

Appendix B

Education Subcommittee Report

Introduction

As the concepts of palliative and end-of-life care enter mainstream education in the United States, consumers, medical professionals and other health care providers are recognizing the comprehensive range of treatments offered to individuals and families coping with end-of-life issues. Some of these treatments remain curative, but many now focus on the palliative aspects of providing care.

People with HD and their families want more sensitive involvement by medical professionals during the end-of-life process. Empathy on the part of health care providers is important to achieve a sense of completion, including resolving conflicts, saying good-bye and contributing to the well-being of others in the context of illness. Medical professionals can provide this support to people and families coping with Huntington's Disease only if they are properly trained and emotionally prepared for dealing with end-of-life issues, and they possess an understanding of the myriad complications that accompany the progression of Huntington's Disease.

The Education Subcommittee conducted an extensive literature review through Medline, CINAHL, PubMed and Virgo, as well as the online annotated bibliography of the *Promoting Excellence in End-of-Life Care* Critical Care Workgroup. Informal surveys of medical professionals and caregivers working within the HD field were also conducted. Other than a few materials published by the Huntington's Disease Society of America

(HDSA) and one substantive article by Moskowitz and Marder in 2001, there is a dearth of available information on end-of-life care issues for Huntington's Disease. Although HDSA has distributed almost 12,000 copies of their *Physicians Guide to the Management of Huntington's Disease* (Second Edition), this 90-page publication includes scant information about end-of-life care.

Deficiencies in Education for Individuals with Huntington's Disease and Their Caregivers

The recent name change of the *National Hospice Organization* to the *National Hospice and Palliative Care Organization* reflects an increased awareness by policy-makers and health care providers of the need to extend quality end-of-life care to those facing life-limiting health conditions that last far longer than six months. People and families coping with HD comprise a unique population that stands to benefit enormously from this new, more broadly defined palliative care movement. According to the World Health Organization, the need for palliative care in people with cancer begins with diagnosis. The same holds true for Huntington's Disease. HD is not just an individual illness. It is a family crisis that begins with the knowledge that any family member may have the HD gene. For some, awareness of the HD gene in the family gene pool signals the end of life as they have experienced or envisioned it. To reduce this trauma, families would benefit from palliative care options early in the disease process. Caregivers in HD families need access to education and to supportive care in order to cope effectively with their suffering and loss. As the spouse of a husband with HD observed:

"Illness and disability is a family affair. The diagnosis that made our family member need care happened to us as well. It is our diagnosis just as much as it is

theirs. I have a psychosocial form of the disease, just as my husband has a clinical one." (1)

Although there is a wealth of palliative care resources that are not specific to HD and published materials about the impact of Huntington's Disease on families (especially on the Internet), there is a lack of materials that educate HD families about palliative care or provide resources that relate to specific features of this long-term genetic family disease.

The impact of predictive genetic testing for HD on family systems cannot be overstated. As Sobel and Cowan found, the consequences of the genetic testing process are often unanticipated and may be disruptive to family functioning. As one family member commented:

"Somebody could've said this [the testing] is going to change everything in your life, not only for you, but change everything for your whole family forever. You can't take this one back." (2)

Sobel and Cowan suggest that families should be involved when a member decides to undergo predictive testing so all members will be prepared for a positive or negative test result. This kind of research information should be available to families who are struggling with the issues surrounding genetic testing.

Appropriate palliative care requires the recognition of loss and grief of people with HD and their families. Unresolved grief interferes with individual and family functioning. While many resources about grief and bereavement exist, including a few specific to HD families (3), little attention has been focused on the many non-death related losses facing people with HD and their families, including the decline of physical, cognitive and social abilities. Less obvious causes for grief include the loss of innocence that follows the

results of predictive testing, as well as a loss of a healthy future, of independence, of marriage and of parenthood. These secondary losses, which often go unrecognized, may lead to unreconciled or disenfranchised grief (4) that may later interfere with the person's ability to cope effectively with other aspects of the disease. For spouses of people with HD, psychosocial losses include the loss of a significant relationship, as well as loss of freedom, of a future and of a supportive partner in old age. As one spouse lamented,

"Less attention...is paid to caregiver grief, that relentless ongoing process that is brought about, not by a loved one's death, but by the changed aspects of their life, and inevitably of our own." (5)

