

## Appendix A

### ***Care Subcommittee Report***

#### **Introduction**

At the time of diagnosis for people with HD, it is critical to proactively address a number of problems that occur throughout the course of the disease. Early attention is especially important because people with HD slowly lose their ability to communicate and may be unable to voice their preferences. Advance Directives and durable power of attorney are two instruments that play important roles in decision making for treatment options in the later stage of this disease. They not only state treatment preferences but also relieve family members from the burden of making those decisions in the absence of input from the person with HD. Protecting patients' dignity often becomes a challenging task as the disease advances. Educating people with HD, caregivers, family members, physicians, nurses and other health care professionals about the various problems caused by HD is essential for good care.

The HD Peer Workgroup defined *end of life* as that stage in which people with HD can no longer care for themselves. As the disease progresses and the person with HD approaches the end of life, many problems in the area of care arise. The Care Subcommittee chose the following domains as they considered specific problems and solutions germane to HD:

- Autonomy,
- Dignity,
- Meaningful Social Interaction,

- Communication,
- Comfort,
- Safety and Order,
- Spirituality,
- Enjoyment, Entertainment, and Well-being,
- Nutrition, and
- Functional Competence.

This report discusses each of these domains in three different settings: home, long-term care facility and hospital. It first defines the domain of care. Then, in each setting, it presents ideal and worst case scenarios of care based on actual patient experience. Both the best and worst case scenarios in this report describe real and actual experiences of people living with Huntington's Disease. Lastly, the report suggests solutions and resources that modify a worst case toward improved outcomes. Vignettes illustrate key points in each domain.

## ***Autonomy***

*The quality or state of being self-governing; right of self-government; self directing freedom and especially moral independence. Autonomy is more difficult to implement since the person with HD is no longer totally self-governing, free or independent. Their preferences and wishes, as stated in Advance Directives or shared with significant others during their life, need to be upheld to respect their independence and individuality.*

## **Home Setting**

This setting provides the most autonomy for the person with HD, prior to entering the end-stage of the disease.

## ***Ideal Scenario***

The person with HD has previously identified, verbally and in writing, their preferences for care at this stage of illness. Although they are no longer autonomous in the physical sense (e.g., self-governing or independent), the family caregivers can reassure the person with HD that care is being provided in ways that they have chosen.

## ***Worst Scenarios***

- The person with HD lives alone. The need for autonomy leaves them without family or friends to assist with care needs. Ultimately, their condition deteriorates to the point of death.
- The person is evicted from their living situation and the courts force admission to a long-term care facility.

- Family or other caregivers assume total control and decision making for the person with HD regardless of personal preferences. No attempts are made to understand the person with HD, or their emotional/psychological needs.

### ***Solution***

Educate physicians, especially neurologists, about resources that are available for people with HD and their families. When a diagnosis is made, information is shared to ensure support, education and continuity of care as the disease progresses. Community resources for HD include, but are not limited to, HD clinics, HDSA support groups, chapters and Centers of Excellence. Families need to have a solid connection to help them meet the challenge of HD.

### **Long-Term Care Facility**

For a person with HD in the end-stages of the disease, the situation is similar to the home setting described above.

### ***Ideal Scenario***

The staff has been educated about HD and the importance of respecting the independence, individuality and autonomy of the person with HD up to and including the time of death. The person with HD has been in this setting for many months and the staff understands their needs and wishes through communication that occurred prior to admission. Written instructions and the involvement of a health care agent or informed family member help staff comply with the person's wishes.

### ***Worst Scenario***

The person with HD has no advocate and is isolated. Staff is not knowledgeable about the person with HD's personality or preferences. Meaningful communication between the person with HD and staff is non-existent. The staff and the primary care provider decide on all care needs.

### ***Solution***

All long-term care facilities have information about HD and local resources. When a person with HD is admitted for care, staff give contact information about local resources to the person and family. An HD social worker interacts with the facility staff and the family in the best interest of the person with HD.

### ***Hospital Setting***

Generally, the person in late-stage HD enters a hospital for acute care (e.g., aspiration pneumonia). There may not be time to educate staff about HD or the person's preferences.

### ***Ideal Scenario***

Involved family members are present upon admission of the person with HD to the hospital. An interdisciplinary team including the primary care physician, attending physician/neurologist, nurse, social worker and hospital chaplain consult with the health care agent and family about the treatment plan. The person with HD's Advance Directives and other written instructions are presented to the team.

### ***Worst Scenario***

The person with HD is admitted without an advocate or written instructions for care.

They are unable to communicate. An attending physician, unfamiliar with the person with HD's medical history, makes decisions based upon the presenting data. The person with HD has no choice in the care administered.

### ***Solution***

Provide family or other caregivers with appropriate information, education, support and resources to reinforce the importance of the autonomy of the person with HD. Keep communication open between all parties involved.

### **Consider**

Jack led a full and vibrant life. He had earned his Ph.D. and two Masters Degrees at a major university and, as a professor, enjoyed influencing the lives of young and old alike with his vibrant, upbeat personality.

Jack's life changed dramatically after his HD diagnosis. Jack felt the loss keenly when he could no longer safely drive his car to work every day or manage the complicated demands of his position. With his autonomy and independence dramatically affected, he fell into a deep depression. With the help of an understanding wife, as well as a qualified and sensitive occupational therapist, Jack learned to utilize public transportation. This adaptation allowed him to maintain his connection with co-workers and continue working in a newly established office arranged by his department for his use.

Returning home from work one day, Jack began to cross the street but with his slowed gait and poor judgment, he did not reach the other side before the light changed, and he was hit by an oncoming car. Although his injuries were minor, he and his family were again faced with changes in his life and lifestyle due to HD.

Jack eventually agreed to attend a local day care center, although he maintained that he did not need anyone watching over him while his wife worked. His outgoing personality and positive attitude quickly endeared him to the staff and residents, however, and Jack soon became a staff assistant rather than a resident. He helped others with activities and chatted with residents and staff. Encouraging and

supporting others allowed him to maintain his dignity and independence to the greatest extent possible.

### **Conclusion**

Although autonomy is more difficult to maintain in the later stages of HD, the concept of limited autonomy, wherein the person continues to act independently to the fullest extent possible, remains a significant aspect of the overall care and functioning of the person with HD.

***Dignity***

*The quality or state of being worthy, honored or esteemed; formal reserve of manner or language; high rank, office or position.*

Symptoms of HD usually manifest between 30 and 50 years of age, when an individual is most productive and able to contribute to self, family, community and society. The majority of people with HD have had the opportunity to take pride in their personal, educational and professional achievements. Many have led lives of accomplishment and distinction. HD is not discriminatory in that it touches the lives of attorneys, accountants, college professors, homemakers, teachers, physicians, and musicians. In a more practical sense, it touches the lives of spouses, children, mothers, fathers, brothers and sisters. The dignity, pride and self-esteem attained by a person with HD during the individual's functional years slowly erode as the cognitive, emotional and physical effects of HD invade the body.

How can dignity be maintained in the later stages of HD? How can others ensure that the person with HD is treated with dignity and respect in all settings?

**Home Setting**

The person with HD is treated with the dignity they deserve. This assumption hinges on the caregivers involved in day-to-day care. A completed *Values History Form* (at the end of this Subcommittee report) defines which family members are sensitive to the HD person's needs and personality. The form also identifies other caregivers with whom the



person with HD has built trust and rapport as well as primary interests in food, entertainment and sensory stimulation.

### ***Ideal Scenario***

The person with HD is surrounded by the comfort and familiarity of home and family. Their participation in and contribution to family, community and profession are evident in pictures, trophies, awards and commendations, which serve as a testimony to who they are beneath the veil of HD. A sense of pride, dignity and self-worth are supported with the knowledge that their accomplishments, large or small, are recognized and respected by others. Family, friends, spiritual advisers and other significant people in the life of the person with HD continue to visit, validating their personhood and maintaining contact and interest with the outside world as the person with HD becomes less able to participate in activities outside the home.

The primary caregiver is physically and emotionally well adjusted and committed to meeting the needs of the person with HD. The caregiver attends HD support group meetings and other HD functions to obtain the knowledge and support needed for meeting the continuing challenges of HD. The neurologist and HD clinic that provide care for the person maintain a good working relationship that ensures consistent quality care for symptom control and disease progression. These contacts identify needed resources. The caregiver has access to respite from the daily demands using other competent family, friends or professional in-home help. Hospice care is available to enhance the care and dignity of the person with HD and their family until death.

### ***Worst Scenario***

The person with HD lives alone, refuses offers of help, or alienates all friends and relatives. Conditions in the home are sub-standard. Nutritional needs are not met and medical care is minimal or non-existent. Dignity and self-esteem spiral downward. Eventually a crisis occurs (e.g., fire from inattentive smoking, infestation, power shut-off from non-payment of bills) and intervention is forced through the court system. Death may occur as a result of choking, aspiration pneumonia, overall neglect of personal and medical needs or suicide.

### ***Solutions***

Caring, committed family relationships are preserved and nurtured. Education and resources are available to family and caregivers. These ideals are best established early in life since they are less available in the later stages when the person with HD needs the most understanding and care.

### ***Long-Term Care Facility***

For many families, a long-term care facility is eventually the most suitable setting for continued day-to-day care. It is extremely important to emphasize with staff that a person with HD is acutely aware of their surroundings, as is a person with severe Alzheimer's disease. The person with HD understands what is being said and what is happening in the environment. Treating a person with HD with dignity and respect, and offering reassurance and support, ensures more acceptable behavior and cooperation.

### ***Ideal Scenario***

The primary caregiving staff has some familiarity with HD. In addition, they arrange for an educational in-service before the person with HD is admitted to familiarize all staff, from the maintenance person to the administrator, about the unique needs and behavioral nuances of a person with HD. The family and the community resources for HD provide the staff with a list of educational resources. The information includes Internet sites and contact information for local, regional and national HD community resources. A copy of the HDSA publication *The Physician's Guide to the Management of Huntington's Disease* (Second Edition) is provided to the facility by the HDSA community resource (chapter, Center of Excellence or support group). The long-term care facility is added to the HDSA mailing list to receive continuing information about HD and educational programs for caregivers and professionals. A social worker affiliated with the community HD resource visits the long-term care facility to update staff about the care needs of the person with HD. Staff turnover is low, since the staff is empowered to make changes for the benefit of the person with HD and the facility. In-services are repeated periodically and a video presentation on the care of a person with HD is available for review by staff. The *Patients' Bill of Rights* is presented to the person with HD and their family during the admissions process to guarantee that dignity and respect are upheld. As a legal agreement, the staff acknowledges the *Bill of Rights* as a part of the care plan.

