



Long Term Care Monograph

Overview of HDSA Long Term Care Workgroup

HUNTINGTON'S DISEASE SOCIETY OF AMERICA

The Huntington's Disease Society of America (HDSA) is a national, voluntary health organization dedicated to improving the lives of people with Huntington's disease (HD) and their families. HDSA promotes and supports research and medical efforts to eradicate the disease, assists people and affected families to cope with the problems presented by HD and educates the public and health professionals about the disease.

HD AND LONG TERM CARE

HD is a chronic progressive brain disease that always causes severe disability with complete dependency in its late stages, requiring 24-hour supervision and care either in the home or in LTC facilities, often for many years. Recognizing challenges in both the provision and acquisition of high quality LTC for Huntington's disease, HDSA convened a workgroup of experts on HD with the goal of preparing a monograph on Long Term Care for the person with Huntington's Disease.

DEFINITION OF LONG TERM CARE

LTC comprises a variety of services which help meet both the medical and non-medical needs of people with a **chronic illness** or **disability** who cannot care for themselves for long periods of time.

LTC is provided in many settings. LTC for most persons with HD begins in the home, as many families prefer to keep the person with HD at home for as long as possible. Services in the home are most commonly provided by family caregivers. This informal network of care offers persons with HD personalized assistance from those who know them, in a setting that is comfortable for them. Families may provide home care services without assistance, or may engage

home care agencies to provide personal assistance in the activities of daily living (ADLs), or in such things as housekeeping, transportation, medical services, nursing services, skilled physical, occupation, or speech therapy services.

Many persons with HD participate in adult daycare services, which may provide transportation, meals, supervision, recreation, some skilled therapies and even some health services on an outpatient basis. For day care programs, the person with HD lives at home, but travels weekdays to a daycare facility. In outpatient settings, the person with HD may receive care from outpatient physicians (some do home visits) or clinics, and receive laboratory or skilled physical, occupational or speech therapy services in clinics or hospital settings.

Some persons with HD, especially those who are over 55 years old, eligible for both Medicare and Medicaid, and dependent enough to need a nursing home, may be cared for in a Program for All-inclusive Care of the Elderly (PACE) program or in similar innovative approaches to LTC. PACE programs are new but can provide more extensive in-home services to prevent or postpone institutionalization. The program can include services that are not traditionally financed through Medicare and Medicaid – for example modifications of the home or a paid companion – because they realize savings from reducing institutionalization in the hospital or a LTC facility. PACE is just now rolling out and may not be available in all areas. Respite services offer those caring for a person with HD at home relief by placing the person in a nursing home for a limited time so the family member can take care of their own health needs, travel for a special event or whatever is needed.

A number of “inpatient” settings also provide care for persons with HD. Assisted living facilities or community group homes provide meal service, offer minor assistance with medications, and provide some level of supervision. Intermediate care facilities offer custodial care, provision of medications and supervision and assistance with ADLs. Skilled nursing facilities (often referred to as “nursing homes”) provide similar assistance, but with a higher level of nursing care, and may also provide skilled therapies such as physical therapy, occupational therapy, and speech therapy. Some skilled nursing facilities are secured to care for persons with dementia or mental illness who may wander or try to leave, but are unable to care for themselves. There are “continuing care communities” that charge a large upfront entrance fee and then care for enrollees at whatever level is needed for the rest of their lives – starting out as independent living, in perhaps an apartment, then progressing through the level of assisted living and even providing skilled nursing services and terminal care at the end of life. These are expensive, rare and typically have no experience with HD. For those who are deemed terminal, hospice may provide services for the person in the home, in a LTC setting, or in a free standing hospice.

LTC can be provided in hospitals or medical or psychiatric facilities in certain circumstances. Although this is not ideal, finding an appropriate placement following an acute hospitalization can be challenging, thus an individual may have a protracted course in a medical or psychiatric hospital. The Veterans Administration offers some veterans services such as hospitals and nursing homes, as well as outpatient services to assist some veterans with LTC needs. Those with HD are also served in more atypical LTC settings such as in prisons from time to time.

Some of the difficulties faced by persons with HD in LTC settings are similar to other diseases. Uneven quality, high costs, patchy reimbursement, and limited availability of services are troubles plaguing the LTC market. However, for persons with HD, there are some unique challenges. LTC is traditionally utilized for those who are elderly, and services are designed to meet the needs and preferences of this population. Because of this, LTC facilities are not designed to care for younger people, those with behavioral issues, and those with gait and balance problems characteristic of HD. In addition to these limitations, very few facilities have experience with HD.

