OPEN ACCESS Freely available online

Tremor and Other Hyperkinetic Movements

Reviews

Palliative Care in Huntington Disease: Personal Reflections and a Review of the Literature

Christopher G. Tarolli[1*], Amy M. Chesire[1] & Kevin M. Biglan[1,2]

[1] Department of Neurology, University of Rochester, Rochester, NY, USA, [2] Early Phase Clinical Development, Neurosciences, Eli Lilly and Company, Indianapolis, IN, USA

Abstract

Background: Huntington disease is a fatal, autosomal dominant, neurodegenerative disorder manifest by the triad of a movement disorder, behavioral disturbances, and dementia. At present, no curative or disease modifying therapies exist for the condition and current treatments are symptomatic. Palliative care is an approach to care that focuses on symptom relief, patient and caregiver support, and end of life care. There is increasing evidence of the benefit of palliative care throughout the course of neurodegenerative conditions including Parkinson disease and amyotrophic lateral sclerosis. However, beyond its application at the end of life, little is known about the role of palliative care in Huntington disease.

Methods: In this article, we discuss what is known about palliative care in Huntington disease, specifically related to early disease burden, caregiver burnout, advance care planning, and end of life care.

Results: We provide a review of the current literature and discuss our own care practices.

Discussion: We conclude by discussing questions that remain unanswered and positing ideas for future work in the field.

Keywords: Huntington disease, palliative care, caregiver burden, advance care planning, hospice

Citation: Tarolli CG, Chesire AM, Biglan KM. Palliative care in Huntington disease: personal reflections and a review of the literature. Tremor Other Hyperkinet Mov. 2017; 7. doi: 10.7916/D88057C7

* To whom correspondence should be addressed. E-mail: christopher_tarolli@urmc.rochester.edu

Editor: Elan D. Louis, Yale University, USA

Received: February 7, 2017 **Accepted:** March 20, 2017 **Published:** April 11, 2017

Funding: None.

Financial Disclosures: K.M.B.: Consultancies: KJT consulting, Teva, International Center for Postgraduate Medical Education (ICPME), North American Center for Continuing Medical Education (NACCME), WebMD Global. Advisory Boards: Teva. Grants: MJFF, NINDS, NPF, HDSA, PCORI, Greater Rochester Health Foundation, AMC Health. Employment: Eli Lilly and Company. Contracts: Presbyterian Home for Central NY, Susquehanna Nursing Home and Rehabilitation Center, Samaritan Keep Home, Sitrin NeuroCare and Focus Otsego.

Conflict of Interest: The authors report no conflict of interest.

Ethics Statement: Not applicable for this category of article.

Introduction

Huntington disease (HD) is an inherited neurodegenerative condition characterized by the combination of a movement disorder, neuropsychiatric features, and dementia. Symptoms of HD most commonly start in the fourth to sixth decade, although disease onset in the extremes of life is also seen.[1] Mood disturbances, subtle cognitive changes, and motor features (with chorea being the most common in adult-onset HD) dominate early in the condition; as the disease advances, behavioral changes, dementia, and postural instability become prominent. HD progresses relatively slowly, but significant physical and psychiatric morbidity exist throughout its course and have major impacts on both patient and caregiver wellbeing. The majority of patients will die within 10–30 years of diagnosis, typically from complications of dementia or worsening motor decline.[2]

Palliative care is an approach to the treatment of patients with chronic disease that focuses on symptom management and psychological, social, and spiritual support for patients and caregivers, rather than cure. Increasing recognition of the role of palliative care in the management of neurodegenerative conditions has emerged given the chronic and progressive nature of many neurological diseases. There has been particular focus on the benefits of palliative care in neuro-oncology, Parkinson disease (PD), and amyotrophic lateral sclerosis (ALS), but minimal exploration of the indications for palliative care or its benefits in the HD population.[3]

With no cure or disease-modifying therapy for HD, all of our currently available therapeutic options for motor and psychiatric symptoms are by definition palliative: aimed at alleviating suffering for patients and caregivers. Neurologists should feel comfortable applying these "primary" palliative care principles to all stages of care for patients with HD; the purpose of this paper, however, is not to review the symptomatic treatment for HD.[4]

Instead, we aim to consider the principles more specific to specialty palliative care. As the disease progresses, neurologists, including movement disorder specialists, are often ill-equipped to oversee and manage intractable non-neurological symptoms, mitigate patient and caregiver burden, or facilitate difficult end-of-life care discussions. Palliative care specialists can effectively manage these features, but many are unfamiliar with neurodegenerative conditions, including HD, limiting provider comfort.[5,6] Effective care at this stage of disease therefore requires the coordinated efforts of a multidisciplinary care team, often including a HD-specialized neurologist, social worker, and nurse, an actively engaged family or nursing facility staff, and, ideally, a neuropsychiatrist, dietician, speech pathologist, physical and occupational therapist, and palliative care specialist; unfortunately, access to this level of care is limited.[7]

Current literature on the application of specialty palliative care to patients with HD is relatively limited, although best practices with multidisciplinary team care in other conditions employ many of these principles. We aim to provide a review of the literature that does exist regarding palliative care in HD, and will also discuss our experience with the management of the condition, its phases, and the unique palliative care needs that should be considered in individuals with HD.

