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Nancy R Downing
*Texas A&M University*

Siera Goodnight
*University of Michigan - Ann Arbor*

Sena Chae
*University of Iowa*

Joel S Perlmutter
*Washington University in St. Louis*

Michael McCormack
*Rowan University*

*See next page for additional authors*

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## Authors

Nancy R Downing, Siera Goodnight, Sena Chae, Joel S Perlmutter, Michael McCormack, Elizabeth Hahn, Stacey K Barton, and Noelle Carlozzi

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# Factors Associated with End of Life Planning in Huntington Disease

**Nancy R. Downing**[a], **Siera Goodnight**[b], **Sena Chae**[c], **Joel S. Perlmutter**[d], **Michael McCormack**[e], **Elizabeth Hahn**[f], **Stacey K. Barton**[d], and **Noelle Carlozzi**[b]

[a]Texas A&M College of Nursing, 8447 Riverside Parkway, Bryan, Texas 77807, USA

[b]University of Michigan, Department of Physical Medicine & Rehabilitation, 2800 Plymouth Road, Ann Arbor, MI 48109-2800, USA

[c]University of Iowa College of Nursing, 50 Newton Rd., Iowa City, IA 52242, USA

[d]HDSA Center of Excellence, Washington University at St. Louis School of Medicine, 660 S. Euclid Avenue, St. Louis, MO 63110, USA

[e]Department of Pathology, Rowan University, 401 South Broadway, Camden, NJ 08103, USA

[f]Northwestern University Feinberg School of Medicine, 633 N. St. Clair St., Chicago IL 60611, USA

## Abstract

**Objective.—**Knowledge of one's gene status for adult onset conditions provides opportunity to make advance end-of-life (EOL) plans. The purposes of these analyses were to: 1) determine the prevalence of EOL plans, including advance directives (ADs) among persons across three stages of Huntington disease (HD); and 2) examine factors associated with having ADs in this sample.

**Methods.—**Data are from 503 participants in the Huntington Disease Quality of Life study. Participants completed an online health-related quality of life survey that included questions regarding EOL planning and self-reported HD symptoms. Frequencies were calculated for EOL planning by HD stage. Bivariate analysis and logistic regression were used to identify variables associated with having ADs.

**Results.—**38.2% of participants stated they had ADs and fewer than half had other EOL plans. Being older, increased HD stage, more years of education, lower anxiety, more swallowing symptoms, and higher meaning and purpose were associated with having ADs.

**Conclusion.—**The prevalence of ADs in our sample is comparable to the general US population, but surprisingly low considering the severity and long disease course of HD.

**Practice Implications.—**Health care providers should develop specific interventions early in the disease process to increase ADs in this population.

**Correspondence to:** Nancy R. Downing, Texas A&M College of Nursing, 8447 State Highway 47, Bryan, TX, 77807, USA. Tel: 979-436-0157; Fax: 979-436-0098. downing@tamhsc.edu.

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**Keywords**

Advance directives; Huntington disease; adult onset conditions; palliative care

## Introduction

Huntington disease (HD) is a devastating neurodegenerative disease for which there is palliative treatment, but no cure. The disease is caused by an autosomal dominant trinucleotide CAG repeat expansion on the *HTT* gene. Symptoms typically begin in middle age, and include progressive decline in cognitive, behavioral, motor, and day-to-day functioning [1] that significantly impact health-related quality of life (HRQOL) [2, 3]. Progression to dementia is inexorable; death occurs approximately 17–20 years following motor diagnosis [1]. Presymptomatic genetic testing has been available since the discovery of the *HTT* gene mutation in 1993 [4]. If a patient does not undergo presymptomatic genetic testing, diagnoses are made based on consistent family history with typical motor signs (e.g. chorea) or confirmation by genetic testing [5]. One of the benefits of knowing one's gene status for an adult onset disease, especially one that involves dementia, is the ability to make advance end of life (EOL) decisions while one still has the cognitive capacity [6].

Advance directives (ADs), including living wills, health care powers of attorney, or health care directives, are legal documents that state an individual's preferences for medical treatment and to appoint a surrogate to make decisions on their behalf if they become incapacitated [7]. Advance directives are effective in decreasing unwanted hospitalization and unwanted life-sustaining treatment, and increasing the use of palliative and hospice care [8]. Since HD affects decision-making capacity in later stages, it is imperative that EOL discussions occur early in the disease course [9]. A working group on EOL planning needs for people with HD funded by the Robert Wood Johnson Foundation recommended healthcare providers educate persons with HD on the need to develop and complete ADs [10].

