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Development of the HD-Teen Inventory

Martha Driessnack[1], **Janet K. Williams**[1], **J. Jackson Barnette**[2], **Kathleen J. Sparbel**[3], and **Jane S. Paulsen**[1]

[1]The University of Iowa, Iowa City, IA, USA

[2]University of Colorado–Denver, Denver, CO, USA

[3]University of Illinois–Chicago, Chicago, IL, USA

Abstract

Adolescents, who have a parent with Huntington Disease (HD), not only are at genetic risk for HD but also are witness to its onset and devastating clinical progression as their parent declines. To date, no mechanism has been developed to direct health care providers to the atypical adolescent experiences of these teens. The purpose of this report is to describe the process of developing the HD-Teen Inventory clinical assessment tool. Forty-eight teens and young adults from 19 U.S. states participated in the evaluation of the HD-Teen Inventory tool. Following item analysis, the number of items was reduced and item frequency and reaction scales were combined, based on the strong correlation ($r = .94$). The resultant tool contains 15 inventory and 2 open-ended response items. The HD-Teen Inventory emerged as a more compact and efficient tool for identifying the most salient concerns of at-risk teens in HD families in research and/or clinical practice.

Keywords

Huntington disease; adolescence; HD-Teen Inventory; field test

Introduction

Adolescence is commonly recognized as the developmental period during which an individual transitions between being a child and becoming an adult (Holmbeck, 2002; Smetana, Campione-Barr, & Metzger, 2006). Current research suggests that for this transition to be successful, teens need ongoing and secure parental attachment (Hair, Moore, Garrett, Ling, & Cleveland, 2008). Thus, health providers caring for teens must be aware of issues affecting both teens and their parents since both can affect the teen's risk for transition difficulties.

One group of adolescents that needs focused attention includes teens with parents who have Huntington Disease (HD). Research reveals these teens can have atypical, complex, and often painful adolescent and family experiences, which increases their risk for a difficult transition through adolescence (Duncan et al., 2007, 2008; Keenan, Miedaybrodzka, van Teigjlingen, McKee, & Simpson, 2007; Keenan, van Teijlingen, McKee, Miedzybrodzka, & Simpson, 2009; Sparbel et al., 2008; Williams, Ayres, Specht, Sparbel, & Klimek, 2009a). Nevertheless, assessing this group of teens and clinical follow-up is particularly challenging,

Corresponding Author: Martha Driessnack, PhD, PNP-BC, RN, College of Nursing, The University of Iowa, 324 NB, Newton Avenue, Iowa City, IA, USA, martha-driessnack@uiowa.edu.

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because HD families often carefully conceal their situation, making it difficult to be aware of potential problems (Quaid et al., 2008).

Background

Huntington disease (HD) is an inherited, neuropsychiatric disease involving progressive, involuntary and voluntary motor disability, a decline in cognitive capability, and psychiatric disturbances (Sturrock & Leavitt, 2010; Warby, Graham, & Hayden, 2010). The mean onset of clinical symptoms is 30 to 50 years of age; and after onset, the median survival time is 17 to 20 years (Roos, 2010). The average age at death for an adult with HD is 55 years of age.

Adolescents who have a parent with HD typically witness the onset of their parent's symptoms, the disease progression, and their parent's decline and eventual death. To date, there is no cure or life-extending intervention for the individual with HD (Richards, 2008). While having a parent with a life-altering and limiting disease is difficult for any teen, symptom progression in the HD parent can be especially disruptive to the process of secure parental attachment, often placing the teen in the role of caregiver rather than receiver.

Affected individuals may present with neurologic psychiatric or cognitive changes. Chorea, an involuntary movement disorder involving irregular jerking of limbs, face, or trunk, is present in more than 90% of individuals (Sturrock & Leavitt, 2010). The choreic movements are continuously present during waking hours, leading to increased caloric needs that are difficult to keep up with. In the early stages of HD, clinical manifestations include subtle changes in coordination, minor involuntary movements, and difficulty in mental planning, irritability, and depression (Cummins, Eggert, Pruitt, & Collins, 2011). In the middle stage, chorea becomes more prominent, voluntary activity becomes increasingly difficult, and dysphagia emerges, along with intermittent outbursts of aggressive behaviors and social disinhibition. In the late stages of HD, motor disability becomes severe and the individual is totally dependent.

In addition, because HD is inherited in an autosomal dominant manner, teens in HD families face an additional challenge as they discover their own genetic risk for the disease (Sturrock & Leavitt, 2010). The inheritance of HD follows an autosomal dominant pattern, meaning each offspring of an individual with HD has a 50%, or 50-50, chance of inheriting the disease. Although genetic testing for HD is available to "at risk" individuals, it is not offered to teens until the individual reaches 18 years of age, the age at which they can give their own informed consent.