We conducted a search of the PubMed database for papers and studies evaluating the role of palliative care in HD as well as other neurodegenerative and chronic medical conditions using search terms related to palliative care in these conditions. The authors also identified articles for inclusion from knowledge of relevant literature in the field. This yielded a non-exhaustive collection of articles on the topics discussed here. Cases included in this article are taken from the authors' personal experiences in treating individuals with HD; all names used have been changed to protect confidentiality.

Early disease burden, unawareness of disease, and the challenge to care

Case 1

Susan was identified as a carrier of the HD mutation following predictive genetic testing. She was initially asymptomatic, but within the first few years following her genetic diagnosis, her motor and psychiatric burden dramatically increased. She developed generalized chorea, impaired coordination, and imbalance with regular falls. Despite her obvious difficulties, she could not see the need for physical support. Her pre-existing depression and anxiety worsened; panic attacks increased and her refractory depression overcame her. She became more erratic at work but was not aware of these difficulties herself, eventually being terminated rather than leaving voluntarily. In addition, her home life deteriorated, cascading into disarray with an abusive relationship and loss of custody of her children. Susan's loved ones and her care team saw all this happening and tried to intervene. But what was so obvious to outsiders, was not seen by Susan.

The physical symptom burden of most oncological and medical diagnoses increases as the diseases advance, driving and reinforcing the misconception that palliative care is meant exclusively for late-stage disease and patients meeting hospice criteria. However, the effects of caregiver burden, psychological distress, and demoralization on quality of life throughout the course of chronic diseases has driven increasing recognition of the role of early palliative care with trends of increased accessibility and utility of palliative care in the chronic management of those with cancer, congestive heart failure, dementia, and PD.[8–11] HD is unique in comparison, with prominent and debilitating early physical and psychiatric burden in addition to the psychological distress common to other chronic diseases. This highlights the potentially greater need for specialty palliative care involvement, or at least the application of dedicated palliative principles, early in the condition.[12]

Motor features are required to make a diagnosis of clinically manifest HD, but many patients are unaware of these symptoms. Patients may deny or under-report motor symptoms and are often unaware of their consequences despite caregiver reports of bothersome, painful, or disabling chorea, dystonia, or gait dysfunction.[13] This poses a unique challenge to the care of individuals with HD as the lack of insight into the features of their disease may make them reluctant to initiate or be compliant with symptomatic therapy.[13]

Psychiatric symptoms can also be significantly disabling in early disease stages. However, patient agnosia extends into the psychiatric manifestations of HD. Many patients do not offer symptoms without directed prompting, resulting in psychiatric disease that is inadequately recognized or managed.[14] Clinically significant mood disorders are highly prevalent throughout the disease course, with depression present in up to 40% of those with stage 1 disease.[14–16] Irritability, anxiety, behavioral disturbances, and subtle cognitive changes are also highly prevalent early in HD.[15,16]

As HD progresses, the social toll of the disease becomes significant as well. Patients invariably need to stop working because of cognitive, psychiatric, or motor disease, and the financial strain can be profound.[2] Caregivers become increasingly frustrated and intimate relationships are strained as the psychiatric and motor manifestations of the disease worsen.[17] And many patients remain unaware of the disarray that can overtake their life.

So where does the problem arise? The early burden of the disease is well recognized, yet unmet needs remain and are common.[18] First, providers less familiar with the care of patients with HD may not be aware of the agnosia common to the condition, resulting in under-recognition of disability. Second, patients and caregivers lack education on HD and cannot adequately advocate for patients who may not advocate for themselves. And, last, there are times, including early in the disease process, when symptom burden extends beyond provider comfort.[19]

In our practice, we attempt to manage these challenges by educating our patients and their families to be advocates for their own care,

through our physicians and an HD-specialized social worker. This empowers those receiving care to know what to expect and how to navigate challenging symptoms. Establishing patient and caregiver trust in provider care is also essential in managing the "nothing is wrong" attitude of many with HD. Education plays a major role here as well, with discussion of the rationale for symptomatic therapy, by directing attention to unnoticed symptoms or caregiver burden, which are necessary for the patient to agree to initiate and continue treatment. Last, we systematically evaluate the presence of common symptoms, including mood, even when patients or caregivers do not mention them. This is done not only during patient visits, but also through communication with concerned family and friends between visits; direct involvement of these individuals in care is essential to identify and understand patient symptoms. And when symptoms are identified, we feel comfortable with the initiation and management of symptomatic therapy for the diverse motor, cognitive, and psychiatric symptoms of HD.