Despite a recognized need, the prevalence of ADs among persons with HD in the US is not known. People usually have prior knowledge of their HD risk because they have affected family members or have undergone HD genetic testing. Knowledge of gene status for adult onset conditions provides the opportunity to make advance EOL plans [6]. People with HD might be more likely than the general population to have ADs since people with chronic diseases in the general US population are more likely to have ADs than those without chronic disease [11]. The purposes of these analyses were to: 1) examine the prevalence of ADs and other EOL plans in a large sample of people at three stages of HD; and 2) examine the demographic, clinical and self-reported HRQOL factors associated with having ADs in this sample.

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## Methods

### Setting/Sample

Data for this analysis come from the first phase of the Huntington Disease Quality of Life (HDQLIFE) Study. The purpose of the HDQLIFE study is to develop computer adaptive tests (CATs) and short forms (SF) to assess patient-reported HRQOL for persons with HD [12–15]. Individuals with prodromal or manifest HD ($N$=503) 18 years and older and able to read and understand English participated in the first phase of the study (12/19/2012–12/14/2014). Participants were recruited at a variety of specialized treatment centers across the US (University of Michigan, University of Iowa, University of California - Los Angeles, Indiana University, Johns Hopkins University, Rutgers University, Struthers Parkinson's Center, Washington University at St. Louis, University of California – San Francisco, and the Cleveland Clinic), from the PREDICT-HD study [16], the National Research Roster for Huntington's Disease [17], and articles and advertisements in HD newsletters and websites. This study was approved by all participating institutions' human subjects research boards and all participants provided informed consent.

There were 505 participants in the HDQLIFE study. Data regarding HD stage was missing for two participants. Thus, analyses that included comparisons between HD stage included 503 participants stratified into three HD stages: prodromal ($n$=197), early ($n$=193), and late ($n$=113). Prodromal stage included individuals who did not have a clinical diagnosis of HD but had a confirmed CAG repeat ≥36, meaning they will eventually develop HD if they have a normal lifespan; manifest HD participants ($n$=308) had a neurologist-confirmed HD clinical diagnosis or received a ≥99% confidence rating on the motor component of the Unified Huntington's Disease Rating Scale (UHDRS), indicating unequivocal signs of HD. Staging of HD for participants with manifest HD was classified according to the UHDRS Total Functional Capacity (TFC) scale [18], a clinician-administered scale evaluating day-to-day function regarding occupation, finances, domestic chores, activities of daily living, and care level. Scores range from 0 (low functioning) to 13 (high functioning). Participants were classified as early stage (7–13) or late stage HD (0–6).

### Assessments

Participants completed the HDQLIFE protocol, including questions regarding EOL planning, including ADs, living wills, healthcare power of attorney, resuscitation preference, conversations about death and dying, location of death preference, nursing home care, palliative care, and hospice care. All participants were asked to answer questions about all these items. Due to confusion regarding the definitions of these terms in the general public that might be relevant in this study, the focus of our analysis was on comparison of participants with and without ADs. Information regarding ADs was assessed by a single item, "Advance directive (also known as a Living will)." Participants were instructed to choose the response that best described where they stood regarding ADs: "I have not thought about getting an advance directive;" "I have thought about getting an advance directive;" "I have taken steps to obtain an advance directive;" and "I have an advance directive." Similar response choices were provided for other items.

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Participants also completed newly developed self-report assessments for the presence of HD symptoms, concerns about HD, impact of symptoms, and concerns on HRQOL. We used SF versions of these measures that were created using item response analyses: HDQLIFE Speech Difficulties SF [15], HDQLIFE Swallowing Difficulties SF [15], HDQLIFE Chorea SF [13], HDQLIFE Concern with Death & Dying SF, and HDQLIFE Meaning & Purpose SF [12]. All HDQLIFE measures generate *T* scores on a *T*-metric relative to other individuals with HD. *T*-scores are standardized scores with mean of 50 and standard deviation of 10; higher scores indicate more of the construct being measured (e.g., more speech difficulty, or greater meaning and purpose). Participants also completed the Patient-Reported Outcomes Measurement Information System (PROMIS) Anxiety SF [19]; higher scores indicate more anxiety and is scored on a *T*-metric with the general population as referent.