Hospice services are enlisted. Hospice care addresses the needs of the person with HD and their family from a holistic rather than a problem-oriented perspective and they

oversee care of the person with HD at end-stage in the long-term care facility.

Throughout the stay, the family remains involved and monitors care.

### ***Worst Scenario***

The long-term care staff is unaware of the care of the person with HD. The long-term care facility is understaffed and unclean. The facility medical director is minimally familiar with HD and is unaware of *The Physician's Guide to the Management of Huntington's Disease*. There are insufficient resources and no contact is made with community HD resources for in-service or information. The family is not involved to advocate for the person with HD and they are ignored. The person with HD is unable to communicate, which becomes increasingly frustrating for them and the staff. The person with HD is isolated, with no social interaction. They die without dignity or comfort.

### ***Solution***

Educate long-term care facilities' staff. Provide educational resources such as *The Physician's Guide to the Management of Huntington's Disease* (Second Edition), *Understanding Behavior in HD* and *A Caregiver's Handbook for Advanced-Stage HD*. Consistently involve the HD social worker or HD volunteers to monitor the needs of the person with HD and staff at the long-term care facility.

### **Hospital Setting**

Every hospital and health care institution must uphold and apply the *Patients' Bill of Rights* and *Code of Ethics* to all patients being served. These rights include the dignity

and respect of the individual. Often, the hospital setting for care of late-stage HD occurs during a time of crisis. Due to the critical care need and short-term stay, the interdisciplinary team may consist of only a physician, nurse and occasionally a social worker and a chaplain. It is difficult to educate during a crisis, since the focus is on the immediate need (e.g., aspiration pneumonia or organ failure) and not on the broader dynamics of HD.

### ***Ideal Scenario***

The primary care physician is familiar with the HD person's medical history and attends to both the family and the person. Family members, including the one appointed by the *Durable Power of Attorney for Health Care Decision*, are present and part of the treatment team. Explicit written instructions defining care preferences (Advance Directives), previously prepared by the person with HD, are reviewed by the medical team. The health care agent informs and educates the team about the person with HD's intentions for care. All agree to respect the expressed wishes of the person with HD and to uphold their dignity.

### ***Worst Scenario***

The person with HD is admitted to the hospital unable to communicate and without an advocate. Hospital staff is unaware of the person's diagnosis and treats only the presenting symptoms (e.g., aspiration or injury due to fall). Because the person with HD is unable to communicate and may be disheveled, the medical team assumes they are demented or mentally ill. The dignity of the person with HD is not a factor in treatment

decisions. Social workers and other advocates are not involved. The person with HD dies alone or is discharged to a mental unit of a local long-term care facility.

### **Consider**

Susan's husband left her when her HD symptoms became pronounced and sometimes problematic. Her three teenage children chose to stay with their father rather than care for Mom who could be unpredictable. Depression and loneliness added to Susan's already difficult life. Her dignity and self-worth were severely affected. With no support system, minimal finances and lack of transportation, life became overwhelming. While walking two miles to the nearest grocery store one day, Susan fell on the side of the busy roadway and broke her arm. The incident prompted several local church members and the HD social worker to become involved.

Networking with area agencies resulted in help and support for Susan. Church members built a trusting relationship with her and helped her to realize that she was not alone, and that she was still lovable and worthwhile. Susan felt she had found a new family.

When independent living was no longer a good option, Susan, only 43 years old, consented to placement in a long-term care facility. She told those advocating for her which facility and location she preferred and her wishes were respected. The staff soon realized Susan had much to offer others. They encouraged her to help in the dining room by handing out bibs to residents and assisting them in small ways. The staff introduced her to other residents of similar age and abilities. She made friends easily and soon had a boyfriend with whom she spent time, shared stories and participated with in activities.

Church members stayed involved, visiting and advocating for Susan's needs. They helped her obtain badly needed dental care, which improved her ability to eat properly and dramatically improved her looks. She smiled without covering her mouth with her hand. She felt pretty again!

Susan says she loves being at the nursing home. Her dignity and emotional well-being have improved dramatically. She is cared for, cared about, confident and safe.

## ***Conclusion***

In each of the above settings, we must advocate for the person with HD and educate providers and caretakers to ensure dignified treatment. Though a person may not be able to communicate verbally by the later stages of HD, they continue to understand emotionally and psychologically whether they are being treated with dignity and respect.

### ***Meaningful Social Interaction***

*Meaningful social interaction is composed of the ability to be seen by the group, the ability to express one's views, the ability to grow and to learn through social interaction and the ability to belong to the group. Most people belong to many overlapping social groups, allowing many different levels of social interaction. When left to fend for themselves in an unstructured environment, the person with HD most likely ends up homeless or in prison. Few are fortunate enough to be placed in a long-term care facility.*

### **Home Setting**

The person with HD participates as fully as possible as a family member.

### ***Ideal Scenario***

The person with HD is provided with well-supervised social contact through their existing social groups from school or church, or through newly created social groups. Mental stimulation is encouraged through hobbies such as assembling a scrapbook of special moments. Physical stimulation is accomplished through the provision of pleasurable activities, such as eating favorite foods. Friends and relatives remember them often with cards and letters. The person with HD is informed about medical strides to find a treatment and a cure for HD. Hope for a future and spiritual well-being are encouraged.

### ***Worst Scenario***

Maintaining social relationships is difficult. Transportation is not available and social contacts are non-existent. Denial is a major factor, complicating the transition into the later stages of HD. Frustration escalates to severe emotional distress and anger, which



decreases the person with HD's ability to cope with the progression of the disease. Family members frequently avoid the person. Authorities mistakenly confuse HD symptoms with intoxication.

### **Long-Term Care Facility**

The person with HD is cared for by staff that is well educated about Huntington's Disease. They are treated with utmost respect and appreciation. Social interaction is encouraged among the person with HD, the staff, other residents as well as family and friends.

### **Ideal Scenario**

The person with HD is placed in an assisted living home where they are comfortable and able to communicate. They are trained to use state-of-the-art communication devices. Staff and caregivers understand what the person is communicating. The person with HD feels comfortable and is more approachable by health care staff, family members and other patients. Every employee of the facility that interacts with the person with HD receives regularly scheduled education about HD. New employees are required to attend an educational orientation on the manifestations of Huntington's Disease, as well as palliative care. Employees are required to attend ongoing sessions about the most current developments on research, drugs, technology and care of the person with HD.

### ***Worst Scenario***

Long-term care staff is unaware of the care of people with HD. The long-term care facility is not equipped to accommodate the needs of a person with HD and the facility is understaffed. Minimal training for staff is provided and they exhibit no real understanding of the disease. Staff turnover remains a problem. The person with HD is isolated and has no social interaction. Suicide is a strong possibility.

### ***Hospital Setting***

The person with HD is admitted to the hospital and is cared for and treated with respect despite their minimal response.

### ***Ideal Scenario***

The family is allowed to be with the patient as much as possible. Staff keep the family informed of their loved one's condition and involved in all decision making. The family is allowed and encouraged to be with the patient and are kept informed of the dying person's changing condition; they understand what is being done for the person with HD and why; they are assured of the comfort of the person with HD; they are reassured and allowed to display emotions; they feel confident that the person with HD is fed, hydrated and rested; and they are assured that their decisions were proper and correct.

### ***Worst Scenario***

The person with HD is admitted to the hospital without the knowledge of their family due to estrangement or lack of notification. The hospital staff does not know the background

of the person with HD. Inappropriate decisions are made about life-sustaining treatment and the person dies alone.

### **Consider**

Marguerite is a beautiful woman who, before the onset of HD, enjoyed ballet, singing, modeling in fashion shows, occasional extra film assignments and participating in stage productions. She married at an early age and has two children. Although her husband loves her and is compassionate, the rest of the world has not been quite as sympathetic nor have people in her community understood the nature of her illness and the accompanying personality and behavior changes. In time, her friends avoid her as do her child's school staff, grocery clerks and even medical staff. It becomes clear that her family needs to orchestrate a support group around her.

Since it is no longer safe for Marguerite to drive, the family secures transportation from a county transportation service for the handicapped.

Her aunt invites her to join her church Bible study class and a women's group that has frequent activities throughout the year. These activities encourage meaningful social interaction and Marguerite feels renewed spiritual strength.

Family gatherings also change. Many now take place at Marguerite's home to make it more convenient, to promote her self-esteem and create happy memories for her husband and her children while she is still living at home. Family and friends respond to the family's invitation to help by orchestrating some of these gatherings. As Marguerite's abilities decline, family and friends adapt tasks and activities to meet her abilities.

These subtle but powerful changes produce a much happier and content person as Marguerite realizes self-worth and happiness. Being appreciated by others reaffirms her as a person. The quality of her life does not decline although it is altered to accommodate the changes brought on by Huntington's Disease.

### ***Conclusion***

A person's psychosocial adjustment to chronic illness is influenced by social factors.

People with HD experience a heightened need for interpersonal support. Furthermore, maintaining close connections with family and friends during the course of the disease is

more likely to enable the person with HD to cope more effectively with the disease than those who are unable to maintain such relationships. For the person with HD and their families, this becomes an increasing challenge, as the disease affects the mind. Therefore, it is imperative that the entire multidisciplinary team, including the families of the person with HD, are well informed of therapeutic interventions available to stabilize dramatic mood swings often expressed by people with HD. Behavior modification encourages and makes it easier for family and friends to be more attentive and to spend more time with the person with HD.