A delicate patchwork of funding from local agencies, Medicare, Medicaid, and private insurance proves inadequate to sustain in-home and day care services, without considerable personal cost. Private insurance and Medicare provide limited funding for rehabilitation services provided in skilled nursing facilities subsequent to an acute hospitalization. Private LTC policies can be used to pay for home-based or institutional care. However, the majority of the costs for LTC in HD are provided by Medicaid.

MISSION AND VISION OF THE HDSA LONG TERM CARE WORKGROUP

Mission

The goal of the HDSA Long Term Care Workgroup is to ensure dignity for those affected by this debilitating and chronic disease. The mission of the Workgroup is to identify critical issues related to LTC for persons with Huntington's disease (HD). This will be accomplished by reviewing existing services and resources necessary to provide access to high quality care in late-stage HD and by defining gaps and barriers to these services for persons with HD and their families.

Vision

The vision of the HDSA Long Term Care Workgroup is to provide information that is timely, relevant, customized, and innovative in delivery regarding the long-term care needs in persons with HD.

HDSA Long Term Care Workgroup Process

The Workgroup, first convened in June 2011, included neurologists, neuropsychologists, medical directors and managers of LTC units, social workers, a bioethicist, an attorney and a family caregiver. HDSA envisioned an 18-24 month process that would assess the current state of the field, as compared to a vision of optimal care, and propose strategies geared toward improving care in this area, essential not only to persons with HD and their families, but also to constituents with other disabling nervous system disorders, providers of LTC, other consumers of LTC, and policy-makers. At the initial in-person meeting in June 2011, participants were convened and Subcommittees to address care, education and policy were formed. The workgroup was chaired by Kathleen M. Shannon, MD.

At the initial meeting, the participants defined the mission of the Workgroup, stakeholders for the report, and the scope of the project and divided into Subcommittees. The participants met at two subsequent in-person meetings and worked independently and by conference call over an 18-month period. Each Subcommittee defined the scope of its contribution to the document, defined the state of the field of LTC in HD, worked to envision optimal care models, and proposed ways to move from current strategies toward a more functional model of LTC. Each Subcommittee prepared a summary document and these were integrated into this comprehensive document.

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This document is addressed to a number of potential stakeholders, including persons with HD and their families and caregivers, health care professionals who care for persons with HD, educators, policy makers, lobbyists, surveyors of health care quality, licensing organizations, ombudsmen, third-party payers, professional societies, and product developers.

INTRODUCTION TO HD

Huntington's Disease (HD) is an inherited neurodegenerative disease with motor, cognitive and behavioral manifestations. The prevalence of HD in the Americas and most of Europe is about 1 in 10,000, with an estimated 30,000 Americans living with HD. For each affected person, it is estimated there are 5-7 persons genetically "at risk" of HD. More than 200,000 Americans suffer with symptoms of HD, provide care for one or more affected relatives, or live under the specter of potentially developing the devastating disease. Symptoms of HD most often start between the ages of 35 and 50 but onset of disease can occur anytime. Age at onset is conventionally defined as the age at which the characteristic motor syndrome can be identified by clinical inspection. However, a burgeoning literature suggests that milder signs of cognitive, behavioral or motor decline may be detectable up to 20 years in advance of diagnosable disease, and that sensitive imaging studies may also be able to detect structural brain changes well in advance of the onset of symptoms or signs. Once the disease begins, there is a progressive decline that typically lasts 15-20 years on average.

Chorea, derived from the Greek word for "dance," is the hallmark motor sign of HD. Choreic movements are rapid, purposeless, involuntary movements that have a flowing quality. Choreic movements affect the muscles of the face,

neck, trunk, arms and legs. Other involuntary movements that can be seen in HD include faster jerking or twitching movements (tics, myoclonus) and slower twisting or writhing movements (dystonia). In addition to these spontaneous involuntary movements, persons with HD have difficulty performing normally coordinated voluntary movements. Movements are slow and clumsy, with abnormalities in amplitude, timing and coordination. Persons with HD have difficulty maintaining constant muscle force (motor impersistence). This leads to dropping things, as well as transient loss of tone in antigravity muscles when walking. Limb rigidity can be detected by passive range of motion. Voluntary motor deficits begin with fine coordinated movements, but over the disease course impairments in gross motor function develop. Every type of movement is affected, including those required for normal speech and swallowing, gait, and balance. Most persons with HD progress from mild changes in movement to a bedbound state characterized by gross impairments in communication and self-care.