Trained clinicians administered the UHDRS Total Motor Score (TMS) in person. Participants completed patient-reported outcome (PRO) measures through PROMIS Assessment Center (https://www.assessmentcenter.net) at a designated computer or tablet during the study visit or were asked to complete it within two weeks of the study visit. Participants and those assisting them were instructed that responses were to come directly from participants (i.e., assistance was limited to logging into the online study platform, reading questions aloud, and/or clicking response options, when needed).

## Data Analysis

Participants were classified as "with ADs" when participants selected the response option "I have an advance directive;" all other responses to that question were classified as "without ADs." We calculated descriptive statistics for demographic variables (age, gender, race, ethnicity, marital status, years of education), and scores on HDQLIFE and PROMIS measures. Analyses of variance (ANOVA) were used to compare demographic differences between prodromal, early, and late stages. Sociodemographic and clinical characteristics of participants with and without ADs were compared using chi-square for categorical data: gender, race, marital status, and HD stage; independent *t*-tests were conducted to compare continuous data: age, years of education, and self-report SF (HDQLIFE Chorea, HDQLIFE Speech, HDQLIFE Swallowing, HDQLIFE Concern with Death & Dying, HDQLIFE Meaning & Purpose, and PROMIS Anxiety). We used an alpha of .05 for all *t*-tests. We used Bonferroni adjusted alpha levels for the variables groups demographics (.05/5 variables = .01) and HDQLIFE/PROMIS measures (.05/6 variables = .008). We did not correct for all variables together (.05/12 = .004) because this is the first known analysis on this topic and we did not want to miss potentially important differences that could be further examined in future studies.

In the logistic regression model, variables that were significantly different ($p < 0.05$) between participants with and without ADs in chi-square and *t*-tests were included to determine which factors independently increased odds of having ADs: age, HD stage, education, HDQLIFE Concern for Death & Dying, HDQLIFE Meaning & Purpose, and PROMIS Anxiety. Age and years of education were entered as continuous variables and *T* scores were used for HDQLIFE and PROMIS Anxiety. HDQLIFE Swallowing was not

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included because the difference between those with ADs (53.83) and without ADs (52.78) was unlikely to be clinically meaningful. Although not significant in *t*-tests, HDQLIFE Concern for Death & Dying was included in the model because this psychological variable might impact whether a person has ADs.

## Results

Participant demographics are shown in Table 1. The age range of participants was 18–81, mean 49.04 years. Most participants were White (96%) and not Hispanic (93.7%). There were some notable differences in ANOVA comparisons between prodromal vs. early and prodromal vs. late stage groups on some variables: There were differences in age, $F(2, 500) = 44.377$, $p < .001$, and years of education, $F(2,498) = 15.415$, $p < .001$, with participants in the prodromal stage being younger and having more years of education than those in the early and those in the late stage. More prodromal stage participants were married compared to early stage, $X^2 (2, 502) = 8.12$, $p = .017$.

Overall, 38.2% of participants reported they had ADs. Frequencies and percentages for ADs and associated EOL planning are presented in Table 2 for each HD stage. In general, higher percentages of participants in the late stage had ADs and other EOL plans.

Chi-square results (Table 3) indicated significant differences between participants with and without ADs for HD stage, $X^2 (2, n=503) = 9.01$. Participants in late stage HD were more likely to have ADs than those in prodromal or early stage HD. In *t*-test analyses (Table 3), there were significant differences between participants with and without AD based on age, $t(503) = 8.63$, $p = <0.001$; years of education, $t(501) = -2.89$, $p=0.004$; and HDQLIFE Meaning & Purpose, $t(493) = 2.92$, $p =0.004$. Participants were more likely to have ADs if they were older, had more years of education, and higher scores on HDQLIFE Meaning and Purpose and HDQLIFE Swallowing, and lower scores on PROMIS Anxiety, although Swallowing and Anxiety were not significant after correcting for multiple comparisons.

The multivariable logistic regression model explained 72.4% of the variance in predicting whether a participant had ADs (Table 4). For each year of age, participants had 6% higher odds of having ADs (odds ratio [OR] 1.06; 95% CI: 1.03, 1.08). Participants in the late stage had almost three times higher odds of having ADs than the prodromal stage (OR 2.87, 95% CI: 1.35, 6.14). Each year of education increased the odds of having ADs by 12% (OR 1.12, 95% CI: 1.02, 1.22). Early stage HD, HDQLIFE Concern for Death & Dying, HDQLIFE Meaning & Purpose, and PROMIS Anxiety were not associated with higher odds of having ADs.