## ***Communication***

*The process by which information is exchanged. Several principles support communication. According to standard communication theory, any message involving use of words, gestures or body syntax has at least three levels - content, emotional expression and the relationships between sender and receiver. A person is classified as having a speech or language impairment when speech is unintelligible much of the time, or after being professionally diagnosed as having speech impairment that requires intervention. In people with Huntington's Disease, stuttering, impaired articulation, impairments in language, voice and swallowing are all part of the symptoms. Intervention with speech therapy is helpful but may never completely repair the disability.*

As the person with HD progressively loses the ability to speak, it is imperative that clear directives regarding critical preferences at the end of life are communicated well in advance. Directions should include care settings, living situations and services.

Technological equipment can make a tremendous difference in the quality of care for the person with HD at the end of life. A best practices scenario at the end of life for a person with HD includes communication among all of the involved parties. This includes caregivers, family members, health care professionals and those providing support from the community. Communication promotes comfort. With adequate technological equipment and education of nursing staff and practitioners, the person with HD can express needs and desires. Communication keeps the person with HD and their family informed of the person's health status and available palliative care options in preparation for the end of life.

Loss of speech is one of the most frustrating symptoms for the person with HD and the caregiver. Technological equipment permits communication and is especially important for people with HD with slurred or incoherent speech. Technology can assist people with disabilities, making a certain amount of communication possible and the end-of-life process more pleasant. State-of-the-art technology must be accessible to the person with HD.

### **Home Setting**

#### ***Communication Traps***

Establishing care goals requires expert communication skills and attention to “Communication Traps” that occur when health professionals are not cognizant of ritualistic patterns of interaction. A particular challenge occurs when the care goals of the person with HD shift from active treatment to palliative care. Five common communication traps can arise when health care professionals attempt to manage exchanges that are socially familiar and safe.

1. In the *We all understand each other* trap, individuals involved in care assume that other caretakers and providers share the same understanding of the goals of care and approach to treatment.
2. The *Let's all be polite* trap occurs when people with HD and families are cognizant of power imbalances between health care providers and themselves. They often comply with treatment and care approaches that are not comfortable for them.

3. *We should be positive* trap occurs when those involved in the care are concerned about taking away hope. They believe that the most appropriate way to manage an advancing illness is to fight it and focus on the positives.
4. *More of the same* trap occurs when the health care team, patient or family are unwilling to examine or accept a palliative care approach. Inappropriate use of active treatment and the transmission of false hope when palliation and comfort are called for lead to disappointment.
5. *Can we be clear?* is characterized by a need of involved individuals to obtain clarity, precision and open agreement about the plan of care. Over-concern with clarity creates difficulties when a situation is simply not certain. In some situations, people with HD and family members feel pressured to come to terms with care goals that they are not ready to accept.

To avoid these five communication traps, it is best to take a slower and more thoughtful approach to defining care goals.

Attention to signs of communication barriers and a willingness to explore different ways to interact with people with HD and families helps to avoid these impasses. Four strategies to facilitate care goals, “Lane Changes,” are offered with recognition that goal instability may combine seemingly disparate care goals at times.

1. *Anticipate roadblocks* - requires an ability to estimate whether a planned care goal is achievable or whether a change is needed.
2. *Gain consensus* - requires tact, diplomacy and respect for alternate points of view.

3. *Time the transition* - requires astute attention to the readiness of people with HD and families to consider changing from treatment goals aimed at modifying the disease to palliative care goals.
4. *Shift gently* - involves the use of communication skills that are honest but gentle and sensitive to the needs of people with HD and their families. At these times, caregivers need reassurance that comfort care remains paramount.

### ***Ideal Scenario***

The person with HD and caregiver are well informed about available services and resources. They are educated about the progression of the disease, and have made all the necessary plans and arrangements for the end of life. They are free of denial, fear and worry and see dying as a natural process.

### ***Worst Scenario***

The person with HD can no longer speak and does not have access to equipment that enables them to communicate. The person with HD, the caregiver and nursing staff become frustrated and their needs are not adequately met. The person with HD feels isolated and alone, unable to express their needs, is often misunderstood and does not receive proper care. Miscommunication builds, resulting in confusion, anger, inconsistent treatment, mistrust, conflict, potential for later guilt, isolation of the person with HD and their family, lack of preparation for advancing illness, loneliness, frustration, missed opportunities, inappropriate treatment plans, false hope, avoidance of in-depth discussion of treatment goals, and regret.



### **Long-Term Care Facility**

It is imperative that the staff be well informed about the progression of HD and the needs of the person with HD.

### ***Ideal Scenario***

The person with HD completed Advance Directives earlier in the course of their disease. Even though the person with HD is slowly losing the ability to communicate, the caregivers, nursing staff and medical practitioner are all clear on the person's needs, wishes and preferences. Staff take the time to effectively communicate with the person and family and attend to the person's basic needs: comfort, nutrition and a stable environment.

### ***Worst Scenario***

The person with HD is unable to communicate. As a result, the person with HD, caregiver, nursing staff and medical practitioner become increasingly frustrated. The person with HD may not get enough food to satisfy a caloric intake requirement. There may be pain from bedsores, discomfort from too much noise or light in the room. They may be positioned uncomfortably, or they may not be able to communicate their wish to see a specific person to the nursing staff. The facility may be understaffed or staff may not have time or patience to listen. Care goals are not identified and "Lane Changes" result in chaos and confusion while "Communication Traps" take over.

### **Hospital Setting**

When the person with HD gets to this point, they and their family would benefit from knowing that the health care system will deliver reliable and humane care giving. Some hospitals have special advanced illness programs that coordinate care, maintaining a consistent care coordinator for the patient.

### **Ideal Scenario**

In the hospital setting, interpersonal or internal conflicts are resolved, and the challenges of dying with honesty, caring and commitment are achieved.

### **Worst Scenario**

The person with HD reaches the advanced stages of the disease without preparing for the progression of the illness. Their needs are not stated and they are unable to make their needs and desires known to those around them. This lack of communication fosters an overall feeling of frustration, preventing the person with HD from dying well.

### **Consider**

Mandy is a young lady with Huntington's Disease. Although she is only 25 years of age, she began showing symptoms at the age of 11. After a formal diagnosis at the age of 17, her attitude was positive. She stayed on top of the latest HD research and dealt with her limitations. She participated in whatever she could and enjoyed a peaceful environment created to accommodate her lifestyle and needs. She participated in Karaoke with the family as backup singers and she danced from a wheelchair with the help of family and friends.

Mandy is now at Stage IV of Huntington's Disease. Her family foresees a transition to the next level of the disease in the near future. Although her speech is difficult to understand, Mandy can still communicate her needs and wishes. When the family misinterprets what she is trying to communicate,

they assume a loving and playful attitude in trying to decipher her words rather than expressing frustration or hopelessness. Mandy is usually the first one to start laughing at their hilarious interpretations! She currently receives speech therapy. The family is now looking ahead to when even limited communication will be impossible. They have contacted a computer company that offers equipment to assist disabled people with communication needs.

The family has talked about Advance Care Directives. They have contacted an elder law attorney to draft the necessary documents. This has all been done in a positive and natural course of life spirit. Mandy would like to be placed in an assisted living facility when a home setting is no longer adequate for the care that she requires. She feels that she will be able to maintain a greater sense of dignity and care in a professional care setting.

The family has, as a group, selected burial plots and discussed the funeral arrangements that they would like to have. Mandy, being of young heart, communicated her desire to wear a red, fitted, spaghetti strap dress for her funeral. Her mother assured her that she would get her dress and the entire topic became a festive discussion, such as planning a party.

One of the most comforting and reassuring communications from family and friends to Mandy is the knowledge that she is loved and that she will be well taken care of and never left alone.

### ***Conclusion***

Inadequate time, space and funds are allocated for communication. Lack of discussion about care plans demonstrates denial on the part of the person with HD as well as the family and health care providers. Early preparation for the inevitable loss of speech and cognitive ability can be made by preparing all legal documents, Advance Directives, instructions for care and services at the end of life. Equally as important is early training in hi-tech equipment to assist a person with HD to communicate when they have lost the ability to speak.

## **Comfort**

*From Late Latin confortare, to strengthen greatly, affording or enjoying contentment and security. Affording or enjoying physical comfort; free from vexation or doubt; free from stress or tension.*

*Discomfort in people with HD includes a feeling of deprivation from not being understood or able to express themselves, from not having services or resources for their many needs. Feelings of discomfort range from any or all of the following: loneliness; isolation; hunger; pain; loss of dignity, respect, mobility, speech, control of personal finances and friends.*

Simple amenities like the presence of tissues, chairs, blankets, coffee, water and a phone, and general attention to the aesthetics of a room, can contribute substantially to the person with HD's and the family's sense of well-being and peacefulness.

## **Ideal Scenario**

A safe and peaceful environment with knowledge, time and patience to take proper care of the person with HD is the most desirable setting. There are several areas of care that are enhanced in an ideal setting. They include home comfort, nutrition, exercise, entertainment and pain management.

1. Mobility: Moving the person with HD from one position to another is extremely difficult for the caregiver. The home is equipped with a wheelchair that can prop the person with HD into an upright position. To protect the caregiver from injuries and unnecessary strain, the home is equipped with hoisters that lift the person with HD and haul them to the shower, bathroom etc., using a remote control. This arrangement makes

the task much more comfortable and enjoyable for the person with HD and the caregiver. Furniture designed for their special needs is available.

As an example, *Home Technologies 1999* is a movement toward the *Home as a One-Stop Healthcare Shop for Retrofitting*. Under this concept, the typical living room is a one-stop health care shop and the coincident safety concerns have been designed along particular guidelines emphasizing simplicity of design and ease of use.

2. Nutritional Education: Proper nutrition is vital not only to the physical well-being of the person with HD but also necessary in making them comfortable. It is essential that long-term care facility staff be knowledgeable that the amount of calories required by a person with advanced HD is greater than that of the typical resident of a long-term care facility.

3. Humor: Social interaction and humor, which serve to boost comfort level, are essential components of daily therapy for a person with HD.

4. Company: Especially at the end of life, company is comforting to the HD patient, even with minimally interactive activities such as watching a favorite movie.

5. Music: Listening is a form of relaxation.

6. Exercise: Exercise supports well-being and relaxation.

7. Comfortable living environment: Fresh air, good company, good lighting, a comfortable setting free of obstacles that can cause accidental falls all help to create a comfortable environment.