According to Ira Byock, M.D.,

"The touchstone of dying well—the sense of growing individually or together in the midst of dying—is that the experience is of value and meaningful for the person and their family." (6)

Market research indicates that a high proportion of Internet usage is by people seeking health information. An “end-of-life care for Huntington’s Disease” Internet search produced many sites that are derivative in nature, i.e., reprints from or links to a small number of sites with original information (www.caregiver.org, www.hdsa.org, www.rwjf.org and www.hsc-ca.org). For the most part, Huntington’s Disease is often mentioned incidentally at these derivative sites. The venerable PubMed site identified three articles, the first of which is essentially the only extant tutorial on end-of-life care for people with HD. (7)

Deficiencies in Education for Medical/Health Care Professionals

Medical education for Huntington's Disease usually occurs during the second and third years of a four-year physician's education program and is most often presented as lectures on movement disorders, degenerative dementias, trinucleotide repeat disorders and genetic diseases. Similar but less extensive training is provided in physician assistant coursework, while neurology and psychiatry residency education programs may offer the most inclusive training for the care of people with Huntington's Disease. These programs often focus on the pathophysiological aspects of disease progression rather than the cognitive and behavioral aspects that are prevalent during the advanced stages of HD.

Lectures on movement disorders, and their associated medical management, are often part of nursing education programs. Clinical psychologists, clinical social workers and other mental health counselors learn about Huntington's Disease during their training, but their education seldom covers the family dynamics of those who care for people with HD during the progressive decline of the disease while they reside in the home. Education programs for occupational therapy, physical therapy and speech-language pathology often address safety, mobility, self-care, communication and swallowing issues of individuals with neurodegenerative disease though most of these programs focus on rehabilitative rather than palliative care.

Continuing Medical Education (CME) about HD for practitioners is often found only in geographic areas served by HDSA-funded Centers of Excellence (currently 17 in the

U.S.), where the multidisciplinary team is empowered to share information about all aspects of this disease with other medical professionals.

Most medical professionals, then, define HD as a movement disorder. Education regarding the less obvious, yet equally disabling, cognitive and behavioral aspects of the disease are not adequately covered, nor are palliative interventions that are so critical for coping with HD throughout the progression of the disease. Medical education in the United States emphasizes medical diagnosis and treatment, while educational offerings from the United Kingdom, Canada and Australia focus more on client-centered care. Because HD treatment in the U.S. occurs primarily in hospitals, clinics or nursing homes, the professional practice literature reflects this attachment to the medical model.

For all care providers, interventions that are addressed in professional literature and educational programming tend to focus on cure rather than comfort, and independence rather than autonomy. This emphasis, coupled with related client and family psychodynamics, can lead to delayed or inappropriate treatment. Earlier and more effective recognition, by health care providers, of the functional difficulties and end-of-life care wishes of people with HD and their families may help to alleviate personal and family distress that occurs with misdiagnosis or when inappropriate or unwanted treatments are provided.

End-of-life care training is available to medical professionals through the American Medical Association's (AMA) Project on Education for Physicians on End-of-life Care

(EPEC). The project's goal is to improve the competency of all U.S. physicians in end-of-life care. EPEC programs have been introduced in the last five years in medical schools across the country. In 1997, The Robert Wood Johnson Foundation launched the *Promoting Excellence in End-of-Life Care* national program office with a mission to improve care and the quality of life for dying Americans and their families. The growing importance of palliative medicine (intended to improve the comfort and quality of life for patients) is supported by professional journals and organizations such as the American Medical Association. Several Web sites, such as the Toolkit for Instruments to Measure End-of-Life Care (<http://www.chcr.brown.edu/PCOC/TOOLKIT.HTM>) and End-of-Life/Palliative Education Resource Center (<http://www.eperc.mcw.edu/start.cfm>), were recently created to provide educational resources in end-of-life care.

Physicians may refer patients for hospice care, but in many areas, hospice admission criteria continue to support entry into the program when the individual has less than six months to live. Furthermore, under CMS guidelines, once someone is in hospice care, attempts at curative health care measures are precluded. Therefore, the most appropriate palliative and end-of-life care for people with HD or other patients is often underutilized until just before a predicted death. Hospice care includes personal, family and spiritual support for the dying patient as well as medical management of symptoms and pain.