## Discussion

Huntington disease is a devastating disease that causes dementia and premature death. People usually have many years to engage in EOL planning since the disease course spans up to 20 years. We report data on the prevalence of ADs among a large sample of people with HD in the US. Among the 503 participants in this analysis, 38.2% responded they had ADs. This exceeds 31.3% of participants in a study of persons with HD in the Netherlands [9], and 26.3% of people in the general US population [11]; however, it is lower than 47.3%

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of participants over 60 in the US with two or more chronic conditions [20]. Considering the severity of HD and its association with premature mortality, it is surprising only 38.2% of participants had ADs.

Older age has consistently been associated with having ADs in prior studies, as it was in ours [9, 11, 20]. Our sample is relatively young (mean age = 49.04). When age is considered, prevalence of ADs may be higher than among persons of similar ages without HD. For example, in a study examining prevalence of ADs in the general population [11], 18.2% of participants ages 18–54 had ADs. In that light, 38.2% of participants in our study with ADs appears relatively high. On the other hand, it is reasonable to assume that people who know they face significant premature morbidity and death would have even higher prevalence of ADs. Self-reported HD symptoms, including the hallmark symptom chorea, were not associated with increased ADs in our study. Having more self-reported swallowing symptoms was associated with having ADs in bivariate analyses, but the difference in symptoms between those with and without ADs was very small and was significant after correcting for multiple comparisons. There are some previously documented phenomena in HD that might explain why prevalence of ADs was not as high as expected. For example, decreased awareness of disease symptoms can occur in HD [21], and a tendency for persons with HD to normalize symptoms [22]. While participants in late stage HD were almost three times more likely than participants in the prodromal stage to have ADs, this could be related more to age than presence of HD symptoms. Our findings suggest that persons with the HD gene mutation are similar to those without HD with regard to EOL planning, despite knowing more about their potential fate. The HDQLIFE study was not designed specifically to examine this issue, however; thus, it is possible that other designs might better reveal the motivations of persons with HD to make EOL plans.

Participants with late stage HD in our study might have being diagnosed based on family history and HD symptoms and not have had presymptomatic testing, either by choice or because presymptomatic testing was not available before they developed symptoms. People who get tested presymptomatically might be different from people who learn their gene status through clinical diagnosis. In our study, participants in the prodromal stage had more years of education than those in early and late stage HD, and more years of education slightly increased the odds of having ADs. Higher education level has previously been associated with having EOL plans in both HD and non HD samples [9, 11, 23–25]. The relationship between education and having ADs might be mediated by literacy [23]. The use of jargon, including multiple terms referring to ADs, might not clearly describe the intent and usefulness of ADs to patients with lower literacy.

Participants in our study with higher self-reported scores on the PROMIS Anxiety SF were less likely to have ADs in the bivariate analysis, although it was not significant after correcting for multiple comparisons and it was not a predictor of having ADs in the logistic regression model. In our study, HDLIFE Concern for Death & Dying was not significantly associated with having ADs. Fear of death, anxiety and concern might, in fact, lead people to avoid EOL planning in an effort to avoid increasing anxiety [26]. Therefore, it would still be important to assess for anxiety, especially disease- and death-related anxiety, in future studies.

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A potential benefit of presymptomatic HD genetic testing is the ability to plan for the future [27, 28]. Participants in the prodromal stage in our study had undergone presymptomatic HD genetic testing, yet had the lowest prevalence of ADs and other EOL plans in our sample. Thus, genetic testing did not appear to strongly impact making EOL planning in the prodromal stage. However, we cannot exclude that many of these participants will make EOL plans while still cognitively able to do so.

Our study has some limitations that might impact generalizability of our findings. Race and ethnicity are limitations because the vast majority of persons with HD in the US are of European descent and non-Hispanic; prevalence rates are higher in North America, Europe and Australia, compared to Asia [34]. Advance directives, living will, and other EOL terminology such as palliative care were not defined in the HDQLIFE online survey. Given that literacy is an identified barrier to ADs [23], participants might have responded without having a clear understanding of these terms. Participants were encouraged to answer all questions but were not forced to answer items. We did not include a response option to indicate when participants did not understand a question. In the future, we could consider defining these terms.