8. Pain Management: Pain management supports respect for and dignity of the patient. The person with HD must communicate their needs relative to pain management. To do this, they may need technological tools that offer them the ability to communicate when they can no longer speak and the services of a health care practitioner well versed in non-verbal communication.

9. Multidisciplinary Team: This approach offers the person with HD expertise and quality care.

### ***Worst Scenario***

In pain, the person with HD is not attended to nor understood. Discomfort is such that they would rather die but cannot communicate even that to anyone.

### **Consider**

Richard is a 42-year-old man in the last stages of Huntington's Disease who lives in an assisted living facility. He has lost his speech but family members know his likes and dislikes in food, music and entertainment.

His wife, Mary, suffers silently as she watches her husband slowly wither away. Though she risks hurting the feelings of others, she is unabashed about letting family members and friends know her wish for them to interact with her husband in a loving and compassionate way rather than with pity and distance. His teenagers visit several times during the week and always greet him with a big smile. They massage his arms and legs and help with his grooming, which Richard finds comforting, physically and emotionally. The staff is well educated about HD and knows how to handle difficulties associated with the disease such as choking and breathing problems. Mary reads articles that she knows Richard would find interesting and his children bring music that they know he appreciates. The children sit by his bed and play board games. Just the sound of their voices and laughter bring a smile to his face.

While his caloric intake was extremely high at one time, he lost his appetite in the late stages of the disease. The family and facility staff keep him well hydrated and have plenty of creamed soups and soft food available. He does not appear to be in pain but he does complain about something or other every once in a while. Medication is readily available for the treatment of discomfort.

Because Richard is now at the end stages of HD, his family spends most of their time with him. The facility has arranged to have the necessary items available for the families' comfort as this is now their home away from home. Richard feels a sense of love, peacefulness and comfort. He feels lucky to be surrounded by a loving family and so many loving and caring friends.

### ***Conclusion***

A training program to improve the knowledge and skills of providers and caretakers in the home, as well as in long-term care facilities and hospitals, is needed. In the past, health care professionals were ill prepared to assume an active role in the care of terminally ill people at home. The health care community has a responsibility to educate itself and others about the dying process and the specific needs of people with HD at the end of life. There is an urgent need to improve end-of-life care as health care delivery continues to shift care to the home. To improve care, health care professionals need educational materials, tools and resources. Important to delivering good end-of-life care in the home is recognizing the importance of end-of-life care in home care settings; discussing principles of pain and other symptom management at the end of life; listing communication strategies with dying patients and family members in home care settings; and discussing nursing interventions about death in these settings. This also applies to long-term care facilities and hospital settings.

Finally, it should be mandatory for elected officials to visit assisted living facilities, long-term care facilities and hospitals on a regular basis (at least once a year) as well as to become fully informed and prepared to allocate federal, state and county funds to meet the needs of critically ill people in this country.



## ***Safety and Order***

*Safety is a state of freedom from danger, injury or damage. In the context of end-stage HD, safety considerations include the potential for physical injury related to cognitive, behavioral and motor aspects of the person's disease as well as the possibility of sexual, emotional, verbal or physical injury inflicted by others.*

*Order means many different things, including a fixed or definite plan, a sequence or arrangement of things or events, a state of peace and serenity and a state or condition in which everything is in its right place and functioning correctly. For the person with late-stage HD, order includes a constant living environment with predictable daily routines, good general health as well as personal order in the form of good hygiene and a clean immediate environment. In many ways, order is easiest to achieve at home where the person with HD's belongings, history, personal comfort and security are familiar. External disruptions are easier for caregivers to prevent or control.*

## **Home Setting**

### ***Ideal Scenario***

In the ideal circumstance, the person with HD is surrounded by familiar objects, sounds, smells and foods provided by loving family members. Family members pose no risk of sexual, verbal or emotional abuse. Appropriate assistive devices, chairs and beds minimize the risk of physical harm and a visiting nurse and aide assist the family in problem-solving and providing personal hygiene. Order in the environment is easily preserved by frequent cleaning, replacing items that are damaged or worn and by maintaining a quiet, peaceful surrounding. Paid assistants or able members with appropriate skills perform cleaning and other heavy chores.

### ***Worst Scenario***

There is no care provider. The person with HD lives in an unkempt house, with limited or no interaction with outsiders. They deny having HD and threaten to shoot anyone who suggests otherwise. The person with HD may threaten to shoot himself or herself. The person with HD falls, eats and bathes haphazardly and drives in a manner that others feel is unsafe.

### ***Solutions***

It is difficult to find solutions for the person with HD who denies symptoms. Adult protective services and county social workers can help document the situation and eventually family or medical providers can pursue commitment hearings if the situation becomes dangerous. Medications help reduce aggressiveness, paranoid ideation and underlying depression that lead the person with HD to resist help.

In the not-so-bad scenario, where caregivers are available but need help, in-home services are increasingly available, either to provide help with household chores or to help with personal care. Physical, occupational and speech therapy assessments help the family provide the best and safest care. These assessments may need to be repeated at intervals, as disease symptoms change over time.

Caregivers must be aware of the unique aspects of HD as they look for assistive devices and programs. Involuntary movements can lead to bruising of limbs and traumatic joint effusions and self-induced injuries, often on the face. Poor control of head and trunk

create a high risk for falling out of bed, sliding out of wheelchairs and recliners, strangling on restraints or being caught in awkward positions under railings, chair arms or legs, or other furniture. Those who are ambulatory are at a high risk for significant injuries from falls. Impaired voluntary control of hand and arm movements increase the risk of injury when the person uses eating or grooming utensils, smokes, or holds or carries heavy items. Dementia and impulsive behavior compound the risks of denial, while combative or aggressive behavior and dementia complicate the caregiver's ability to exert control over an unsafe situation. In addition, physical disorder is common and almost universal in the form of poor oral, body, menstrual or excretory hygiene, unclean clothes, and a sloppy or unclean immediate environment. A chaotic living environment compounds a sense of disorder. Hospice care, which can be provided in the home, may be helpful in imposing some order in the terminal stages of HD.

### **Long-Term Care Facility**

#### ***Ideal Scenario***

The staff thinks of the facility as the home of the person with HD. They have familiar and personal possessions and adequate space, increasing their sense of control and order.

Personalized arrangements are made for sleeping, seating, hygiene, feeding and day activities to accommodate the person with HD's specific motor or cognitive problems.

Needs are reassessed regularly as the disease progresses. A regular daily routine creates a sense of order. A stable set of health professionals works with the person with HD, and similar events occur at similar times each day or week. Issues such as falls, unsafe smoking and terminal care directives (e.g., feeding tubes) are discussed in advance of

progression to the next stage with the person with HD and family members. The family is updated about indicators in the progression of the disease.

### ***Worst Scenario***

The staff is unfamiliar with HD and interprets involuntary movements as hitting. The unattended bed-bound person with HD wriggles between the mattress and the headboard, strangling himself or herself. Overworked staff feed the person with HD quickly and then report that they vomit after meals. Falls are frequent and skin tears and bruises are poorly documented and treated. There is a question about an episode of sexual abuse between a staff person and the person with HD. They smell of urine and have crusted food on their face and clothes. The facility has no funds to replace the chair or bed that broke when the person with HD flung themselves into it last week. The drink stains on the wall are still present months later. The family sues the long-term care facility after the person with HD falls and sustains a subdural hematoma and fractured collarbone.

### ***Solutions***

The team approach is critical, including input from the neurologist or HD specialist, the primary care or long-term care facility physician, everyone involved in the care of the person with HD as well as the person and the family. Frequent reassessment of the person with HD's problems and needs is necessary.

## **Hospital Setting**

### ***Scenarios***

Like a hotel, a hospital is a place to visit but not to live. Personal order is relatively easy to establish in a hospital as patients are more or less molded to the institution's routines (e.g., they wear hospital gowns and sleep on identical beds whose linens are changed daily). Devoid of personal belongings, rooms are cleaned daily. However, the patient exerts no control over their own order in the hospital but rather, is at the mercy of test schedules and treatments. The daily routine may be disruptive, unfamiliar and upsetting.

Personal safety is difficult to achieve in the hospital as individualized approaches to seating, bedding, positioning, restraint, feeding and bathing are unlikely to be available. Lack of familiarity of hospital staff about the unique aspects of HD complicates the provision of excellent care. The symptoms of the person with HD are likely to be exaggerated in the context of a critical illness.

### ***Solutions***

Solutions are difficult to attain in any practical way since no hospital will experience a high enough volume of people with HD to warrant a commitment of significant time or staff to specialized HD care. Developing and marketing easy-to-read and/or easy-to-access literature about practical issues of HD care to hospitals, long-term care facilities and nursing departments would be beneficial to people with HD and their families.

## **Consider**

Leslie, 45, is admitted to a long-term care facility with Stage IV HD. She has severe chorea, falls multiple times daily, and refuses to let male nursing aides help her with personal care. She frequently falls out of her single bed and she often screams, especially when the resident across the hall plays loud rock 'n' roll music.

Nursing staff has a care conference with the family. They learn that Leslie was a hairdresser prior to becoming disabled from HD, as well as a violinist with the community orchestra for many years. As a result of the conference, Leslie is moved to a room down the hall, away from the noisy rock music. The family purchases a queen-sized bed for her. She is given a Q-foam chair to sit in during the day and a hockey helmet to protect her head when she falls. Violet, an aide who also works as a hair stylist, trims her nails every two weeks to help prevent Leslie from scratching herself. The physician treats her with an antidepressant and an anti-anxiety medication, as needed. The speech pathologist evaluates her swallowing and recommends a change in the texture of her foods. The recreational therapist engages her in an ongoing art project that the residents on the unit are undertaking.

After a few weeks, Leslie's weight increases by 10 pounds and her movements have decreased. She smiles readily and shows everyone her contribution to the quilt project. She won't let anyone near her Q-foam chair. Although Leslie still lurches when walking and frequently falls, she no longer falls out of bed.