Although these teens' genetic risk to develop HD is already fixed, their developmental and health trajectories are not. Accordingly, through qualitative interviews and focus groups, their experiences have been explored in-depth, highlighting the need to identify teens who can be facing difficult challenges and provide support (Duncan et al., 2007, 2008; Keenan et al., 2007, 2009; Sparbel et al., 2008; Williams et al., 2009a). Nevertheless, no mechanism has yet been developed that allows clinicians or researchers to readily access these teen experiences in the clinical setting. To address this shortfall, we developed, field-tested, and revised, a novel, clinical inventory tool—the HD-Teen Inventory (HD-TI). Here, we report the development of the HD-TI and findings from the field test.

Method

This section begins with a brief overview of the process to develop items for the HD-TI, followed by a discussion of the field test and subsequent revision of the HD-TI as it was transitioned to its current format as a clinical tool.

Development of the HD-TI

The development of the HD-TI began within a larger study focusing on the needs of family members of individuals with HD linked with the PREDICT-HD study (Paulsen et al., 2006; Williams, 2001). In the first phase of the larger study, data from focus groups were used to develop items. The focus group data and analysis with teens (Sparbel et al., 2008; Williams et al., 2009a) and adult family members (Williams & Ayres, 2007; Williams et al., 2007, 2009b) are reported elsewhere (Skirton, Williams, Barnette, & Paulsen, 2010; Williams, Skirton, Barnette, & Paulsen, 2012). Here, we focus on the final stage of evaluation and the transition of the HD-TI to its final format (Figure 1).

Field Test

The HD-TI items were identified from the first phase qualitative analysis of data from six focus groups that involved 32 teens at five HD Centers across the United States and Canada (Sparbel et al., 2008; Williams et al., 2009a). To assure the initial items were clear and relevant; we validated the item content using nine experts in HD, family and/or pediatric clinical management, or research, as well as 9 teens who participated in the initial focus groups. The resultant HD-TI contained 29 *Personal Concerns* items and two open-ended questions. The openended questions asked respondents to identify (a) the top-ranked concern from the items listed and (b) any item that was missing that we needed to know about.

The next step was to field test the HD-TI in a new sample to identify potential sources of problems in terms of item concepts, response format, and/or admin- istration (Groves et al., 2009). For this study, the research team was particularly interested in fine-tuning the HD-TI, to make it user-friendly and ensure it cap- tured only the aspects of the teens' experiences that are unique in HD families by carving these aspects out from the larger experience of adolescence. Of particular concern was the inclusion of teen preferences. This concern was rooted in the evidence-based practice movement's emphasis on the application of best research evidence to clinical care *in conjunction with* the preferences and wishes of the patients to which the evidence is being applied. Clinical assessment tools developed using this approach are more likely to direct clinician's attention to those items most important to patients, which in this case were teens in HD families (Gilgum, 2004; Morse et al., 1998).

The study was approved by the University of Iowa Institutional Review Board (IRB). The sampling goal was to obtain equal distribution of teens (14 to 17 years of age) and young adults (18 to 30 years of age). The two groups were purposively sought not only to ascertain what concerns are unique to teens currently living in families with HD but also to identify concerns that persist over time in the memory of young adults recalling their teen years. Eligible participants (a) had a family member with HD, (b) could communicate in English, and (c) were 14 to 30 years of age. The study was announced through the University of Iowa HD Registry and Huntington Disease Society of America (HDSA). Participants received their choice of either a phone card or an *iTunes* gift card for participating.

Forty-eight teens and young adults from 19 different U.S. states participated. Of the 48 participants, 66.7% were female and 91.7% were White. The mean age was 18.8 years (SD = 4.41). Twenty three (47.9%) were teens (14 to 17 years old) and 21 (43.8%) were young adults (18 to 30 years old).

Findings

In the initial version of the HD-TI, each respondent was asked to indicate two things: (a) the frequency with which each of the 29 items occurred and (b) their personal reaction. For example, one item was *I worry I will develop HD*. Respondents first indicated how

frequently this concern occurred in the past month (0 = *never occurred at any time*, to 4 = *several times a week, daily, or more*). They were then asked to share their personal *reaction*, in terms of how much this concern bothers them (0 = *not at all*, to 4 = *extremely*). For each item, an overall index score was also calculated to represent the product of these two ratings (*frequency* × *reaction*). Internal consistency measures and 95% confidence intervals were computed using the ScoreRel CI program (Barnette, 2005). All values were well above common standards used to assess internal consistency: (a) frequency (α = .96; CI [.94, .97]), (b) personal reaction (α = .96; CI [.95, .98]), and (c) overall index score (α = .96; CI [.94, .97]).