Palliative, symptom-directed care does clearly have a role in early HD. In our practice, we provide many of these palliative services ourselves, and specialty palliative care is typically reserved for those with intractable pain, difficulty coping with their diagnosis, or late-stage disease. For the majority of providers without these resources, being aware of these common features of HD, and how to manage them, represents an important first step in mitigating early disease burden. Beyond this, palliative care can be an appropriate adjunct to care, particularly when psychiatric or somatic symptom burden extends beyond the comfort of the treating provider.[3]

This is not to say palliative care should be utilized for every patient with difficult to manage symptoms. Motor features are best treated by a neurologist, and intractable mood or psychotic symptoms best treated by a psychiatrist. But for those with pain, anxiety, sleep disturbances, and difficulty coping with their diagnosis, referral to palliative care should be strongly considered.

It is currently unclear which symptoms or at what point a referral to palliative care for symptomatic management should be made in HD, and future research should attempt to establish guidelines and standardize care.

Caregiver burden

Case 2

Annie was diagnosed with HD at age 52, more than 20 years into her marriage with John. John was a loving husband and willing caregiver, but within 5 years of her diagnosis he was experiencing the burden that HD exerts not only on patients, but also on their loved ones. He felt confined by Annie's needs; he didn't have personal time; he was overwhelmed by thoughts of long-term care planning and the inevitable inability to care for his wife. John felt an immense sense of guilt about asking others for help. The care team intervened, helped to increase Annie's social activities, and provided support for her and John. But her symptoms progressed. Her sleep became erratic, her gait dysfunction worsened, and she became fully dependent on John for all activities of daily living.

John continued to provide care at home until the day he realized he had taken on too much. He helped Annie use the bathroom one night, but fell asleep; she stayed on the toilet until morning, when John realized what had happened. John was distraught and had reached his breaking point: he was depressed, frustrated, and overwhelmed. He was no longer able to provide the care his wife needed.

Caregiver burden and burnout is a well-described phenomenon across chronic diseases, and particularly neurodegenerative conditions including HD.[20] The psychiatric burden of knowing a loved one is suffering from a life-limiting illness is stressful for caregivers, with caregivers having up to a 60% increase in mortality compared to non-caregivers.[21] In neurodegenerative conditions, caregiver quality of life, burnout, and clinically significant depression are highly correlated with the severity of patient physical disability and neuropsychiatric symptoms.[22,23]

Burnout is prevalent among caregivers of those with HD and evidence suggests that the effect of HD on caregiver mood and quality of life is more significant than in other neurodegenerative conditions.[24] Self-reported quality of life among caregivers of individuals with HD is similar to that of patients.[18] The reasons for this significant impact on caregivers may reflect the fact that family caregivers provide the majority of skilled care for individuals with HD; are in the unique position of having to worry about the risk of illness on future generations; are children of the affected patient, not emotionally equipped to take on this role reversal; may lack the knowledge necessary to navigate a complicated disease process; and that HD affects patients in their prime of life, causing significant financial strain and a drastic adjustment in family dynamics, lifestyle, and expectations.[25–27] Despite these challenges, caregivers willingly take on these roles with the relentless and often unexpected demands having a significant impact on caregiver quality of life.[17]

The need for caregiver support has been well recognized in HD for at least two decades, although gaps in service remain.[28] Available supports include educational materials, home health care or skilled nursing support, respite care, advocacy organizations, and local or online support groups. However, caregiver knowledge of or ability to take advantage of these services is highly variable and often dependent on local resources.[29,30] It is therefore imperative for neurologists and other providers to educate patients and caregivers about the availability of these services and to provide access. And it is equally important for providers to enquire not only about patient-related symptoms, but also about caregiver wellbeing.[3]

Beyond these services, ensuring adequate symptomatic control for patients, actively involving caregivers in care planning, presenting information systematically at a level appropriate for the caregiver, and asking directed questions about caregiver mood and burden can dramatically impact quality of life.[26] Systematic interventions, such as web-based educational seminars, improved provider and caregiver communication, and dedicated nursing care benefit caregiver quality of life in other chronic diseases including cancer and PD, and may have a role in the management of HD as well.[31,32]

For caregivers with needs beyond these interventions, specialty palliative care can be offered as an adjunct to care. Outside the psychological and symptom-based support for the patient, palliative

care also directs support to caregivers.[33] Specialty palliative care is documented to have a positive impact on caregiver quality of life and psychological distress in other chronic medical and neurological conditions including cancer, dementia, and PD.[34,35] Particularly for caregivers of patients with high symptom burden, referral can both primarily improve caregiver burden through counseling, reassurance and normalization, and offering respite services, and secondarily improve burden through directed treatment of patient symptomatology.