### ***Spirituality***

*Spirituality is a sense of one's place and comfort in the larger universe. When we cannot control the environment around us or do not understand it, our spiritual system provides a framework and an explanation. Although spirituality is often associated with organized religion, it also exists outside of a formal religion or religious place.*

HD inhibits one's spirituality primarily by causing dementia, which reduces a person's capacity for abstract thinking, and through its effects on communication, ultimately results in a mute state. It is likely, however, that people with HD, even when mute and incapable of abstract thought, can recognize and appreciate familiar rituals, songs, pictures and prayers.

### **Home Setting**

#### ***Ideal Scenario***

Presumably, a person's spiritual needs are best met at home, surrounded by people who know them well and by objects that are familiar and reassuring. Members of the religious community come frequently. Family and friends take time to talk with the person with HD, review scrapbooks, ask questions and wait for answers or responses, even though the person is almost mute.

#### ***Worst Scenario***

It has been several years since the person with HD attended religious services, even though it was once a big part of their life. Conversation in the home revolves around

custodial care needs and nobody has or takes time to ask anyone how they are feeling. No one talks to the person with HD except to ask about bathroom or sustenance needs.

### ***Solutions***

In his book *When Bad Things Happen to Good People*, Rabbi Harold Kushner makes the point that religion provides the framework to move forward and to continue living after a tragic loss. Caregivers should be encouraged to attend to their own emotional and spiritual needs. Religious communities can be of great assistance to the family through day programs, respite care and in-home help.

### **Long-Term Care Facility**

Providing for the spiritual needs of the person with late-stage HD challenges even the best of long-term care facilities. Spiritual care does not fit well into the medical model of care that underlies nursing care plans. Residents come from a variety of social, ethnic and religious backgrounds. In the absence of family members, it is difficult to determine their spiritual needs. People with HD benefit when their family is encouraged to inform the facility staff of special beliefs, comforts or desires and to include them in special family events. Families with a strong religious affiliation are encouraged to use facilities that have matching religious affiliations. Residents also benefit from being surrounded by things that have special meaning to them such as photographs, household items and music. Hospice programs typically include a component of spiritual assessment and care, which helps the family and the person with HD in the terminal stage.



## **Hospital Setting**

The hospital probably represents the antithesis of spirituality, in its relentless quest for scientific and clinical explanations and treatments for health problems. Even in the hospital, though, there is a growing awareness of the need to provide culturally and spiritually sensitive care. Patients benefit from the use of interpreters, having the family present as much as possible and thoughtful physicians and nursing staff. Hospitals typically have pastoral care available and most encourage the patient's own clergy to participate in supporting the patient and the family.

## **Consider**

Dorothy, age 45 with Stage IV HD, has been in the long-term care facility for six months. As the Christmas holidays approach, Dorothy's screaming and irritable behavior increase. She won't even talk to Violet, her favorite aide.

The nursing staff contacts the family to describe the behavior change. During the conversation, the daughter reminds the nurse that, of course Dorothy is upset. Dec. 4 is the date that Dorothy's son was killed in a motorcycle accident three years ago. Since that accident, the holidays have been difficult for the whole family. The nurse is surprised, as nobody has mentioned this son before.

The chaplain meets with Dorothy several times to discuss her son, his death, the holidays, her religious beliefs and her holiday traditions. The daughter, who hadn't really realized that she could take an active part, arranges to take her mother to the cemetery to visit the grave. She brings pictures for her mother's room and she arranges to bring her mother home for Christmas Day. Dorothy's behavior improves dramatically.

### ***Enjoyment, Entertainment and Well-Being***

*Well-being is a feeling of happiness, comfort and contentment. Since these internal feelings may be impossible to measure in a person who is mute, enjoyment and entertainment have been included with well-being in a single domain. Access to entertainment and exposure to items or events that a person was known to have enjoyed in the past can be measured, even if it is difficult to determine whether the person experienced well-being as a result of the exposure.*

### **Home Setting**

#### ***Ideal Scenario***

The person with end-stage HD is surrounded by items that please them, favorite music, pictures, colors, clothing and television shows. Food preferences and tastes are taken into consideration when meals are planned. The person with HD is included in family activities such as picnics and holiday celebrations. When they are no longer able to attend these activities, the family brings the activities to the person in an appropriate manner. The minister or representatives of the congregation stop by when they are able, as do friends and relatives.

#### ***Worst Scenario***

A financially strapped, overworked daughter, who has a baby and is working two jobs, cares for the person with late-stage HD. Her boyfriend is available most of the time to watch the baby and the person with HD. A sister is also staying in the home for a while. Meals occur somewhat randomly. Nobody pays much attention to the person with HD, who fortunately spends almost all their time watching TV and smoking. As long as the caregivers give the person with HD cigarettes, they behave, and although there are

cigarette burns all over their clothes, hands and the furniture, the house has not caught fire yet. The person with HD has not been out of the house for months, and most of the conversation in the house consists of someone telling someone else to do something.

### ***Solutions***

Once again, the home caregiver must be recognized as a stressed individual, with support made available in any way possible. Day programs provide a safe haven, structured activities and nutritional meals for the person with HD while allowing caregivers some respite. A public health nurse can assess the safety and appropriateness of the home situation and help a disorganized family establish some order. Often the biggest hurdles, however, are financial and logistical. While programs support the medical and nursing care needs of a chronically ill person, few programs provide support for a family to go to the fair, a movie or a restaurant. Social connections outside the home for a person with HD may be quite low on the priority list for a stressed care provider. Programs such as the Make-A-Wish Foundation provide a one-time opportunity for ill individuals and their families to have a special, memorable entertainment.

Many individuals are willing to help a family with HD, but are not certain as to what they can do to help. They can be encouraged to take the person with HD out for a ride, a visit to a park, a movie or other pleasurable activities. Caregivers need to be encouraged to accept this form of help as a way to provide enjoyment for the person with HD and as respite for the caregiver.

## **Long-Term Care Facility**

### ***Ideal Scenario***

The ideal situation in a long-term care facility is not different from the ideal home situation. The person with HD's preferences for clothing, music, food, activities and personal care are known and attempts are made, whenever possible, to accommodate these preferences. Daily entertainment and exercise programs are available and varied so as to address the interests of different individuals from day to day.

### ***Worst Scenario***

The worst-case scenario is again similar to the worst-case scenario at home. The person with HD is only one of 25 residents that require staff care. Because of unusual and sometimes violent behavior, the staff tends to leave care of the person with HD until the end of the shift. Nobody ever asks what the person with HD wants, since they can't talk anyway. The TV is turned up loud to the roommate's favorite show and stays on for hours, while the person with HD is trapped in the bed unable to communicate or move.

### ***Solutions***

One solution to this issue is to include in the care plan a list of patient preferences in a number of categories, such as food, clothing, colors, styles of music, TV shows, hobbies or activities and so on. It would then be possible to document how many of these preferences are present in the room or offered to the person with HD during a week or month. Facility staff ought to be alert to conflicts between roommates or between people with HD and staff members. Documentation of behaviors that suggest a lack of well-

being, such as screaming, wailing or hitting must be followed by an assessment and plan to improve the environment of the person with HD.

### **Hospital Setting**

Patient entertainment, enjoyment and well-being are not major priorities in the hospital, where acute medical care is provided. People with HD deserve the standard care to which all hospitalized patients are eligible, with standard means to address any grievances.

### **Consider**

Dorothy, who was described earlier, is 45 with Stage IV HD. She has made plans to go home with her daughter for Christmas Day. Violet, her favorite aide, who also works as a hair stylist, asks whether she would like to get her hair cut and styled, her nails done, her face made up and some new clothes for the special day.

Dorothy and Violet spend two weeks planning the special day. The recent HD fundraiser has given each HD resident in the facility a \$25 gift card to the local department store. With that money and \$25 from the resident's own funds, Violet purchases an inexpensive Christmas outfit, some lipstick, nail polish, and a stuffed toy animal for the resident's granddaughter. Ten days before the big day, the recreation therapist helps her wrap the gift and make a card to go with it. A week before the big day, Violet washes Dorothy's hair, cuts and styles it. Five days before the big day, they try on her new clothes. Two days later, Violet paints Dorothy's nails, both her hands and her feet, cherry red. On the big day, Violet makes sure that Sally, the aide on that morning, helps Dorothy with the blush and lipstick.

Dorothy's daughter almost faints when she sees her mother, who has the biggest smile ever on her face!

***Nutrition***

*The act or process of nourishing or being nourished; the sum of the processes by which a subject takes in and utilizes ingredients to grow their body and/or maintain their health.*

Nutrition is particularly important because people with HD require an unusually high caloric intake to maintain body weight. Although a recent study showed that most people with HD gain body weight in an early stage of the disease, maintaining or even gaining weight is beneficial to people with HD, particularly in the late stage of the disease.

Factors involved in weight loss in people with HD include:

- Increased metabolism - high calorie consumption,
- Dysphagia,
- Poor motor skills for eating,
- Anorexia,
- Depression,
- Vomiting/Constipation/Absorption,
- Poor oral hygiene,
- Lack of attention to calorie intake by caregivers,
- Side effects of medications, and
- Inability of patients to get food due to psychological, physical and financial constraints.

**Home Setting**

The caregiver, visiting nurse and social worker play important roles in ensuring adequate nutrition. The caregiver must be keenly aware of nutritional problems associated with the

disease. The physicians and nurses ought to discuss problems with feeding, dysphagia, oral/dental hygiene, aspiration, depression, anorexia and other gastrointestinal complications in an earlier stage of the disease with the person with HD and caregiver. Caregivers and social workers assess the person's psychological, physical and financial ability to get enough food at home and provide appropriate help, if necessary.

### ***Ideal Scenario***

The person with HD, caregivers, visiting nurses and aides are aware of potential nutritional problems associated with the disease. The weight of the person with HD is monitored, at least every two to three weeks, under the supervision of a dietician. If a significant weight reduction is noted, the physicians and nurses assess the cause of weight loss, such as decreased caloric intake, malabsorption and increased metabolism. When decreased caloric intake occurs, motor coordination, dysphagia, oral/dental hygiene, depression, anorexia and current medications are assessed while physical therapy, occupational therapy, speech pathology evaluation and psychiatric intervention are provided as needed. Gastrointestinal diseases that cause malabsorption, anorexia, constipation and abdominal discomfort are explored, if appropriate. Caregivers and social workers ensure that enough food is available in the home. Other concomitant conditions including endocrinological disorders, such as diabetes mellitus and hyperthyroidism, chronic infections and malignancy are evaluated, if necessary.