Our sample included more participants in prodromal and early stages than late stage. This is understandable, as decreased cognitive ability in later stages of HD could preclude capacity to provide informed consent. Participants in the prodromal stage in our study might not be representative of most people at risk for HD because they had independently undergone presymptomatic HD testing while the uptake of presymptomatic genetic testing for HD is estimated to be under 20% [35]. Many participants were recruited from sites that have HD Centers of Excellence (COE), which are affiliated with the Huntington Disease Society of America (HDSA). The COE provide specialized care for people with HD and the HDSA has published materials on ADs available to members. Therefore, our sample might include people more likely to seek and receive information regarding HD. While prevalence of ADs was lower than expected, our participants might be more likely to have ADs than people who have not undergone genetic testing, do not have access to a COE, or do not participate in HD-related research.

We cannot exclude the possibility that family members answered items on behalf of participants, despite instructions that answers should come directly from participants. The purpose of the HDLIFE study is to develop a HRQOL measure for people with HD; the examination of factors associated with having ADs was not a primary aim. Therefore, there are several variables associated with having ADs that we did not measure that have been found to be associated with ADs, including knowledge of ADs, literacy, self-efficacy, religiosity, and illness-related fear [20, 23, 26, 36–38].

Despite the 2004 recommendations of the Huntington’s Disease Peer Workgroup, our findings suggest that prevalence of ADs is low in this population. Interventions designed to increase patient-provider discussions of ADs are effective in increasing development of and use of ADs [29]. An important question remains whether health care providers provide adequate patient education to persons with HD regarding ADs, and at an appropriate level of literacy. Having ADs is often not considered an important issue in HD until the disease

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symptoms have progressed significantly and the patient and family are considering long-term care placement. Unfortunately, this can result in the patient being unable to voice their preferences due to advancing dementia [30].

There is a recognized need for EOL planning for patients with HD to reduce risk of emotional distress from unwanted, aggressive and costly EOL care [10, 31]. While we examined factors associated with having ADs among persons with HD, future studies should identify patient and health care provider barriers to creation of ADs in this population. Data from our study could inform development of specific interventions to increase prevalence of ADs and EOL planning. A promising method involves using vignettes to illustrate disease-specific EOL scenarios to help patients work through their preferences [32]. This could be adapted for use in HD by providing patients with scenarios based on common causes of death in persons with HD. This technique could aid literacy by explaining often misunderstood medical terms and end-of-life interventions such as cardiopulmonary resuscitation, feeding tubes, and ventilators [33]. Medicare reimbursement for EOL discussions is a positive step, although these discussions should begin long before patients with HD become Medicare eligible. Prevalence of ADs and EOL plans is surprisingly low considering the severity and long disease course of HD. Health care providers should actively educate and assist patients with HD in making ADs and EOL plans to specify treatment preferences while they still have the cognitive ability.

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## Acronyms

| | |
|---|---|
| **AD** | Advance directives |
| **CAT** | Computer adaptive tests |

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| | |
|---|---|
| **COE** | Centers of Excellence |
| **EOL** | End of life |
| **HD** | Huntington disease |
| **HDQLIFE** | Huntington Disease Quality of Life study |
| **HRQOL** | Health-related quality of life |
| **PRO** | Patient-reported outcome |
| **PROMIS** | Patient-Reported Outcomes Measurement Information System |
| **SF** | Short form |
| **TFC** | Total Functional Capacity scale |
| **TMS** | Total motor score |
| **UHDRS** | United Huntington Disease Rating Scale |

