personal scale of the HDFCSS can be used as a screening tool to identify components of family member well-being that may compromise the abilities of the family to manage or provide care for the person with HD as well as for themselves and the family. The Strategies scale can be used to identify effective and ineffective efforts to manage family member concerns. For example, this scale can be used to identify constructive or health-promoting strategies to manage stressful situations for the family member. Identifying management strategies will not only assist the health care provider in providing resources but also assist the family member in assessing his or her own strengths and limitations in protecting one's health as well as the health of the person with HD. The scale may identify topics that the family member had not previously considered or may help normalize feelings or behaviors that were previously thought to be inadequate or abnormal.

Increasing the sample size will allow further psychometric testing, including factor analysis of the larger scales within the HDFCSS. Construct validity testing is also needed to identify how family members' scores on this measure compare to those on established measures for

specific constructs, such as caregiver burden or anxiety, that may pertain to these families. The survey may be useful in populations outside the United States, particularly in countries where health resources and systems differ from those in the United States.

In summary, genetic aspects of neurodegenerative disorders continue to be identified in some families, which allows for detection of gene mutations prior to the diagnosis of clinical symptoms. Recently developed theoretical frameworks guide research and clinical practice for families who manage these chronic conditions. Valid and reliable measures are needed to address family members' concerns and to identify how families manage neurodegenerative conditions with genetic risk. The HDFCSS has the potential to identify concerns and strategies of family members who manage multiple challenges of HD.

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TABLE 1Focus Group Sample ($N=91$)

Stage of HD in person with HD	Number (%)	Prodromal (at home)
		19 (21%)
Symptomatic (at home)	42 (46%)	
Long-term care facility	30 (33%)	Participant age
Mean	50 years	
Range	25–79 years	
Gender		
Female	67 (74%)	
Male	24 (26%)	
Ethnicity		
White	87 (96%)	
Other	4 (4%)	
Relationship to person with HD		
Spouse	59 (65%)	
Other	32 (35%)	
Location and participation		
United States: four sites	68 (75%)	
Canada: two sites	23 (25%)	

TABLE 2

HDFCSS Sample Items and Format

Personal Concerns Scale		
Item	Frequency	Reaction
It is difficult to distinguish HD from the person	0 1 2 3 4 ?	0 1 2 3 4 ?
I worry that my children will develop HD	0 1 2 3 4 ?	0 1 2 3 4 ?
Person with HD Scale		
Item	Frequency	Reaction
Uses poor judgment	0 1 2 3 4 ?	0 1 2 3 4 ?
Has difficulty walking	0 1 2 3 4 ?	0 1 2 3 4 ?
Community Health Care Services Scale		
Item	Disagree/Agree	Reaction
I am concerned he/she is not getting the health care he/she needs	SD D A SA ?	0 1 2 3 4 ?
It is difficult to find appropriate nursing home, assisted living or adult daycare that would accept him/her	SD D A SA ?	0 1 2 3 4 ?
Strategies to Manage Concerns Scale		
Item	Don't Use	How Helpful
I avoid being around him/her	0	1 2 3 4
I instruct our children how to deal with him/her	0	1 2 3 4

Note. Frequency: 0 = never occurred at any time; 1 = not in the past month; 2 = just a few times (1–3 times in the past month); 3 = about once or twice a week in the past month; 4 = several times a week, daily, or more in the past month; ? = don't know/not applicable. Reaction: 0 = not at all; 1 = a little; 2 = moderately; 3 = very much; 4 = extremely; ? = don't know/not applicable. Disagree/Agree: SD = strongly disagree; D = disagree; A = agree; SA = strongly agree. How Helpful: 0 = I don't do this; 1 = do this but it is not helpful; 2 = do this and it is somewhat helpful; 3 = do this and it is very helpful; 4 = not applicable.

TABLE 3

Validity Indices

	Clarity		Relevance	
	Average	Range	Average	Range
Personal concerns of caregiver	72%	33%–100%	77%	50%–100%
Caregiver concerns for persons with HD (includes Community Health Care services)	92%	71%–100%	93%	33%–100%
Strategies	90%	43%–100%	92%	62%–100%

TABLE 4

Correlations of Scales

Scales	Personal Concerns (Reaction)	Concerns Persons With HD (Frequency)	Concerns Persons with HD (Reaction)	Community Health Service (Agreement)	Community Health Service (Reaction)	Strategies Usefulness
Personal Concerns (Frequency)	.87	.57	.57	.20	.30	-.30
Personal Concerns (Reaction)		.50	.64	.19	.33	-.30
Concerns Persons With HD (Frequency)			.82	.14	.23	-.20
Concerns Persons With HD (Reaction)				.15	.37	-.24
Community Health Service (Agreement)					.74	-.08
Community Health Service (Reaction)						.00

TABLE 5

Internal Consistency Estimates and Confidence Intervals (CI) of the Scales

Scales	N	Number of Items	Alpha	0.95 CI Lower Limit	0.95 CI Upper Limit	Minimum Alpha if an Item Deleted	Maximum Alpha if an Item Deleted	Mean Item-Total Correlation Range
Personal Concerns (Frequency)	245	31	.91	.89	.92	.90	.91	.46 .09–.67
Personal Concerns (Reaction)	245	31	.91	.90	.93	.91	.92	.48 .08–.69
Concerns Persons With HD (Frequency)	245	36	.94	.93	.95	.94	.95	.54 .27–.80
Concerns Persons With HD (Reaction)	245	36	.94	.93	.95	.94	.94	.523 .17–.75
Community Health Service (Agreement)	245	14	.83	.80	.86	.81	.84	.47 .22–.66
Community Health Service (Reaction)	245	14	.89	.87	.91	.88	.89	.58 .42–.72
Strategies Usefulness	245	82	.94	.93	.95	.94	.94	.37 –.06–.58