### ***Worst Scenario***

The person with HD is unable to buy food due to physical, mental or financial problems and no one brings food to their home. Even if enough food is available, the person with HD and caregivers are not aware of the problem of weight loss or, if they are, they do not seek medical attention. When they finally visit a health care professional, the professional does not know that weight loss is a frequent problem in HD and does not take appropriate action. Malnutrition, aspiration and other complications threaten the life of the person with HD.

### ***Long-Term Care Facility***

The role of the staff increases to prevent and identify nutritional problems. Education of staff and physicians about weight loss in HD is important.

### ***Ideal Scenario***

The staff is well informed about nutritional problems of HD. The person with HD is weighed regularly and a dietician provides useful input. Signs of poor nutrition are promptly spotted and appropriate medical, dental, nursing and allied health expertise is obtained from health care professionals in a timely manner. The cause of weight loss is defined and appropriate treatment is given.



### ***Worst Scenario***

The facility staff has no understanding of the nutritional problems of HD and weight loss is overlooked. Necessary actions are not taken. The person with HD continues to lose weight and becomes malnourished with various medical complications.

### ***Hospital Setting***

Malnutrition is one reason for acute hospitalization of people with HD. Consultations with a dietician, speech pathologist and occupational therapist are obtained as needed. Nursing staff must help people with HD who have poor eating skills. The attending physician should offer symptomatic pharmacotherapy for anorexia, depression and constipation. Appropriate evaluation for malabsorption and other gastrointestinal diseases ought to be obtained, if appropriate. When people with HD are hospitalized with any acute medical problems, body weight must be monitored regularly, and if malnutrition is detected, appropriate treatment must be given to them. Gastric tube feeding is an important option, and the person with HD ought to prepare a living will and power of attorney for the decisions regarding this option in an earlier stage of the disease.

### ***Ideal Scenario***

On admission, the nutritional status of the person with HD is assessed. Based on the living will and power of attorney, the triage/code status is determined. A well-informed team of professionals, including the attending physician, nurses, dietician, speech pathologist, occupational therapist, gastroenterological consultant and pharmacist,

implements preplanned procedures for nutritional treatment. Psychologists, counselors and clergy provide psychological support.

### ***Worst Scenario***

The person with HD does not have a living will or power of attorney, nor were their desires discussed with family members earlier in the course of HD, when communication was easier. The nutritional assessment is either not performed or is delayed. There are no specific plans for nutritional treatment. The attending physician and nurses are unfamiliar with HD and fail to recognize nutritional problems associated with the disease.

Consequently, no consultations are requested. When consultants are called in, they have a poor understanding of the disease and the unique nutritional needs of the person with HD are not adequately addressed.

### **Consider**

Sara has just had her 48<sup>th</sup> birthday. It has been seven years since her son, Jack, first noticed something was wrong. Though everything was going well at the time, his mother simply became depressed. She gained some weight and blamed the “happy pills” her doctor gave her for depression. A few years later, she started showing fidgety movements. Since then, she has experienced rapid progression of the disease and can no longer walk unassisted. She started choking and when that increased in frequency, her neurologist ordered a modified barium swallow test to assess swallowing ability. A speech pathologist was also asked to evaluate the patient. As results revealed a significant problem with Sara’s swallowing, the speech pathologist provided several techniques to improve swallowing and eating. A dietician made further recommendations for daily menus. Although Sara followed the instructions and did well, her son noticed that she continued to lose weight. The dietician added high calorie supplements to the diet. Sara initially liked the taste of this canned supplement and she gained some weight. Her doctor was still concerned.

Though Sara has a master’s degree in mathematics, her thinking and memory began to show signs of deterioration. On one clinic visit, her doctor talked

about Advance Directives and a durable power of attorney. It was shocking to both Sara and Jack, when she had difficulty understanding the concept of Advance Directives and durable power of attorney because both thought that Sara was still doing well intellectually. A few months later, Sara suddenly refused to take the high calorie supplement, stating that someone was trying to poison her. She began to show other uncharacteristic behaviors such as often becoming angry with Jack. She ate very little and before long, her weight dropped to 95 pounds.

On her 48<sup>th</sup> birthday, Sara choked on soup. By the next day she developed a high fever and was admitted to a nearby hospital. Her chest X-ray showed aspiration pneumonia. To maintain her blood oxygen level, she was placed on a respirator. Fortunately, antibiotics gave her a quick recovery from pneumonia and she was out of the Intensive Care Unit after four days. After her doctor consulted with the dietician, speech pathologist, gastroenterologist, and otolaryngologist, he recommended that a gastrostomy for tube feeding be performed, based on the risk of recurrent aspiration pneumonia and nutritional concerns. Sara refused tube feeding. While Jack knew that tube feeding was necessary, he regretted not having an Advance Directive and durable power of attorney in place for Sara. After many arguments with Jack, Sara finally agreed to the gastrostomy. After tube feeding began, her weight increased to 120 pounds.

### ***Conclusion***

Nutrition is an important factor at the end of life for people with HD because they suffer from progressive weight loss. Multiple problems affect intake and consumption of nutrition, especially in the late stages of the disease. A proactive approach will alleviate problems. The person with HD would do well to prepare for the time when they can no longer make their desires known, through a living will and power of attorney. In these documents it is important to address nutritional care at the end of life. Availability of services and financial support is critical and best explored in the early stages of the disease. Education of involved professionals is essential for good nutritional care of people with HD.

## ***Functional Competence***

*Possessing and using the ability to integrate thinking, feeling and behavior to achieve tasks and outcomes valued in the host context and culture. In the setting of HD, the following are relevant:*

- *Social competence (for social tasks and outcomes);*
- *Professional competence (for professional tasks and outcomes); and*
- *Driving competence (for driving tasks and outcomes).*

It is important to note that different competencies are required and valued in different contexts. Behaviors that are dysfunctional and disapproved of in one context, might be functional and approved of in another (e.g., anger outbursts are more easily tolerated in a home setting than at work or while driving). Depending upon settings, cognitive, as well as emotional, psychiatric and motor capabilities influence competence in HD.

What parameters are used to determine whether people with HD are competent in each context noted above? No literature was found on this subject except for that which related to legal competence. For the general population, there are some guidelines that may not be directly applicable to people with HD because of the complexity of the disease.

- *Social competence* has a direct impact on one's autonomy in daily life and is a key factor that defines the end of life. It is a critical factor in the institutionalization of patients to chronic care facilities.
- *Professional competence* is directly relevant to disability and social security issues. The medical assessment of professional competence provides critical information for disability. However, because of a poor understanding of HD by medical assessors, the disability is often underestimated or overlooked, especially when cognitive or psychiatric problems are the first to appear. The authorities that make the final decisions regarding disability must be educated about HD.
- *Driving competence* is a complex social problem, particularly in some geographic areas of the United States where public transportation is inadequate. A serious dilemma between public safety and the patient's autonomy and independence must be recognized. There has been no systematic effort to define driving competence in HD.

Different states have different regulations. For example, since 1988, California has required physicians to report dementia cases to the local Division of Motor Vehicles (DMV). Based upon the medical evaluation, as well as vision, knowledge and a 30 to 45 minute driving test, a decision is made to revoke, suspend or limit driving privileges versus placement on probation or unlimited privileges. However, the frequency of reporting has been less than expected. Six other states have statutes that mandate that medical conditions that impair driving be reported. In 10 other states, medical reporting is not mandatory but physicians who choose to report an impaired patient (in good faith) are protected from litigation by the driver in question and the report is considered confidential. Driving competency is, however, an issue that arises before the final stages of HD.

Different levels of competency must be considered in different settings. There are no effective medical means to maintain competency of the person with HD although providing a good environment, according to their needs, may delay the loss of competence. In each setting, however, it is important to recognize that maintaining all competencies for as long as possible may not always be in the best interest of the person with HD, especially when it becomes a significant burden. Rather, having insights about competency, preparing for losing competency, recognizing the loss of competency and implementing contingency plans for that loss are keys to alleviating problems. Proactive solutions are much more effective than passive ones. If the competency of the person with HD is already in question and they lack insight, the next important steps are to identify instruments to assess competence and to educate people who are involved in decision making of competency status.

### **Home Setting**

Professional and driving competencies are first questioned, hinting at the beginning of the end of life. When social competencies are lost, people with HD are definitively nearing the end of life.

### ***Ideal Scenario***

Before the person with HD loses their professional competence, they are fully informed about available options through various information resources and consultations with social workers, financial advisors, legal experts and psychologists. These experts are fully informed about problems associated with HD. When the person with HD's professional competency is questioned, they maintain their insight into the situation and implement the option that they have already selected. The person with HD seeks medical and legal advice for their driving competency and voluntarily terminates driving privileges when it becomes necessary. The person with HD also prepares a living will and power of attorney with legal consultants and a trustworthy companion. To prepare for social incompetence, they consult with social workers about assisted living, nursing homes and other chronic care facilities. Caregivers or professional home help are available to assist them with household chores and the activities of daily living.

### ***Worst Scenario***

The person with HD is in denial of the disease and has no desire to prepare for competency issues. There are no caregivers to address competency issues with them. Resources such as social workers, financial advisors, legal experts and psychologists are not available or they do not understand the needs of people with HD. Consequently, the person with HD is fired from work and becomes unemployed and without disability income. They keep driving and cause motor vehicle collisions. Social isolation and lack

of income force the person with HD to become homeless. There are no options to mitigate the consequences. No living will or power of attorney is prepared.

### **Long-Term Care Facility**

Professional and driving competency are no longer issues. People with HD in long-term care facilities have already lost social competency and many have lost legal capacity as well.