Advance care planning in HD

Case 3

Elaine was adopted, and although her adoptive parents knew about a family history of HD, they wanted to shield her from the burden of knowing she might be affected. However, by her mid-20s, Elaine had developed motor manifestations and was diagnosed through genetic testing. Her parents' desire to protect their daughter continued, avoiding difficult conversations about advancing disease or care preferences. But as Elaine's motor and cognitive disease progressed, the need to address these issues became increasingly apparent. Again, her parents tried to protect her, having conversations without Elaine's input, but they eventually saw the importance of involving Elaine herself.

By this point, Elaine had significant motor burden and her agnosia to her disease was profound; she was confident a cure was just around the corner and could not see how far she had already progressed. It was difficult to engage her in care planning conversations, although with persistence Elaine's wishes became clearer. Her parents led the conversations with the team's assistance, and Elaine's desire for better quality of life, without the use of artificial ventilation or feeding, was realized.

Advance care planning is an approach to end-of-life care that engages patients in decision-making regarding future care, while they are still able to make decisions for themselves. There is strong evidence supporting early advance care planning in individuals suffering from serious medical illnesses including cancer, congestive heart failure, chronic obstructive pulmonary disease, and end-stage renal disease.[36,37] The approach has been shown to improve patient quality of life throughout illness trajectory, increase adherence to personal patient goals, improve bereavement outcomes for family members, reduce the use of life-sustaining therapies near death, prompt earlier hospice referral, and even prolong survival.[36]

Patient opinions on the optimal timing of care planning are more variable in neurological diseases, although many of the principles still hold, offering patients the option to engage in early discussions regarding goals of care that are recommended across neurological diagnoses including glioblastoma, ALS, and PD.[38,39]

Unfortunately, current practice patterns fall short of achieving this goal. Medical and neurological providers are uncomfortable leading the conversation, citing inexperience and the lack of a structured framework.[40] Time constraints, lack of consensus between patients and family, and provider concerns about confronting patients with their own mortality also lead to delayed or ineffective conversations.[41]

Beyond these, advance care planning has unique challenges in HD. Apathy and cognitive decline may limit patient willingness and ability to engage in these conversations; patients may have an unexpected rapid decline; and patients and caregivers may not recognize the need for these conversations until it is too late because of misperceptions of disease severity.[42] Most challenging in HD, however, are the questions that remain around the timing of these conversations. Unlike most other neurodegenerative conditions, patients with HD may receive a diagnosis years before symptom onset through predictive testing. So is an "early" conversation at the time of genetic diagnosis, reminding a patient of their inevitable disease? This scenario is particularly challenging given the timing of disease onset and progression of disease is often unclear. Should we wait until a patient has manifest clinical symptoms, a time that is often fraught with the emotions of coping with the reality of a chronic and life-limiting diagnosis? Or is there a time later in the disease course, and if so, when is that and when is it too late given the impact of disease on cognitive function and decision-making?

These questions are currently unanswered, and further evaluation of patient and caregiver preferences for advance care planning in HD is necessary. However, what is clear from the literature is that the conversation should be initiated by the physician and regularly readdressed.[33] In our practice, we conduct these conversations ourselves and generally reserve them until patients show signs of advancing disease, with worsening gait or motor function, early swallowing difficulties, or early cognitive changes or behavioral disturbances. For those patients and caregivers not ready for these conversations, education about the rationale for addressing care preferences is undertaken, and the conversation is readdressed until they are ready. Topics discussed include wishes regarding artificial feeding, mechanical ventilation, cardiopulmonary resuscitation, end-of-life care settings, and patient and caregiver concerns, goals, and fears.

Neurologists caring for individuals with HD should feel comfortable initiating and leading these conversations. However, for providers with limited experience or for more complicated end-of-life care discussions, specialty palliative care again represents a useful tool. Palliative care specialists are trained and experienced in navigating these difficult topics. Although the conversation should be initiated by the patient's treating provider, involvement of a palliative care specialist in a neurologist-led advance care directive discussion or through dedicated referral can enhance the doctor–patient relationship and can enhance patient and caregiver confidence in their future care.[33]

Care settings in advanced disease and at the end of life

Case 4

Tony was fiercely independent, and for him driving and being at home had always been representations of that independence. He was diagnosed with HD at age 53, and by age 60 his disease had progressed to the point that he was no longer safe on the road. After numerous accidents, the care team needed to contact the Department of Motor Vehicles, and Tony lost his license. He suddenly felt trapped at home, became more depressed, and increased his smoking to an essentially nonstop stream of cigarettes. Tony's behavior became more erratic, and his motor

function decompensated. He was hospitalized after a fall, and had to move to a nursing home given his increasing dependence on others. Although he made some functional gains there, he was unable to return home because of his impaired sight, his skilled nursing needs, and the financial pressures of providing care at home. He felt abandoned, lonely, and dejected by his loss of independence, the one thing he had valued so greatly.

