

Brief Report

Utilization of Hospice Services in a Population of Patients With Huntington's Disease



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Abstract

Context. Although the early and middle stages of Huntington's disease (HD) and its complications have been well described, less is known about the course of late-stage illness. In particular, little is known about the population of patients who enroll in hospice.

Objectives. Our goal is to describe the characteristics of patients with HD who enrolled in hospice.

Methods. This is a retrospective cohort study of electronic medical record data from 12 not-for-profit hospices in the United States from 2008 to 2012.

Results. Of the 164,032 patients admitted to these hospices, 101 (0.06%) had a primary diagnosis of HD. Their median age was 57 (IQR 48–65) and 53 (52.5%) were women. Most patients were cared for by a spouse ($n = 36$, 36.6%) or adult child ($n = 20$, 19.8%). At the time of admission, most patients were living either at home ($n = 39$, 38.6%) or in a nursing home ($n = 41$, 40.6%). All were either bedbound or could ambulate only with assistance. The most common symptom reported during enrollment in hospice was pain ($n = 34$, 33.7%) followed by anxiety ($n = 30$, 29.7%), nausea ($n = 18$, 17.8%), and dyspnea ($n = 10$, 9.9%). Patients had a median length of stay in hospice of 42 days, which was significantly longer than that of other hospice patients in the sample (17 days), $P < 0.001$. Of the 101 patients who were admitted to hospice, 73 died, 11 were still enrolled at the time of data analysis, and 17 left hospice either because they no longer met eligibility criteria ($n = 14$, 13.7%) or because they decided to seek treatment for other medical conditions ($n = 3$, 3.0%). Of the 73 patients who died while on hospice, most died either in a nursing home ($n = 29$; 40%) or a hospital ($n = 27$; 37%). Seventeen patients (23%) died at home. No patient that started in a facility died at home.

Conclusion. Patients with HD are admitted to hospice at a younger age compared with other patients (57 vs. 76 years old) but have a significant symptom burden and limited functional status. Although hospice care emphasizes the importance of helping patients to remain in their homes, only a minority of these patients were able to die at home. *J Pain Symptom Manage* 2018;55:440–443. © 2017 American Academy of Hospice and Palliative Medicine. Published by Elsevier Inc. All rights reserved.

Key Words

Huntington's disease, hospice, CHOICE, Coalition of Hospices Organized to Investigate Comparative Effectiveness

Introduction

Huntington's disease (HD) is an autosomal dominant, progressive, and ultimately fatal, neurodegenerative disease. The median time course from diagnosis to death is approximately 15 years.¹ The most common

phenotype includes a combination of movement abnormalities, psychiatric changes, and cognitive decline. Movement abnormalities include chorea, ballism, motor impersistence, and dystonia. There are no disease-modifying therapies that cure or delay HD

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progression, and the focus remains on supportive care. Given the wide variety of symptoms, management of this population remains complex, especially in the advanced stages of the disease.

Although the trajectory of HD and its complications have been well described, much less is known about the course of late-stage illness. In particular, little is known about the population of patients who enroll in hospice. The Huntington Study Group (HSG) develops and maintains the HSG natural history database.² However, there is a considerable lack of information in the HSG database for subjects with late- and end-stage HD.³ Therefore, the goal of this study, using available hospice data, is to further characterize this population with regard to their demographics, symptom burden, length of stay, and where they die.

Methods

Patient data were extracted from the electronic medical records of 12 hospices in the CHOICE network (Coalition of Hospices Organized to Investigate Comparative Effectiveness). CHOICE is a research-focused collaborative of 72 hospices that all use Suncoast Solutions Electronic Health Record Software and have agreed to share their data for research purposes. CHOICE projects are defined and approved by a steering committee comprising leaders from all hospices in the network. Participating hospices range in size from 400 to 1700 patients/day and are located in New Mexico, California, Kentucky, Florida, Pennsylvania, Wisconsin, Michigan, Ohio, Texas (three hospices), and Kansas/Missouri. All organizations involved in this study are not-for-profit.

CHOICE obtains data from a data warehouse that participating hospices use for tracking, quality measurement, and benchmarking. Warehouse data reside on a secure server managed by Suncoast Solutions. Extracted data are stripped of identifiers to create an HIPAA-compliant limited data set that is transferred as an encrypted file to the University of Pennsylvania for analysis.

Patients were included if they were admitted to a participating hospice between January 1, 2008, and May 15, 2012. We first extracted a data set containing basic demographic variables (age, gender, and race) and diagnoses (admitting diagnosis and up to three additional diagnoses). Extraction also included site of care at the time of enrollment (home, long-term care facility, hospital, hospice inpatient unit).