Items were ranked *by statistical* mean (*M*). The top-ranked item was *I worry I will develop HD* (*M* = 2.50), with 76% reporting this concern occurred any- where from once a month to several times a day. The second ranked item, *It is difficult to distinguish HD from the person* (*M* = 2.36), followed close behind, with more than 66% of respondents reporting this concern also occurred any- where from once a month to several times a day. Items identified by intensity/reaction scores were also ranked by statistical mean. Of note, was the extremely strong correlation between item frequency means and the reaction/intensity means (*r* = .94). Also of note was a clear delineation of item frequency between the top 14 and remaining 15 items.

Items were further analyzed by examining differences between the teens' responses and the experiences young adults recalled from their teen years. Using Fisher Exact tests, 16 items had significantly ($p < .05$) higher frequencies in the young adults recalling their teen experiences. Fifteen of the 16 items also had significantly higher intensity/reaction scores ($p < .05$). Twelve of the 16 items were already among the 14 top-ranked teen group items, validating their prevalence and importance in teen's lives. Females reported six concerns significantly more often ($p < .05$) than their male counterparts. However, because there was a 2:1 female to male ratio in the numbers of participants, male respondents were examined as a subgroup to ensure their experiences were being not masked by the larger female presence. The team was reassured as the top-ranked male concerns, across both teens and young adults, were found to be identical to the overall ranking of items.

The final evaluative step compared top-ranked items to the two open-ended questions. Of the 48 participants, 31 (62.5%) responded. Three concerns were mentioned most often and were already identified in the top-ranked 14 items: *(a)I worry that I will develop HD* (10 comments), (b) *I feel worried that my quality time with him or her is running out* (6 comments), and (c) *I feel no one knows what I am going through* (6 comments). Seven (22%) of the 48 participants indicated a concern they felt was missing: *having an intimate relationship*.

To understand how concerns about intimacy had not been included in the original items, the research team returned to the transcripts and analysis from the Phase 1 teen focus groups. A close evaluation revealed intimacy was identified as an important teen concern, but the team felt it was a concern common to most teens, not unique to teens in HD families, and therefore had not been included. However, after close rereading and write-in comments during the field test, the teens' concerns about becoming intimate went beyond typical adolescent concerns about sexual and/or intimate relationships as these teens were concerned about becoming intimate because they did not yet know their genetic HD-risk status. Therefore, the decision was made to add this item to the HD-TI, using the participants' written responses to craft it.

Refining the HD-TI

The first step in refining the HD-T was to establish whether the internal consistency of the HD-T would be compromised if, out of the original 29, only the 14 top-ranked items were included. The Cronbach α for the 14 items remained high at .93 (frequency) and .94 (reaction). Second, because of the extremely strong correlation between frequency and reaction scores ($r = .94$), the research team felt that either score could be used. Furthermore, the participants suggested collapsing scaled responses into binomial response categories (yes/no) was more user-friendly. Having binomial categories also ensured male responses, which scaled lower than females, were represented equally. Furthermore, other researchers are beginning to report that teens prefer categorical responses over Likert-type scales (Flicker et al., 2010; Gallo, 2010).

After field testing, the final format that emerged for the HD-TI is a targeted, 15-item categorical response inventory, with two open-ended questions that allow for individuation and continued evolution (Figure 2).

Summary

The research team used a carefully sequenced, evaluative process that placed teens at the forefront at each stage, thus producing a clinical inventory tool that captures the unique aspects of teens in HD families. This is particularly important because while individuals directly affected by HD receive health care from specialists, the adolescents within these families typically receive their health care from primary care providers who may be unaware that these teens have specific risks that can impede their transition through adolescence. The process used in developing the HD-TI may be relevant to other researchers seeking to capture experiences of teens in families with other adult-onset and/ or progressive chronic conditions that could interfere with a teen's ability to maintain secure parental attachment during adolescence.

The HD-TI emerged from field testing as a more compact and efficient tool for assessing the most salient concerns of at-risk teens in HD families. The open-ended questions provided formative comments, which not only validated but also revealed an important omission. The open-ended questions were retained to capture individual experiences that may not be anticipated and to create a mechanism through which the tool can continue to evolve. By adding an item about intimacy, the HD-TI was further enhanced. The final inventory format is easy to administer, can be completed within the time constraints of clinical practice, and provides health care providers and researchers with the ability to identify sources of personal concerns for teens growing up in families with members who have HD. Awareness of these concerns is an essential first step in building supportive and health promoting interventions for this population.

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Biographies

Martha Driessnack, PhD, PNP-BC, RN, is an assistant professor at the College of Nursing, The University of Iowa, Iowa City, IA, USA.

Janet K.Williams, PhD, RN, FAAN, is the Kelting Professor of Nursing at the College of Nursing, The University of Iowa, Iowa City, IA, USA.