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**Table 1.**

Participant demographics by HD stage

| | | Stage | | | |
|---|---|---|---|---|---|
| **Variable** | | **Prodromal (*n*=197)** | **Early (*n*=193)** | **Late (*n*=113)** | **All (*n*=503)** |
| Age | | 42.80 | 51.98[a] | 54.95[a] | 49.04 |
| Mean (SD) | | (12.17) | (12.41) | (12.04) | (13.23) |
| Years of Education | | 15.91 | 14.69[a] | 14.24[a] | 15.07 |
| Means (SD) | | (2.94) | (2.78) | (2.64) | (2.89) |
| Gender (%) | Female | 64.0 | 45.1 | 41.6 | 59.2 |
| | Male | 36.0 | 54.9 | 58.4 | 40.8 |
| Race (%) | White | 97.5 | 96.4 | 92.9 | 96.0 |
| | Other | 2.0 | 3.6 | 7.1 | 3.8 |
| | Unknown | 0.5 | 0.0 | 0.0 | 0.2 |
| Ethnicity (%) | Not Hispanic or Latino | 92.4 | 92.7 | 94.3 | 93.7 |
| | Hispanic or Latino | 1.5 | 4.1 | 0.9 | 2.4 |
| | Not Provided | 6.1 | 3.1 | 1.8 | 4.0 |
| Marital Status (%) | Married | 67.0 | 52.8[a] | 62.8 | 60.8 |

[a] = significantly different than prodromal, all $p<0.05$

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**Table 2.**

End of life planning by HD stage

| | HD Stage Frequency Number (%) | | | |
|---|---|---|---|---|
| | **Prodromal** | **Early Stage** | **Late Stage** | **Total** |
| **Advance directive** | | | | |
| I have not thought about getting an advance directive | 35 (17.8) | 28 (14.5) | 14 (12.4) | 77 (15.3) |
| I have thought about getting an advance directive | 83 (42.1) | 75 (38.9) | 22 (19.5) | 182 (36) |
| I have taken steps to obtain an advance directive | 20 (10.2) | 18 (9.3) | 15 (13.3) | 53 (10.5) |
| I have an advance directive | 59 (29.9) | 72 (37.3) | 62 (54.9) | 193 (38.2) |
| Total | 197 | 193 | 113 | 503 |
| **Living will** | | | | |
| I have not thought about preparing a living will | 30 (15.3) | 29 (15.2) | 18 (16.1) | 78 (15.6) |
| I have thought about preparing a living will | 90 (45.9) | 77 (40.3) | 22 (19.6) | 190 (37.9) |
| I have taken steps to prepare a living will | 18 (9.2) | 14 (7.3) | 9 (8.0) | 41 (8.2) |
| I have a living will | 58 (29.6) | 71 (37.2) | 63 (56.3) | 192 (38.3) |
| Total | 196 | 193 | 112 | 501 |
| **Healthcare power of attorney (POA)** | | | | |
| I have not thought of getting a healthcare POA | 55 (27.9) | 32 (16.6) | 14 (12.4) | 102 (20.2) |
| I have thought of getting a healthcare POA | 71 (36.0) | 64 (33.2) | 16 (14.2) | 152 (30.1) |
| I have taken steps to identify a healthcare POA | 21 (10.7) | 19 (9.8) | 11 (9.7) | 51 (10.1) |
| I have a healthcare POA | 47 (23.9) | 70 (36.3) | 69 (61.1) | 186 (36.8) |
| Total | 194 | 185 | 113 | 492 |
| **Resuscitation preference** | | | | |
| I have not thought about my preference for resuscitation if I stop breathing | 55 (28.1) | 49 (25.8) | 26 (23.2) | 131 (26.2) |
| I have thought about my preference for resuscitation if I stop breathing | 66 (33.7) | 60 (31.6) | 27 (24.1) | 154 (30.8) |
| I have made my preference for resuscitation clear to others if I stop breathing | 75 (38.3) | 81 (42.6) | 59 (52.7) | 215(43) |
| Total | 196 | 190 | 112 | 500 |
| **Conversations about death and dying** | | | | |
| I have not thought about starting a conversation about death with my friends, family, or members in the community (e.g., church/synagogue) | 78 (39.6) | 49 (25.4) | 48 (42.5) | 175 (34.7) |
| I have thought about starting a conversation about death with my friends, family or members in the community | 27 (13.7) | 45 (23.3) | 16 (14.2) | 186 (36.8) |
| I have taken steps to start a conversation about death with my friends, family or members in the community | 33 (16.8) | 32 (16.6) | 11 (9.7) | 28 (5.5) |
| I have had a conversation about death with my friends, family or members in the community | 56 (28.4) | 64 (33.2) | 37 (32.7) | 53 (10.5) |
| Total | 194 | 190 | 112 | 496 |
| **Location of death preferences (i.e., at home, in the hospital)** | | | | |
| I have not thought about where I would like to die | 96 (49.5) | 85 (45) | 48 (42.9) | 230 (46.3) |
| I have thought about where I would like to die | 75 (38.7) | 77 (40.7) | 33 (29.5) | 186 (37.4) |
| I have taken steps to arrange where I would like to die | 10 (5.2) | 10 (5.3) | 8 (7.1) | 28 (5.6) |
| I have identified a location where I would like to die | 13 (6.7) | 17 (9.0) | 23 (20.5) | 53 (10.7) |
| Total | 194 | 189 | 112 | 497 |
| **Nursing home care** | | | | |