Results

At the time of data collection, 164,032 patients were included. Of these, 101 patients (0.06%) had a

Table 1

Characteristics of HD Patients on Hospice (2008–2012)	
# Patients	101 (0.06%)
Median age	57 (IQR 48–65)
Gender	
Male	<i>n</i> = 48 (47.5%)
Female	<i>n</i> = 53 (52.5%)
Median days enrolled	42
Symptoms	
Pain	<i>n</i> = 34 (33.7%)
Anxiety	<i>n</i> = 30 (29.7%)
Nausea	<i>n</i> = 18 (17.8%)
Dyspnea	<i>n</i> = 10 (9.9%)
Location at death	
Home	<i>n</i> = 17 (23%)
Nursing home	<i>n</i> = 29 (40%)
Hospital/inpatient hospice	<i>n</i> = 27 (37%)

HD = Huntington's disease; IQR = interquartile range.

diagnosis of HD (Table 1). The median age of the 101 patients admitted to hospice with a diagnosis of HD was 57 (interquartile range 48–65) compared with 76 years old in the larger hospice population; 53% were women. Many were cared for by a spouse (*n* = 36, 35.6%) or adult child (*n* = 20, 19.8%). At the time of hospice admission, most were living in a long-term care facility (*n* = 41, 40.6%) or at home (*n* = 39, 38.6%). Based on palliative performance scores, all were either bedbound or could ambulate only with assistance.

Of those who died on hospice, 29 (40%) were in a long-term care facility at the time of death, compared with 28% in the total population of hospice enrollees. There were 27 (37%) who died in an inpatient hospice or hospital setting and 17 (23%) died at home. As a point of comparison, non-HD hospice patients in the data set were more likely to die at home (56%). The median length of hospice enrollment was 42 days, compared with 17 days in hospice for patients with all other diagnoses ($P \leq 0.001$).

The most commonly reported symptom during hospice admission was pain (*n* = 34, 33.7%), followed by anxiety (*n* = 30, 29.7%), nausea (*n* = 18, 17.8%), and dyspnea (*n* = 10, 9.9%). Other symptoms, specific to HD, like movement abnormalities and mood changes were not included in the data set.

Of the 101 patients, there were 73 who died and 11 who were still enrolled at the time of data analysis; 17 left hospice either because they no longer met eligibility criteria (*n* = 14, 13.9%) or because they decided to seek treatment for other conditions (*n* = 3, 3.0%). All three patients were hospitalized for acute care on the day of discharge: two were hospitalized for pneumonia and one for a hip fracture after a fall.

Discussion

We found that patients with HD were more likely than other hospice patients to be enrolled longer

and to die in a long-term care facility. We hypothesize that this may have been because of the insidious nature of the disease progression, the medical complexity of symptom management (especially with regard to psychiatric and behavioral disturbances), caregiver burden, and profound family psychosocial distress.

We found that the median length of hospice enrollment in HD patients was 42 days, compared with 17 days in hospice for patients with all other diagnoses. We propose several hypotheses to explain this difference. This may be explained by the fact that prognosis determination is challenging, in part because the typical time from diagnosis to death is approximately 15 years after onset.¹ Unlike other conditions (chronic obstructive pulmonary disease, congestive heart failure, etc.), there are no specific evidence-based criteria for determining a six-month prognosis for HD patients. Patients may also be more willing to forgo life-sustaining treatment given the lack of disease-directed therapies for HD and therefore enroll earlier in their disease course. One possibility is that the relatively younger age (i.e., potentially fewer comorbidities) of the HD population (57 vs. 76 years) may explain the longer duration on hospice. However, the CHOICE database does not maintain sufficient data on patient comorbidities to confirm this.

Early hospice referral may also be explained by the need for complex supportive care for patients and their caregivers. The need for highly complex care leads to long-term care placement before HD becomes more advanced compared with other chronic neurological conditions. This is consistent with previous data regarding HD patients not on hospice.^{4,5} Providers at a specialized, long-term care unit for people with HD report that chorea is not as disabling as the underlying emotional problems of stress, anxiety, or depression.⁶ A comparison of HD patients living at home to those living in a nursing facility did not identify significant differences in rates of depression, anxiety, or suicidal ideation. However, obsession and compulsions, delusions, auditory hallucinations, and disruptive behavior were more common in the nursing home population.⁷

This study found that the most common symptom among HD patients at the time of enrollment was pain. The prevalence of pain in HD is unknown.⁸ Multiple studies have demonstrated that same areas of the brain that modulate pain perception are also affected by HD.^{9,10} However, the pathophysiology of pain processing in HD is remains under investigation.¹¹ There is no specific literature on the assessment and management of pain in this population. However, pain assessment and management is an integral part of end stage HD and further research is needed in this area.

Anxiety was reported in 29.7% of HD patients on hospice. The prevalence of anxiety in HD has been previously reported as 13%–71% and appears to be more prevalent in HD compared with the general population.¹² Reports of successful management included olanzapine, a psychological intervention, and an intensive multidisciplinary program, but none of these methods were controlled or randomized.¹² Further research may help determine whether anxiety is associated with the pathology of the disease itself or whether other psychosocial factors primarily contribute.

Caregiver exhaustion and burnout is well known among families affected by HD, but little has been formally studied. There may be a higher psychosocial burden than in other populations given the prominence of behavioral and psychiatric symptoms and the genetic heritability and anticipation seen in this disease. It has been well established that inclusion of palliative care can reduce the burden of the caregivers. Therefore, inclusion of palliative care early on could be a valuable asset for HD caregivers.

Hospice services should be considered for all patients with advanced disease. If history predicts the future, patients will continue to die on hospice delivered in long-term care facilities rather than at home until we are better able to manage and treat their unique symptoms and psychosocial needs in the later stages of disease. Until we can delay the progression of this fatal disease, all treatment remains palliative, and patients would benefit from greater involvement of hospice in end-of-life care. Increased education about HD for hospice workers and administrators will be important in improving the care of these patients.

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