HD is broadly divided into five stages, with advancing disease stages corresponding with advancing cognitive and physical symptoms. Stages IV and V typically begin 10–20 years after disease onset, and are characterized by the need for advanced nursing care and complete or near-complete dependence on caregivers for all activities of daily living.[2,43] Given these changes, the majority of individuals with advanced HD live outside the home, most commonly in skilled nursing or long-term care facilities.[44] However, as previously noted, severe behavioral disturbances including psychosis or agitation may be present in earlier disease stages and may also dictate the need for higher-level care facilities.[45]

Dying in a person's preferred place of death is one of the central tenets of palliative care. Between 50% and 87% of all individuals indicate that home is the preferred place of death; this preference holds in those with cancer and dementia diagnoses.[46,47] Current data on place of death in HD are lacking. However, with the majority of those with advanced disease living in care facilities, most are likely to die in these settings or in hospitals. A European cohort found that less than 25% of those with HD die at home.[48]

Although the home is often an inadequate setting to provide care leading up to the end of life in HD, there are alternatives for patients to die at home or in a home-like environment. For those who have lived in a nursing facility for many years, it can feel like home for patients and their families, making it the most comfortable place to be at the end of life. Those receiving hospice benefits have an option to transition care to the home, with the support of hospice nursing. Hospice homes offer a comfortable home-like environment at the end of life and can also be considered in those with HD.[49] In our experience, for those patients and families who oppose the transition to a higher-level care facility, care in the home, with the support of home nursing, a multidisciplinary HD team, and significant social supports, can be achieved.

End of life care: an unpredictable progression

Case 5

Jim had been diagnosed with HD nearly 13 years earlier and he had progressed to late-stage disease. He continued to live at home with home nursing and a supportive family. Jim remained able to communicate, despite his advanced disease, but he fell frequently, was increasingly depressed, and was losing weight. Jim had been hospitalized three times in the same number of months with episodes of unresponsiveness in the setting of recurrent aspiration pneumonias. With each hospitalization, Jim's family had been told that it was the end, yet Jim would surprise everyone, recovering and returning home. A hospice was repeatedly discussed, but Jim didn't feel ready, worried that enrolling in a hospice meant giving up.

Jim eventually met the hospice admission criteria and agreed to referral with more rapidly advancing swallowing difficulties and an inability to maintain an adequate weight. He and his family agreed to hospice admission, and he was moved to a hospice home. He became unresponsive again one day, and his family prepared to say goodbye for the last time. Yet Jim recovered again, becoming more conversant, but also irritable and agitated. His mood and behavior continued to fluctuate over the following days and his hospice team questioned whether he was still eligible for their services. Our care team and Jim's family advocated for him: that the terminal phase of HD is unpredictable, and Jim remained there, dying peacefully within the week.

Hospice care refers to a purely palliative approach to end-of-life care, specifically directed toward improving quality of life for those with a prognosis of 6 months or less. Beyond symptom-directed care, the hospice also provides dedicated nursing, counseling, respite care, and familial support, including bereavement services. It can be an invaluable asset for dying patients.

Modern hospice care was conceptualized more than 50 years ago and the 6-month requirement for hospice candidacy developed from the relatively predictable decline in patients with terminal cancer, the population traditionally enrolled.[50] Unlike oncologic diagnoses, however, most other terminal illnesses including HD have a less predictable trajectory at the end of life, with significant fluctuation and variance in patient course once they enter "end-stage" disease.[51] Patients approach what seems like the end of life, then recover; or patients have a sudden worsening that results in death unexpectedly.

Individuals with HD can qualify for the Medicare hospice benefit through criteria for dementing illnesses (Table 1).[52] However, these

Table 1. **Medicare Hospice Benefit Criteria for Huntington Disease**

All of the Following	AND	At Least One of the Following
Inability to ambulate without assistance		Aspiration pneumonia
Inability to dress without assistance		Pyelonephritis
Urinary and fecal incontinence, intermittent or constant		Septicemia
No consistent meaningful verbal communication		Decubitus ulcers (stage 3–4)
		Fever, recurrent after antibiotics
		Inability to maintain sufficient fluid and caloric intake
In the absence of one or more of these findings, rapid decline or comorbidities may also support eligibility for hospice care		

restrictive admission criteria have limited access to hospice services for those with HD and other dementias. Although overall Medicare beneficiaries receiving hospice benefits have increased significantly over the past two decades, the shift is driven by enrollment of individuals in their last days of life. Only around 30% of those in a hospice with dementia have access to 3 or more days of hospice care.[26,49,53]

Despite recognition of the limited reliability of these criteria to predict 6-month mortality in dementia patients, attempts to create alternative criteria have been unsuccessful.[54] Further, it is unclear if there are disease-specific predictive features that may be important to consider in various forms of dementia, including HD. Ongoing advocacy for patients with HD is therefore important, to ensure that those with HD and their families have the support needed at the end of life.