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| | HD Stage Frequency Number (%) | | | |
|---|---|---|---|---|
| | **Prodromal** | **Early Stage** | **Late Stage** | **Total** |
| I have not thought about nursing home care | 116 (59.2) | 93 (48.9) | 52 (46) | 263 (52.5) |
| I have thought about nursing home care | 61 (31.1) | 73 (38.4) | 28 (24.8) | 162 (32.3) |
| I have taken steps to arrange nursing home care | 13 (6.6) | 11 (5.8) | 6 (5.3) | 30 (6.0) |
| I have established nursing home care | 6(3.1) | 13 (6.8) | 27 (23.9) | 46 (9.2) |
| Total | 196 | 193 | 113 | 501 |
| **Palliative care** | | | | |
| I have given little or no thought to palliative care | 116 (60.4) | 101 53.7) | 63 (56.8) | 282 (57.0) |
| I have thought about palliative care | 69 (35.9) | 72 (38.3) | 23 (20.7) | 164 (33.4) |
| I have taken steps to arrange palliative care | 7 (3.6) | 12 (6.4) | 7 (6.3) | 26 (5.3) |
| I am receiving palliative care | 0 (0) | 3 (1.6) | 18 (16.2) | 21 (4.3) |
| Total | 192 | 188 | 111 | 493 |
| **Hospice care** | | | | |
| I have not thought about hospice care | 119 (61.3) | 101(52.9) | 65 (57.5) | 286 (57.2) |
| I have thought about hospice care | 73 (37.6) | 81 (42.4) | 42 (37.2) | 197 (39.4) |
| I have taken steps to arrange hospice care | 2 (1.0) | 9 (4.7) | 5 (4.4) | 16 (3.2) |
| I am receiving hospice care | 0 | 0 | 1 (0.9) | 1 (0.2) |
| Total | 194 | 191 | 113 | 500 |

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**Table 3.**

Bivariate analyses results comparing participants with and without advance directives

| Variable | Statistic | *p* value |
|---|---|---|
| Age | $t(503)=-8.632$ | <0.001[a] |
| Gender | $X^2$ (1, $n$= 505) = 0.63 | 0.426 |
| Race | $X^2$ (1, $n$= 504) = 2.48 | 0.115 |
| Marital status | $X^2$ (1, $n$= 504) = 3.84 | 0.050 |
| HD stage | $X^2$ (2, $n$= 503) = 9.01 | <0.001[a] |
| Years Education | $t(501) = -2.89$ | 0.004[a] |
| HDQLIFE Chorea SF | $t(490) = -1.66$ | 0.097 |
| HDQLIFE Speech SF | $t(499)=-1.51$ | 0.132 |
| HDQLIFE Swallowing SF | $t(498)=-2.110$ | 0.035 |
| HDQLIFE Concern for Death & Dying SF | $t(491)=1.943$ | 0.053 |
| HDQLIFE Meaning & Purpose SF | $t(493)=-2.915$ | 0.004[a] |
| PROMIS Anxiety SF | $t(330)=2.353$ | 0.019 |

[a]Note: = remains significant after Bonferroni Correction,SF = short form

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**Table 4.**

Multivariable logistic regression model

| Factor | Odds Ratio | 95% Confidence Interval |
|---|---|---|
| Age | 1.06 | 1.03 , 1.08 |
| Early Stage HD* | 1.45 | 0.76 , 2.76 |
| Late Stage HD* | 2.87 | 1.35 , 6.14 |
| Years of Education | 1.12 | 1.02 , 1.22 |
| HDQLIFE Concern for Death & Dying SF | 0.98 | 0.95 , 1.02 |
| HDQLIFE Meaning & Purpose SF | 1.01 | 0.99 , 1.05 |
| PROMIS Anxiety SF | 0.99 | 0.96 , 1.02 |

*Note: Prodromal group is the index group; SF = short form; all $p$ values <0.02.