Delays in providing services to people with HD might be minimized if physicians were better educated about standards for hospice care, which are listed on hospice Web sites. Additionally, people with HD would benefit from provider knowledge about two rating scales used to justify referral to hospice care:

- Functional Assessment Staging (FAST) with scores at or below level 7.A., and
- Karnofsky Performance Status Scale with scores at or less than 30 percent (see Addendum C of Care Subcommittee Report).

These rating scales are helpful to those less experienced in determining the appropriateness of hospice referral, and are useful to all health care providers in justifying the changes in medical benefit funding.

Deficiencies in Education for First Responders

In a crisis, first responders such as firefighters, police officers, paramedics and emergency medical technicians are often the first points of contact between a person with HD and the medical system. Misperceptions at this juncture often lead to the inaccurate conclusion that the person may be intoxicated. Placement in a holding cell not only delays adequate medical assessment and treatment, but also places the person with Huntington's Disease at risk of harm.

A trained examiner is capable of recognizing the bloodshot eyes and odor of alcohol in a person who is intoxicated or the small pupils of a person on illegal medications. Slurred speech, poorly coordinated movements and misperceptions of reality by a person with HD closely resemble those of a person under the influence of alcohol or illicit medications. At a minimum, first responders would benefit from training that enables them to question whether abnormal behavior is due to mental or organic illness, rather than intoxication.

Current training of first responders varies greatly in different municipalities. Some national organizations like the American Red Cross sponsor training and provide materials, but all too often, it is the first responder's hard-won field experience that guides accurate evaluation. A review of materials for first responders by Anthony C. Stein, Ph.D., a psychologist involved in training first responders, found that the information on mental and organic illnesses was severely limited. People with HD are often not recognized as having an organic illness and inappropriate care is consequently rendered despite the best intentions of first responders.

Medical alert bracelets and necklaces are inexpensive, inoffensive and well-accepted methods of providing crucial medical information for use in emergencies. First responders and medical personnel are trained to look for these during initial evaluations because a person with HD is often unable to produce a card from a wallet or pocketbook, and family members are not always available. Education of first responders during training and re-certification, and the use of medical alert bracelets, would ensure people with HD receive appropriate evaluation and treatment during periods of crisis.

Families should be aware of the *Safe Return Program* of the American Alzheimer's Association. This low-cost technology uses bracelets, necklaces and clothing tags to identify individuals with dementia who are at risk of wandering and may benefit those with HD as well. (8)

Deficiencies in Nursing Home Care

Most people with late stage HD are cared for in nursing homes or other long-term care facilities. Unfortunately, several factors impede quality end-of-life care for people with HD in these settings.

Due to the small number of HD residents, most long-term care facilities do not have a separate wing for people with HD. These residents, who often are younger than most long-term care residents, may lack age-appropriate activities and peer groups with whom to interact.

Lack of sufficient staff training in understanding the symptoms of HD and appropriate treatment/intervention strategies for people with HD often results in ineffective treatment for the person with HD and feelings of frustration and guilt on the part of staff when interventions are not successful.

Lack of understanding of the symptoms associated with HD on the part of other long-term care residents and their families may result in a negative response to people with HD. Fear of being displaced from the long-term care setting to a psychiatric facility further reduces the quality of life for residents with HD and their families.

Finally, state officials who evaluate and monitor long-term care facilities are often unaware of the challenges that staff face in keeping residents with HD safe. Thus, they

may inappropriately be critical of the use of physical restraints for example, which staff deems necessary for the resident's well-being.