J. Jackson Barnette, PhD, is a professor of biostatistics & informatics at the Colorado School of Public Health, University of Colorado–Denver, Denver, CO, USA.

Kathleen J. Sparbel, PhD, FNP-BC, RN, is a clinical assistant professor at the College of Nursing, University of Illinois–Chicago, Chicago, IL, USA.

Jane S. Paulsen, PhD, is professor of psychiatry, neurology, psychology, and neurosciences at the Carver College of Medicine, The University of Iowa, Iowa City, IA, USA.

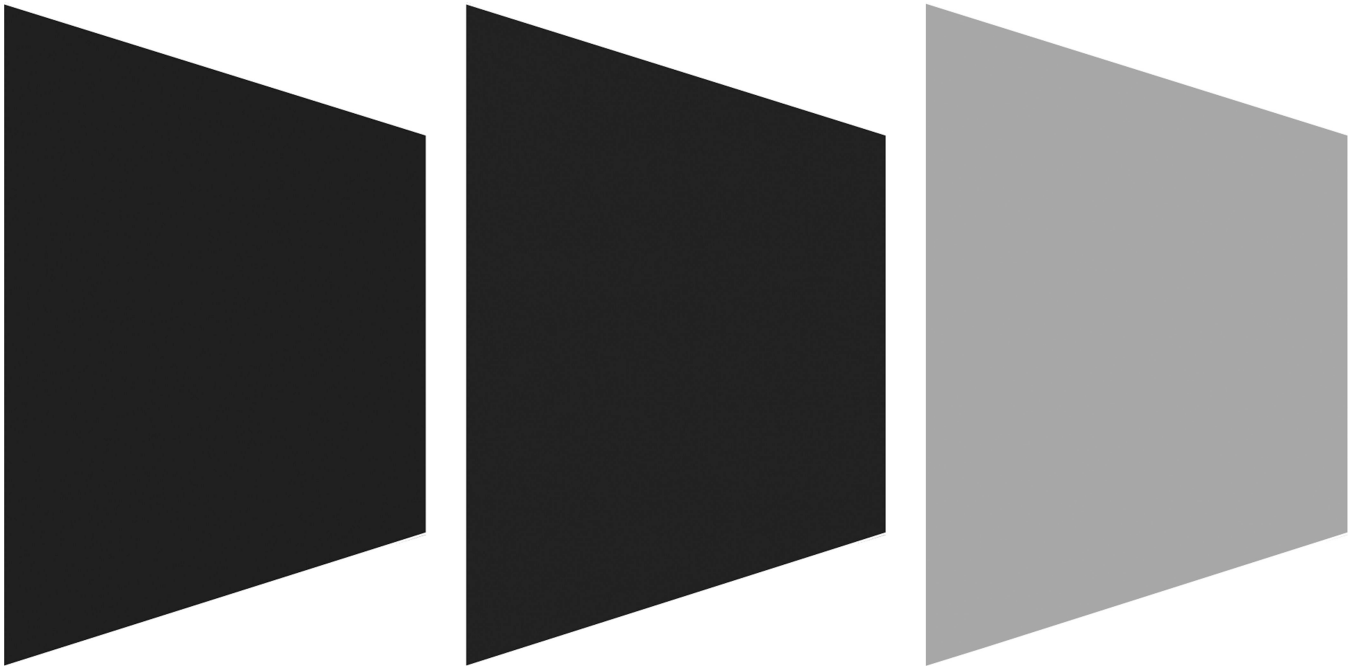

Figure 1.
Sequenced,3-phase evaluative process used in developing the HD-TI

Directions:
There are 15 different types of feelings,worries, and experiences listed below. Check each one that you live with. Next, share any feeling, worry, or experience you have that is *not* on the list. Then, circle the item that is of most concern to you right now.

- o I worry I will develop Huntington Disease
- o I feel no one knows what I am going through
- o I miss the relationship I had with my mother/father
- o I worry about our family's finances
- o I have a hard time distinguishing Huntington Disease from the person o I don't know whether to have genetic testing for Huntington Disease
- o I feel sad in my family
- o I worry that my quality time with my parent is running out
- o My friends have a hard time understanding the stressors in my family
- o I worry about having an intimate relationship with someone because of my risk for Huntington Disease
- o It feels like I need to act like an adult rather than like a teen
- o I worry that my parent may not live to experience important milestones in my life
- o The relationships in my family are strained due to the stresses of Huntington Disease
- o I don't plan to have a family because of the risk of Huntington Disease for my children
- o I worry about handling all of the stresses as my parent's symptoms get worse

Please write in the space below any feeling, worry, or experience you have that is *not* on the list:

Of all of these feelings, worries, and experiences (including the one(s) you wrote in above): CIRCLE the one that is of *most* concern to you right now.

Figure 2.
HD Teen Inventory (HD-TI)