We recommend hospice for any patient who has significant weight loss in the setting of swallowing dysfunction (with feeding tube not desired), or those with likely fatal comorbid disease. However, beyond these more traditional criteria for enrollment, hospice is discussed with patients and caregivers of any individual with HD who has become fully dependent on others to carry out activities of daily living. In our experience, advocacy for hospice admission for patients at this stage is successful in obtaining services, increasing access to care, and improving symptom control and quality of life for patients and caregivers.

Conclusions and future work

A palliative approach to care is an ideal match for the management of those with HD. The chronic and degenerative nature of the disease, the significant early symptomatology, and the psychological impact of HD on patients and caregivers demonstrate potential indications for this care throughout the disease course. In our experience, and that of most other groups specialized in the care of patients with HD, palliative principles are already integrated through a multidisciplinary care team, with an HD-specialized neurologist and social worker recognizing patient and caregiver needs and coordinating care with the various other specialties. What is unclear in the literature or in practice is whether there are circumstances before the end of life where specialty palliative care is a superior alternative or adjunct to this team-based approach. Identifying these factors is important not only for patients and providers with access to integrated team care, but also, and potentially more importantly, for the majority without this access.

Significant gaps remain in our knowledge of the role of specialty palliative care in the management of patients with HD. Ongoing work should evaluate this role in both early and late-stage HD, and should strive to answer the following questions:

1) Are there symptoms or transition points in the course of HD that should be used to standardize referral to specialty palliative care?
2) How can we better educate neurologists, palliative care specialists, and other providers who may interact with individuals with HD to optimize patient care and caregiver support throughout the disease course?
3) Are there reliable indicators or points in the disease course to initiate advance care planning discussions?
4) Where are patients with HD dying, and is this congruent with patient and caregiver preferred place of death?
5) How can we establish less restrictive and more predictive criteria for hospice admission in HD to improve access to care?

Ongoing work to systematically evaluate and establish better guidelines for palliative care utilization and hospice admission are necessary. However, even without these, providers can work to offer patients and families a meaningful and dignified approach to HD: by providing support for families through an unpredictable course; by establishing realistic goals and expectations about the illness and end of life; and by maintaining a commitment to patient care preferences and quality of life.

References

1. Pringsheim T, Wiltshire K, Day L, Dykeman J, Steeves T, Jette N. The incidence and prevalence of Huntington's disease: a systematic review and meta-analysis. *Mov Disord* 2012;27:1083–1091. doi: 10.1002/mds.25075

2. Dorsey ER, Beck CA, Darwin K, et al. Natural history of Huntington disease. *JAMA Neurol* 2013;70:1520–1530. doi: 10.1001/jamaneurol.2013.4408

3. Boersma I, Miyasaki J, Kutner J, Kluger B. Palliative care and neurology: time for a paradigm shift. *Neurology* 2014;83:561–567. doi: 10.1212/WNL.0000000000000674

4. Creutzfeldt CJ, Robinson MT, Holloway RG. Neurologists as primary palliative care providers: communication and practice approaches. *Neurol Clin Pract* 2016;6:40–48. doi: 10.1212/CPJ.0000000000000213

5. Turner-Stokes L, Sykes N, Silber E, Khatri A, Sutton L, Young E. From diagnosis to death: exploring the interface between neurology, rehabilitation and palliative care in managing people with long-term neurological conditions. *Clin Med* 2007;7:129–136. doi: 10.7861/clinmedicine.7-2-129

6. Manu E, Marks A, Berkman CS, Mullan P, Montagnini M, Vitale CA. Self-perceived competence among medical residents in skills needed to care for patients with advanced dementia versus metastatic cancer. *J Cancer Educ* 2012;27:515–520. doi: 10.1007/s13187-012-0351-2

7. Phillips W, Shannon KM, Barker RA. The current clinical management of Huntington's disease. *Mov Disord* 2008;23:1491–1504. doi: 10.1002/mds.21971

8. Temel JS, Greer JA, Muzikansky A, et al. Early palliative care for patients with metastatic non-small-cell lung cancer. *N Engl J Med* 2010;363:733–742. doi: 10.1056/NEJMoa1000678

9. Ritchie CS. Ushering in an era of community-based palliative care. *J Palliat Med* 2013;16:818–819. doi: 10.1089/jpm.2013.9493

10. Miyasaki JM, Long J, Mancini D, et al. Palliative care for advanced Parkinson disease: an interdisciplinary clinic and new scale, the ESAS-PD. *Parkinsonism Relat Disord* 2012;18(Suppl. 3):S6–9. doi: 10.1016/j.parkreldis.2012.06.013

11. Odenheimer G, Borson S, Sanders AE, et al. Quality improvement in neurology: dementia management quality measures (executive summary). *Am J Occup Ther* 2013;67:704–710. doi: 10.5014/ajot.2013.676004