Suggestions for Educational Enhancements

The Education Subcommittee has identified significant gaps in education regarding palliative care and HD. These gaps result in inadequate knowledge, insufficient skills and improper attitudes that negatively impact effective palliative care interventions along the continuum of HD. Education about palliative care options for people with HD and their families must become part of the mainstream education of physicians, psychotherapists, occupational therapists, physical therapists, speech-language pathologists, social workers, nurses and others who provide end-of-life care for people with HD. The HD Peer Workgroup Education Subcommittee suggests the following:

1. Recommendations for Enhanced Education of Health Care Providers

- Develop a core curriculum for use in the training of health care professionals that emphasizes cognitive and behavioral complications of HD as well as the physiologic symptomology. Such curriculum would include basic features of HD with special emphasis on cognitive and behavioral aspects of care at the later stages of the disease. An additional focus of the curriculum would be methodology for assessment of independent function and decision making about palliative and end-of-life care issues.
- Increase medical education stressing palliative care in neurodegenerative disorders. Methods might include modification of the AMA's EPEC program or development of a mini-internship program, offered through HDSA Centers of Excellence for medical practitioners and other health care providers. Education would address specialized training needs for HD and other neurodegenerative disorders, an increased emphasis on palliative care in board review training programs and publication of articles in peer reviewed journals and chapters in neurology/palliative care textbooks.
- Develop specific training programs for physicians and genetic counselors that address the implications of HD genetic testing. These programs must teach techniques of pre- and post-test counseling for at-risk individuals and how to include the entire HD family, when possible and appropriate.
- Increase awareness of the referral process for admission into hospice and palliative care systems. Recommendations include the development of standardized admission criteria for entry into hospice programs and

development of standardized prescriptions for hospice management of people with Huntington's Disease.

- Increase training in evaluating, selecting and acquiring adaptive equipment and assistive technology for people with HD at the various stages of the disease. Educational offerings might include articles or presentations that address the progressive functional decline associated with HD and the need to evaluate long-term device safety as well as convenience before recommending devices.
- Increase training in effective home and facility modifications that promote autonomy of people with HD while accommodating the progression of the disease.
- Produce publications and presentations that stress the efficacy of individualized treatment techniques designed to improve quality of life, and recognition of a person's right to refuse intervention.
- Provide professional care providers with resources to help them improve their ability to help HD families attain several important goals: personal safety during activities of daily living coupled with decline in functional mobility; injury prevention while fostering independence; and maintenance of a healthy balance between determination and acceptance of functional decline.

2. Recommendations for Enhanced Education of the First Responder

- Develop educational products that enable the first responder to question whether aberrant behavior is due to mental or organic illness. One such product is a *Quick Reference Card for Dementias and Mental Illnesses* for use by first responders.
- Develop education about steps to take when a first responder encounters an individual with an HD medical alert bracelet. Educational offerings must include programs for police and emergency workers that explain both the motor-sensory and cognitive-behavioral components of advancing HD.

3. Recommendations for Enhanced Education for HD Families

Provide quality palliative and end-of-life care for family members to promote a better understanding of:

- Salient dimensions of palliative care;
- Importance of executing Advance Directives that promote autonomy and death with dignity;
- Nature of the psychological suffering of people with HD and their families as physical symptoms increase for the person with HD;
- Impact on family members of genetic testing for the HD gene among asymptomatic members;
- Nature of losses and concomitant grief for families dealing with HD;
- Resources to insist on utilizing health care providers with expertise in the palliative care aspects for living with HD; and
- Pamphlets, Internet sites, presentations at support group meetings and reprints of scientific journal articles as sources of information.

4. Recommendations for Long-Term Care Facilities Providing Care to People with HD

- Where possible, create separate wings for people with HD, or establish age-appropriate residential wings. This enhances the quality of life and encourages the development of specialized staff to provide care for people with HD.
- Familiarize staff with resources available from the Huntington's Disease Society of America. Staff frustration and burnout in working with HD residents is correlated with lack of training about the nature of HD and appropriate interventions.
- Provide regional in-service programs for staff that work with HD residents in long-term care settings.
- Create a pocket reference card which long-term care facility staff would carry that describes stages of HD, symptoms and appropriate functional interventions.
- Provide pamphlets and other printed information about HD to families of other residents to reduce negative responses to disruptive actions of some people with HD.
- Educate state inspectors of long-term care facilities about the specific characteristics of HD and appropriate interventions employed by staff.

Conclusion

The research and development agenda for this educational topic is in its infancy. These recommendations are offered as a first step to help promote excellence in end-of-life care for Huntington's Disease. The goals are to empower the person with HD and the family in acquiring a personal and family-centered approach to care that provides support for autonomy and self-direction in decision making, alleviates physical, emotional and spiritual suffering and offers bereavement support for all family members.