### ***Ideal Scenario***

The person with HD is in a long-term care facility where the nature of HD is well understood by the entire staff. The person with HD maintains insight about the limitations of their competency. Appropriate professional help is readily available, including a social worker, psychologist, financial advisor, legal expert as well as religious support. The person with HD has made financial and legal arrangements for secured income prior to the admission to the facility. A living will and power of attorney have been prepared.

### ***Worst Scenario***

The long-term care facility has never had a person with HD and the staff has limited understanding of the disease. Problems are handled in a crisis management fashion without identifying real problems associated with HD. Accessibility to a social worker, psychologist, financial advisor, legal expert and religious support is limited. If these professionals are available, they are poorly informed about HD. The person with HD lacks insight about their competency and refuses to accept professional help. Family and

friends are not available to address competency issues. A living will and power of attorney have not been considered.

### **Hospital Setting**

The situations and needs are similar to the long-term care facility setting. Because the hospital setting will likely involve life-threatening situations, the issue of social competency is further diminished while a living will and a power of attorney become critical. At the very late stage, people with HD are not expected to be competent, although some maintain partial competency for some tasks.

### ***Ideal Scenario***

The person with HD has prepared a living will and power of attorney. The hospital staff understands the needs of the person with HD. Family and caregivers agree about terminal care of the patient.

### ***Worst Scenario***

Family members or caregivers disagree about terminal care, particularly the triage status of the person with HD. There is no living will or power of attorney. The hospital staff lacks understanding of the needs of the person with HD.



## **Consider**

Jack, a 52-year-old corporate accountant, had been experiencing fidgety movements for five years, although the disease had not affected his job performance. His boss, who knew that Jack had Huntington's Disease, was supportive. When Jack began to make minor mistakes at work, he sensed the beginning of cognitive impairment caused by HD. He became depressed. His colleagues noted that he was unusually quiet and socially withdrawn. Jack lost his appetite and ability to sleep and he started drinking. His boss noted these changes and asked him to see a counselor. He did not go. His wife, Karen, also noticed that he was becoming increasingly irritable.

Unfortunately, when Jack's company merged with another, it became evident that the merger would eliminate many jobs. Jack took an early retirement package, which his boss strongly recommended. Because this resulted in a substantial decrease in household income, Jack looked for another job, but nobody hired him because of his overt chorea. He eventually found a job that paid less but he was fired a week later because he could not manage his work responsibilities. He started drinking heavily, which triggered frequent domestic disputes. One day, after an argument with his wife, he stormed out of the house. An hour later, Karen received a call from the police. Jack had been involved in a traffic collision. Although his car flipped three times, no one was hurt. Because he was drunk, his driver's license was suspended. His wife consulted with a social worker at the local chapter of the Huntington's Disease Society of America. After extensive counseling and treatment, Jack stopped drinking and his depression abated. He applied for social security disability. Jack followed through with the social worker's suggestion to prepare an Advance Directive and a durable power of attorney.

Three years have passed and Jack's disease has clearly worsened. He now has an increasing number of falls when walking, although his chorea is controlled relatively well with medications. He is no longer able to handle his finances. Karen is making all household decisions. Jack has not driven a car since the accident. He is still irritable and has occasional delusions but he has never been physically violent. He remains at home most of the time, doing most of the simple household chores but is no longer competent to handle anything complex. Since the accident, Karen has returned to work after being a housewife for 25 years. This was a major adjustment for her, but the situation is under control for the time being.

## ***Conclusion***

Functional competence is a complex issue, which involves different functions at different stages of the disease in different settings. Preparation for loss of competence, recognition

of the loss and implementation of contingency plans in a proactive fashion are important steps. Insight of the person with HD and caregivers is a key factor for successful execution of these steps. If the person with HD is in denial, the solution becomes challenging. The availability of services and financial support is critical. Education of professionals involved in these steps is essential to handling issues of functional competence.

## Addendum A

### **Advance Directives**

Several issues that are unique to the disease challenge the person with HD. These include nutrition, communication, abandonment and the young age of the person with HD. Therefore, the need for complete and definitive written and verbal Advance Directives is paramount to good quality care at the end of life as is respect for the person's autonomy.

Every state has its own set of Advance Directives. For example, Wisconsin offers the option of adding personal instructions to the document for the physician under the section "Other Directions." It has been determined that this is important since it is unclear when a patient becomes terminal under Wisconsin Law.

However, for many other states, the question *should I add personal instructions to my Durable Power of Attorney for Health Care?* is met with caution. The Partnership for Caring organization ([www.partnershipforcaring.org](http://www.partnershipforcaring.org)) advises people not to add instructions to Advance Directives, and especially not to add instructions to the Durable Power of Attorney: "One of the strongest reasons to appoint a Power of Attorney is to have someone who can respond flexibly as your medical situation changes and respond to situations you may not foresee." Instructions can restrict that person's power to act in your own best interest at a given time. It is extremely important to reassess Advance Directives over time since preferences change as a person's condition changes.

People with HD are urged to talk to their family, talk to their agent-in-fact (some states have acceptance statements that the agent reads, agrees to and signs) and talk to their physicians, so that all concerned understand patient preferences well ahead of time (preferably from the time of diagnosis).

In addition, it is helpful to:

- Get an explanation of treatments and procedures;
- Make sure the physician knows the quality of care/life the patient wants; and
- Make sure they are willing to follow the Advance Directives of the person with HD. (Note: The law does not force physicians to follow an Advance Directive if they have a moral or ethical disagreement with its content.)

A helpful tool that clarifies and supports individual wishes is the *Values History Form* developed by the Health Sciences Ethics Program at the University of New Mexico ([http://hsc.unm.edu/ethics/advdir/vhform\\_eng.shtml](http://hsc.unm.edu/ethics/advdir/vhform_eng.shtml)).

This form provides individuals with the opportunity to address multiple personal preferences and issues. The completed document, when attached to a person's Advance Directive, gives the appointed Health Care Agent and the physician a specific and comprehensive guide to desired care.

Topic areas include (with as many as 10 specific questions under each heading):

- Overall attitude toward life and health,
- Personal relationships,
- Thoughts about independence and self-sufficiency,
- Living environment,
- Religious background and beliefs,
- Relationships with doctors and other caregivers,
- Thoughts about illness, dying and death,
- Finances,
- Funeral plans, and
- Optional questions.

Another helpful document for people with HD is *Tube Feeding: Making the Decision: Frequently asked Questions* by Betsy Gettig, M.S., D.G.C. and Toni Finney, B.S. ([www.kumc.edu/hospital/huntingtons/tube.html](http://www.kumc.edu/hospital/huntingtons/tube.html)). This excellent guide helps people with HD and their families with this difficult decision. Long-term feedings do not reverse the course of illness or improve quality of life. Withdrawing treatment is acceptable when no longer beneficial. This document can also be included as part of the Advance Directive.

## Addendum B

### **Euthanasia and Physician-Assisted Suicide**

Euthanasia refers to an act by a third party that directly and intentionally results in death. In the United States and in all countries except the Netherlands and Belgium, euthanasia is illegal.

Physician-assisted suicide refers to a situation in which the affected individual performs the final act that results in death. For instance, by taking an overdose of pills or an injection of some medication, but where the physician has helped in some way, such as by inserting an intravenous line or writing a prescription for the lethal dose of medication, with the intent of helping the patient achieve death. Physician-assisted suicide is illegal throughout the United States except in Oregon.

Many physicians are uncomfortable discussing death. However, people with HD and their families must be permitted, even encouraged, to discuss the terminal stages of their disease with their physician, as the end result of HD in all cases is death. Physicians who attend long-term care facilities may be more familiar with the process of imminent death than others, and may be a helpful resource at that stage of the disease. Hospice teams are available in some communities, and they provide an important bridge for the person with HD and their family during the immediately terminal stages of the disease.

Even prior to the very late stages of the disease, all individuals with HD benefit from developing Advance Directives, and discussing their care needs and wishes with their next of kin and their physician and medical care providers. By having adequate Advance Directives, one can often avoid the helplessness and despair that leads families and people with HD to consider suicide.

## Addendum C

### **Hospice Care in Relation to the Huntington's Disease Patient**

#### ***Background***

Hospice care is provided under Medicare Part A. The Hospice Medicare Benefit states that *if an illness runs its normal course*, life expectancy will be six months or less (the six-month rule). The patient is required to sign a statement choosing hospice care over curative care, thus aggressive treatments (chemotherapy, radiation, blood transfusions etc.) are no longer in the plan for care and will not be paid through the Medicare Hospice Benefit. Comfort measures such as oxygen, pain medications and sometimes blood products, continue to be used if they aid in the comfort of the patient. Medicare pays for all care related to the terminal illness (hospice care) and continues to pay for health care needs that are not related to the terminal illness.

#### ***Medicare Payment Structure***

The issue is not the provision of care but rather payment for care. The six-month rule was established by The Center for Medicare & Medicaid Services (CMS, formerly HCFA) for coverage of hospice services under Medicare. Most hospice agencies and insurance plans have adopted the six-month rule as a guideline for admission to hospice care. It is common for this time frame to be extended to a year or longer if a physician certifies that the patient continues to decline and is still considered terminal. (See criteria for Huntington's Disease.)

Under Medicare, the attending physician, as well as the hospice medical director, sign a statement certifying the individual for hospice care. After the first 90 days, the patient can be re-certified for another 90-day period followed by a 60-day period and then an unlimited period until death occurs. Thus, a patient may be under the Medicare Hospice Benefit for as long as a year or more. As long as the physician documents continual decline with no improvement, lengthy stabilization or available cure, a patient can be re-certified under Medicare for hospice services. Posing the question, *would you be surprised if this patient were still alive in six months?* can help the physician in their decision to re-certify at a given time.

In the event that a patient's medical condition improves, it is expected that the patient will be discharged from hospice care and provided for by home health care services or receive custodial care from the family system or long-term care facility staff. The patient can be re-admitted to hospice care when symptoms increase and decline is again evident.