12. Kirkwood SC, Su JL, Conneally P, Foroud T. Progression of symptoms in the early and middle stages of Huntington disease. *Arch Neurol* 2001;58:273–278. doi: 10.1001/archneur.58.2.273

13. McCusker E, Loy CT. The many facets of unawareness in Huntington disease. *Tremor Other Hyperkinet Mov* 2014;4. doi: 10.7916/D8FJ2FD3

14. Paulsen JS, Nehl C, Hoth KF, et al. Depression and stages of Huntington's disease. *J Neuropsychiatry Clin Neurosci* 2005;17:496–502. doi: 10.1176/jnp.17.4.496

15. Watt DC, Seller A. A clinico-genetic study of psychiatric disorder in Huntington's chorea. *Psychol Med.* 1993;(Suppl. 23):1–46.

16. van Duijn E, Kingma EM, van der Mast RC. Psychopathology in verified Huntington's disease gene carriers. *J Neuropsychiatry Clin Neurosci* 2007;19:441–448. doi: 10.1176/jnp.2007.19.4.441

17. Domaradzki J. The impact of Huntington disease on family carers: a literature overview. *Psychiatria Polska* 2015;49:931–944. doi: 10.12740/PP/34496

18. Ready RE, Mathews M, Leserman A, Paulsen JS. Patient and caregiver quality of life in Huntington's disease. *Mov Disord* 2008;23:721–726. doi: 10.1002/mds.21920

19. Singer C. Comprehensive treatment of Huntington disease and other choreic disorders. *Cleve Clin J Med* 2012;79(Suppl. 2):S30–34. doi: 10.3949/ccjm.79.s2a.06

20. Adelman RD, Tmanova LL, Delgado D, Dion S, Lachs MS. Caregiver burden: a clinical review. *JAMA* 2014;311:1052–1060. doi: 10.1001/jama.2014.304

21. Schulz R, Beach SR. Caregiving as a risk factor for mortality: the Caregiver Health Effects Study. *JAMA* 1999;282:2215–2219. doi: 10.1001/jama.282.23.2215

22. Takai M, Takahashi M, Iwamitsu Y, Oishi S, Miyaoka H. Subjective experiences of family caregivers of patients with dementia as predictive factors of quality of life. *Psychogeriatrics* 2011;11:98–104. doi: 10.1111/j.1479-8301.2011.00354.x

23. Pickett T AE, Paulsen JS. Caregiver burden and Huntington's disease. *Rehab Psychol* 2007;52:311–318. doi: 10.1037/0090-5550.52.3.311

24. McCabe MP, Firth L, O'Connor E. A comparison of mood and quality of life among people with progressive neurological illnesses and their caregivers. *J Clin Psychol Med Settings* 2009;16:355–362. doi: 10.1007/s10880-009-9168-5

25. Skirton H, Williams JK, Jackson Barnette J, Paulsen JS. Huntington disease: families' experiences of healthcare services. *J Adv Nurs* 2010;66:500–510. doi: 10.1111/j.1365-2648.2009.05217.x

26. Kanuit A. The experiences of Huntington's disease caregivers. Social Work, St. Catherine University, 2015. Available from http://sophia.stkate.edu/msw_papers/476.

27. Kavanaugh MS, Noh H, Zhang L. Caregiving youth knowledge and perceptions of parental end-of-life wishes in Huntington's disease. *J Soc Work End-of-life Palliat Care* 2016;12:348–365. doi: 10.1080/15524256.2016.1252828

28. Kessler S. Forgotten person in the Huntington disease family. *Am J Med Genet* 1993;48:145–150. doi: 10.1002/ajmg.1320480306

29. Roscoe LA, Corsentino E, Watkins S, McCall M, Sanchez-Ramos J. Well-being of family caregivers of persons with late-stage Huntington's disease: lessons in stress and coping. *Health Commun* 2009;24:239–248. doi: 10.1080/10410230902804133

30. Meissen GJ, Maguin E, Woodruff A. An assessment of the psychosocial needs of Huntington's disease families. *J Commun Psychol* 1987;15:245–252. doi: 10.1002/1520-6629(198704)15:<245::AID-JCOP2290150213>3.0.CO;2-V

31. Kaltenbaugh DJ, Klem ML, Hu L, Turi E, Haines AJ, Hagerty Lingler J. Using Web-based interventions to support caregivers of patients with cancer: a systematic review. *Oncol Nurs Forum* 2015;42:156–164. doi: 10.1188/15.ONF.156-164

32. Wosinski J, Delmas P, Bouwers B, Stormacq C, Kiszio B. Feasibility, appropriateness, meaningfulness and effectiveness of nursing interventions on the well-being of people with Parkinson's disease and their caregivers living in the community: a mixed-methods systematic review protocol. *JBI Database System Rev Implement Rep* 2015;13:14–29.