Parallel to the increased motor disability, cognitive decline begins early in the disease with difficulties in attention, concentration, multitasking, prioritizing, planning, and learning. This constellation of deficits in higher "executive" functions evolves into a generalized dementia with severe disability. Across the course of dementia, there is a pronounced loss of communication skills and, eventually, persons with HD are generally non-verbal, though they maintain some ability to comprehend.

Superimposed on these progressive changes is a panoply of behavioral symptoms. Irritability, impulsiveness, impatience and anxiety are

common. Affective disturbances are nearly universal. Depression is most common, though mania, obsessive compulsive features, and psychosis may be seen. Suicidal ideation is common, though its severity may fluctuate across disease stages, occurring most often around the time of diagnosis and around the time of disability milestones (loss of employment or driving ability, for example). It has been reported that as many as 25% of persons with HD have attempted suicide at some time in their disease course. Unlike motor and cognitive changes that show gradual progressive decline, behavioral changes show a fluctuating course and persons with HD who have mild cognitive and motor disability may show severe psychiatric symptoms.

In about 10% of persons with HD, symptoms begin before the age of 20. Juvenile onset HD has a different motor appearance from that seen in adults with prominent dystonia, parkinsonism, myoclonus and seizures. As in the adult form of the illness, there is an inexorable progression to a bedbound, dependent state.

The genetic basis of HD has been known since 1993. HD results from an autosomal dominant mutation in the coding region of the huntingtin gene on the short arm of the 4th chromosome. The mutation is an expanded, unstable repeat of the trinucleotide cytosine-adenine-guanine (CAG) repeat. Repeat lengths of 40 or more are associated with complete penetrance. Incomplete penetrance is seen in repeat lengths of 36-39. Persons with 28-35 repeats are considered to have a premutation, and will not generally become ill over the course of a normal lifespan. Because the mutation is unstable, the CAG repeat length may increase in subsequent generations. This may lead to the *de novo* appearance of disease

in a family, when a premutation expands into the fully penetrant range, as well as to the phenomenon of anticipation, where onset in a subsequent generation is earlier than in the prior generation. Instability is more common with male transmission, accounting for the observation that most very early onset cases have been inherited from an affected father. While the discovery of the huntingtin gene and its mutation in 1993 paved the way for groundbreaking research into the causes and potential treatments of HD, it also introduced the possibility of identifying those who inherited the gene mutation in the presymptomatic state. Absent an effective therapy that would forestall the appearance of HD, fewer than 10% of persons at risk elect to undergo predictive testing.

HD AND CAREGIVING AT HOME & IN INSTITUTIONAL SETTINGS

HD strikes at an age that is typically characterized by high productivity in the workplace and at home. Persons with HD lose the ability to work and drive relatively early in the disease course. They become unable to perform domestic and childcare activities, and their normal roles must be assumed by other family members. Moreover, their presence in the home is a constant reminder to the healthy spouse and children that other family members may meet the same fate. While persons with HD can provide their own self-care for several years, they become progressively dependent on caregivers for their personal needs. Most are initially cared for at home by family members, but increasing needs over time force most families to secure 24-hour supervision and care, often in institutional settings, for the person with HD.

Several studies have addressed the clinical characteristics of persons with HD in LTC facilities. These studies suggest that persons

with HD are younger and have better overall health than other residents. Persons with HD in LTC have an average disease duration of 9-10 years and have more severe motor impairments and more psychiatric and behavioral problems than those cared for in the community. Other studies have defined features that predict LTC placement. These include the severity of motor and cognitive impairment and impaired ability to perform activities of daily living. Multiple studies have examined barriers to LTC. These include lack of specialized units, multidimensional clinical features of HD, lack of staff experience, poor access to specialty services, family opposition due to lack of confidence in the quality of care provided, and problems with financing care.

The following Subcommittee reports on Care, Education and Policy will examine the state of the field, present a gap analysis and a vision of optimal care, and make recommendations to the field aimed at improving the accessibility, affordability and quality of LTC for persons with HD. ■■■

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SUBCOMMITTEE REPORTS:

Care Subcommittee Report

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REPORT ON THE STATE OF THE FIELD

There is a paucity of research about the care of persons with Huntington's disease (HD) within LTC settings.

Several studies have addressed the clinical characteristics of persons with HD in LTC settings. Nance and Sanders (1996) conducted a retrospective review of 97 persons with HD in LTC settings and found that they tended to be younger, had relatively minimal serious medical disorders (other than HD), and had a duration of illness of about 10 years. The major challenges that providers in LTC settings faced included management of behavior, dietary management, and adjustment by persons with HD, family, and staff to progressive neurological and cognitive decline. The most common LTC issues for persons with HD included: smoking, incontinence, maintaining personal hygiene, decubitus ulcers/skin tears, pneumonia (which was the second largest cause of death), urinary tract infections, and recurrent fever.