#### ***Report on Hospice Care***

A recent report on hospice care (November 2002) by Dartmouth Atlas of Health Care and the National Hospice and Palliative Care Organization states that terminally ill patients are spending fewer of their last days in hospice care. Patients are entering hospice later in their illness. The average length of stay in hospice has declined from 64 days in 1992 to 51 days in 2002. In 2001, 37 percent of those served by hospice died within seven days or

less of initial referral. “Members of Congress two years ago called the decline in hospice lengths of stay troubling. They urged Medicare to remind doctors that if a patient lives longer than the six months initially expected, re-certifying that he or she remains close to death ensures that payment continues.” (See article on Last Acts Web site [www.lastacts.org](http://www.lastacts.org)).

Recently, CMS Chief Administrator Tom Scully urged all health care agents to help CMS engage the physician community and others in health care to think more about hospice care as they provide for terminally ill people.

### ***State Licensure Laws***

Each state has its own laws governing the licensure of hospices within their state. Within their own regulations, they can, to some extent, establish different criteria for the provision of hospice care. Rather than *six months or less*, Florida hospice care allows for *one year or less until death may occur*. States have influence over terminal care in their state.

### ***Private Insurance***

Private insurance may also have variations of the six-month rule. For example, there may be a capitation for palliative and hospice care that spans more than a year. However, when that amount is met, there will be no more coverage for terminal care whether the person still needs care or not. Private insurance can allow increased time for terminal care by comparing the cost of care outside of a hospice program versus care provided within a hospice program to determine which is most cost effective for patients.

### ***Assessments to Determine Appropriateness for Hospice Care***

There are a number of Functional Assessment Scales that determine readiness for hospice care. Most hospice programs have long used the Modified Karnofsky Performance Status Scale (MKS). The rating is by number and related to functionality, decreasing from 100 points for *No Complaints, No Evidence of Disease* to 10 for *Actively Dying*. Diagnosis is a factor in the number of points needed to qualify for hospice care.

“Vista Care,” a national for-profit hospice agency with locations in 14 states outlines the following criteria for hospice admission for a person with the diagnosis of HD:

The patient has end-stage HD with both 1 and 2:

1. Stage VII or beyond, according to the functional Assessment Staging Scale with all of the following:

- Inability to ambulate without personal assistance;
- Inability to dress without assistance;
- Urinary and fecal incontinence, intermittent or constant; and
- No meaningful consistent verbal communication.

AND

2. Has had at least one of the following conditions within the past 12 months:

- Aspiration pneumonia;
- Pyelonephritis or other upper urinary tract infection;
- Septicemia;
- Decubitus Ulcers, Multiple, Stage 3-4;
- Fever, recurrent after antibiotics; and
- Inability to maintain sufficient fluid and caloric intake with one or more of the following during the preceding 12 months:
  - a. 10 percent weight loss during the previous six months;  
OR
  - b. A serum albumin greater than 2.5 gm/dl;  
OR
  - c. Significant dysphagia with associated aspiration measured objectively (e.g., swallowing test or history of choking or gagging with feeding).

In the absence of one or more of these findings, rapid decline or co-morbidities support eligibility for hospice care.

### ***What can be done Regarding the Person with HD's Need for Hospice Care?***

- Physicians need to be educated to refer people with HD to hospice care when functional assessment criteria for hospice can be met.
- Patient/family need to be aware of their private insurance coverage regarding home care/hospice care and access it in a timely manner.
- People with HD and their families would benefit from awareness of individual state regulations on hospice care.
- HDSA Centers of Excellence physicians need to be educated about hospice referral – for patients in the home and in long-term care facilities.
- HDSA Centers of Excellence and HDSA Chapter social workers need to promote hospice care when educating long-term care facility staff during in-service programs.
- Hospice agencies need to be educated about HD. In-service programs must be provided to assist them in identifying the unique care needs of HD.
- The six-month rule pertains primarily to payment through Medicare. As noted, hospice care is not accessed often enough, given the need. The six-month policy is a guideline that can be extended to serve the patient until death. The public and health



care providers must be educated about the benefits of hospice in order to effectively utilize this important service.

### ***What Is Being Done***

#### ***Benefits Improvement & Protection Act of 2000 (BIPA)***

This legislation continues to support the established six-month rule. Under “Subtitle C, Hospice Care” in the summary of BIPA, Section 322 states: “The primary care physician and the Hospice Medical Director must certify that the individual has six months or less to live. The certification of terminal illness shall be based on their clinical judgments regarding the normal course of the individual’s illness.”

([www.ppsv.com/issues/givebackdoc.htm](http://www.ppsv.com/issues/givebackdoc.htm).)

#### ***National Hospice and Palliative Care Organization***

The National Hospice Organization originated in 1978 and changed its name to the National Hospice and Palliative Care Organization (NHPCO) in 2000. Palliative care extends the principles of hospice to a broader population that could benefit from receiving this type of care earlier in the disease process.

## Addendum D

### **Policy Addendum Regarding HIPAA and End-Stage Care in HD**

The enactment of the Health Insurance Portability and Accountability Act (HIPAA) in 1996, modified by the Department of Health and Human Services under the Bush administration in 2002, contains significant new regulations designed to protect the privacy of individual medical records and to control the electronic storage and transmission of those records.

Because Huntington's Disease has some unique characteristics that distinguish it from other neurodegenerative diseases such as Parkinson's and Alzheimer's, several policy issues must also be considered:

1. *Protection of privacy by care services that are provided through non-medical employees.* The care center or setting must ensure that any contracts with quality of life organizations include an agreement to abide by the same privacy and confidentiality rules that the care center follows. The same contract is required with volunteers. These groups would be considered "contractors" and would engage in agreements detailing the services that would be provided, as well as include a confidentiality agreement.
2. *Obtaining authorization and consent for information-sharing between health care groups or providers and quality of life providers.* Consent forms must have a clause to the effect that the care facility may provide information to certain "contractors."
3. *Accounting for the disclosure of information.* Upon request, a facility must provide a report documenting to whom information has been sent whether the information is forwarded to medical or non-medical providers.
4. *Consent for the family to obtain information about the patient.* According to HIPAA, oral authorization from the patient is sufficient to determine who may be involved in care. Without oral authorization by the patient, no information may be released under any circumstances in order to protect the patient's privacy.

## Addendum E

### Values History Form

NAME: \_\_\_\_\_ DATE: \_\_\_\_\_

If someone assisted you in completing this form, please fill in his or her name, address, and relationship to you.

Name: \_\_\_\_\_

Address: \_\_\_\_\_

Relationship: \_\_\_\_\_

#### **Overall Attitude Toward Life and Health**

What would you like to say to someone reading this document about your overall attitude toward life?

What goals do you have for the future?

How satisfied are you with what you have achieved in your life?

What, for you, makes life worth living?

What do you fear most? What frightens or upsets you?

What activities do you enjoy (e.g., hobbies, watching TV, etc.)

How would you describe your current state of health?

If you currently have any health problems or disabilities, how do they affect: You? Your family? Your work? Your ability to function?

If you have health problems or disabilities, how do you feel about them? What would you like others (family, friends, doctors) to know about this?

Do you have difficulties in getting through the day with activities such as: Eating? Preparing food? Sleeping? Dressing and bathing?

What would you like to say to someone reading this document about your general health?

#### **Personal Relationships**

What role do family and friends play in your life?

How do you expect friends, family and others to support your decisions regarding medical treatment you may need now or in the future?

Have you made any arrangements for family or friends to make medical treatment decisions on your behalf? If so, who has agreed to make decisions for you and in what circumstances?

What general comments would you like to make about the personal relationships in your life?

### **Thoughts About Independence and Self-Sufficiency**

How does independence or dependence affect your life?

If you were to experience decreased physical and mental abilities, how would that affect your attitude toward independence and self-sufficiency?

If your current physical or mental health gets worse, how would you feel?

### **Living Environment**

Have you lived alone or with others over the last 10 years?

How comfortable have you been in your surroundings? How might illness, disability or age affect this?

What general comments would you like to make about your surroundings?

### **Religious Background and Beliefs**

What is your spiritual/religious background?

How do your beliefs affect your feelings toward serious, chronic or terminal illness?

How does your faith community, church or synagogue support you?

What general comments would you like to make about your beliefs?

### **Relationships With Doctors and Other Health Caregivers**

How do you relate to your doctors? Please comment on: trust, decision making, time for satisfactory communication, and respectful treatment.

How do you feel about other caregivers, including nurses, therapists, chaplains, social workers, etc.?

What else would you like to say about doctors and other caregivers?

### **Thoughts About Illness, Dying and Death**

What general comments would you like to make about illness, dying and death?

What will be important to you when you are dying (e.g., physical comfort, no pain, family members present, etc.)?

Where would you prefer to die?

How do you feel about the use of life-sustaining measures if you were: Suffering from an irreversible chronic illness (e.g., Alzheimer's disease)? Terminally ill? In a permanent coma?

What general comments would you like to make about medical treatment?

### **Finances**

What general comments would you like to make about your finances and the cost of health care?

What are your feelings about having enough money to provide for your care?

### **Funeral Plans**

What general comments would you like to make about your funeral and burial or cremation?

Have you made your funeral arrangements? If so, with whom?

### **Optional Questions**

How would you like your obituary (announcement of your death) to read?

Write yourself a brief eulogy (a statement about yourself to be read at your funeral).

What would you like to say to someone reading this Values History Form?

### **Legal Documents**

What legal documents about health care decisions have you signed? (Each state has its own special form—feel free to add yours to this list.)

Advance Directive for Health Care – [Enter Specific State] Yes \_\_\_\_\_ No \_\_\_\_\_

Where and with whom can it be found?

Name \_\_\_\_\_

Address \_\_\_\_\_

Phone \_\_\_\_\_

Living Will? Yes \_\_\_\_\_ No \_\_\_\_\_

Where and with whom can it be found?

Name \_\_\_\_\_

Address \_\_\_\_\_

Phone \_\_\_\_\_

Durable Power of Attorney for Health Care Decisions? Yes \_\_\_\_\_ No \_\_\_\_\_

Where and with whom can it be found?

Name \_\_\_\_\_

Address \_\_\_\_\_

Phone \_\_\_\_\_

Health Care Proxy? Yes \_\_\_\_\_ No \_\_\_\_\_

Where and with whom can it be found?

Name \_\_\_\_\_

Address \_\_\_\_\_

Phone \_\_\_\_\_