33. Hudson PL, Aranda S, Kristjanson LJ. Meeting the supportive needs of family caregivers in palliative care: challenges for health professionals. *J Palliat Med* 2004;7:19–25. doi: 10.1089/109662104322737214

34. Sun V, Grant M, Koczywas M, et al. Effectiveness of an interdisciplinary palliative care intervention for family caregivers in lung cancer. *Cancer* 2015;121:3737–3745. doi: 10.1002/cncr.29567

35. Lanoix M. Palliative care and Parkinson's disease: managing the chronic-palliative interface. *Chronic Ill* 2009;5:46–55. doi: 10.1177/1742395309102819

36. Bernacki RE, Block SD. Communication about serious illness care goals: a review and synthesis of best practices. *JAMA Intern Med* 2014;174:1994–2003. doi: 10.1001/jamainternmed.2014.5271

37. Brom L, Pasman HR, Widdershoven GA, et al. Patients' preferences for participation in treatment decision-making at the end of life: qualitative interviews with advanced cancer patients. *PloS One* 2014;9:e100435. doi: 10.1371/journal.pone.0100435

38. Tuck KK, Brod L, Nutt J, Fromme EK. Preferences of patients with Parkinson's disease for communication about advanced care planning. *Am J Hosp Palliat Care* 2015;32:68–77. doi: 10.1177/1049909113504241

39. Connolly S, Galvin M, Hardiman O. End-of-life management in patients with amyotrophic lateral sclerosis. *Lancet Neurol* 2015;14:435–442. doi: 10.1016/S1474-4422(14)70221-2

40. Rizzo VM, Engelhardt J, Tobin D, et al. Use of the Stages of Change transtheoretical model in end-of-life planning conversations. *J Palliat Med* 2010;13:267–271. doi: 10.1089/jpm.2009.0281

41. Sharp T, Moran E, Kuhn I, Barclay S. Do the elderly have a voice? Advance care planning discussions with frail and older individuals: a systematic literature review and narrative synthesis. *Br J Gen Pract* 2013;63:e657–668. doi: 10.3399/bjgp13X673667

42. Klager J, Duckett A, Sandler S, Moskowitz C. Huntington's disease: a caring approach to the end of life. *Care Manag J* 2008;9:75–81. doi: 10.1891/1521-0987.9.2.75

43. Shoulson I, Fahn S. Huntington disease: clinical care and evaluation. *Neurology* 1979;29:1–3. doi: 10.1212/WNL.29.1.1

44. Rosenblatt A, Kumar BV, Margolis RL, Welsh CS, Ross CA. Factors contributing to institutionalization in patients with Huntington's disease. *Mov Disord* 2011;26:1711–1716. doi: 10.1002/mds.23716

45. Nance MA, Sanders G. Characteristics of individuals with Huntington disease in long-term care. *Mov Disord* 1996;11:542–548. doi: 10.1002/mds.870110509

46. Higginson IJ, Sarmento VP, Calanzani N, Benalia H, Gomes B. Dying at home—is it better: a narrative appraisal of the state of the science. *Palliat Med* 2013;27:918–924. doi: 10.1177/0269216313487940

47. Gomes B, Higginson IJ. Factors influencing death at home in terminally ill patients with cancer: systematic review. *BMJ* 2006;332:515–521. doi: 10.1136/bmj.38740.614954.55

48. Rodrigues F, Abreu D, Damasio J, Ferreira J. Causes of death in a European Huntington's disease cohort (registry). *J Neurol Neurosurg Psychiatry* 2014;85(Suppl. 1):A64–A65. doi: 10.1136/jnnp-2014-309032.183

49. NHPCO's Facts and figures: hospice care in America. National Hospice and Palliative Care Organization, 2015. Available from http://www.nhpco.org/sites/default/files/public/Statistics_Research/2015_Facts_Figures.pdf:

50. Brown GM. This porridge is too thin. *Camb Q Healthc Ethics* 1998;7:206–207. doi: 10.1017/S0963180198002163

51. Hanrahan P, Luchins DJ. Access to hospice programs in end-stage dementia: a national survey of hospice programs. *J Am Geriatr Soc* 1995;43:56–59. doi: 10.1111/j.1532-5415.1995.tb06243.x

52. Luchins DJ, Hanrahan P, Murphy K. Criteria for enrolling dementia patients in hospice. *J Am Geriatr Soc* 1997;45:1054–1059. doi: 10.1111/j.1532-5415.1997.tb05966.x

53. Christakis NA, Escarce JJ. Survival of Medicare patients after enrollment in hospice programs. *N Engl J Med* 1996;335:172–178. doi: 10.1056/NEJM199607183350306

54. Brown MA, Sampson EL, Jones L, Barron AM. Prognostic indicators of 6-month mortality in elderly people with advanced dementia: a systematic review. *Palliat Med* 2013;27(5):389–400. doi: 10.1177/0269216312465649