Wheelock and colleagues (2003) investigated predictors of LTC placement using the large, extensive HD database developed by the Huntington Study Group (HSG). At the time of this study, the HSG database contained clinical and demographic information on over 3,000 persons with HD. Of these, 228 were placed in a LTC facility. Again, these persons with HD tended to be younger (average age of 52 years), predominantly female, and had a mean duration of disease of nearly nine years. They also displayed more severe clinical features of HD, including greater motor impairment and more psychiatric or behavioral problems than persons with HD living at home. The main predictors of long-term placement included bradykinesia, impaired gait and mobility, and the presence of poorly controlled behavior.

More recently, Rosenblatt et al (2011) investigated factors contributing to placement for persons with HD. These authors conducted a retrospective chart review of their HD center which contained a database of over 4,300 persons with HD followed for several years. Of these, 88 were placed in LTC facilities after being followed for about nine years. Motor impairment, with diminished capacity to complete most activities of daily living (ADLs), had the strongest correlation with time until placement. Interestingly, the genetic expansion score (i.e. CAG repeat length) of the person with HD was associated with disease duration at placement when controlling for age at onset. In addition, these persons with HD had a higher CAG repeat expansion and lower educational attainment than those who avoided institutionalization for at least 15 years after disease onset. The major predictors of LTC placement included ADL score, impaired cognitive function, and impaired motor function.

Dubinsky found that few hospitalized persons with HD are able to return to the home setting following discharge. Fifty five (55%) percent of hospitalized persons with HD transitioned to a LTC facility after hospitalization. It is thought that hospitalizations may follow a crisis (injury, behavioral problem or illness) or a failure of caregiving – both situations indicating a decline that prompts a more rapid consideration of a new LTC option.

There have been multiple studies which have examined barriers to LTC. The major findings of these studies suggest that there currently are few specialized residential care options for persons with HD. Most persons with HD are placed in geriatric, traumatic brain injury or severe psychiatric LTC settings. The unique clinical features of persons with HD (e.g. younger age, greater motor/behavioral problems, etc.)

are common obstacles to LTC placement. These studies also found that lack of staff experience with taking care of persons with HD contributed to determining whether or not a person with HD would be admitted to the facility. Many LTC facilities also lack access to HD specialists such as movement disorder neurologists, neuropsychiatrists and neuropsychologists who can best identify and treat the major sources of disability in persons with HD. Given that persons with HD in LTC settings tend to be younger and relatively healthier, there is a failure by LTC providers to recognize that HD is a terminal illness. Thus, for persons with HD, palliative care is typically reactive rather than proactive. In addition, many long term care facilities do not have ready access to the unique needs of persons with HD such as speech therapy, OT/PT, and recreation therapy. A somewhat surprising barrier to long-term placement for persons with HD comes from person/family opposition to placement which is based upon a lack of confidence in the LTC facility. Finally, and perhaps the most salient barriers to LTC placement for persons with HD, are reimbursement and financial issues.

Long term care is costly. The relatively young age at placement for persons with HD increases costs, because persons with HD may spend many years in LTC. The issue of health care utilization costs as a barrier to LTC placement for persons with HD was illustrated in the study by Murman et al (2002). These investigators compared health care utilization and direct costs among persons with Alzheimer's disease (AD), persons with Parkinson's disease (PD) with dementia, and persons with HD. The LTC costs accounted for the majority of direct costs in all 3 groups. However, the presence of dementia with Parkinsonism resulted in significantly greater utilization of LTC

services and higher total direct costs. Medicare payments for direct costs were significantly higher for AD (74%) and PD w/dementia (78.3%) compared to HD (51%) which suggests that more health care dollars are being allocated for older patients with neurodegenerative disorders other than HD. There was a significantly lower number of persons with HD in LTC (42%) compared to AD (53%) and PD with dementia (77%). Persons with HD also had a significantly lower percentage of paid home care (36%) compared to AD (49%) and PD with dementia (76%). These facts suggest that in the case of HD, a greater burden of the costs of LTC is falling on families in terms of the unpaid care they provide and the out-of-pocket expenses they incur.

In 2004, the results of the Robert Wood Johnson Foundation Promoting Excellence in End of Life Care HD Workgroup, created in collaboration with HDSA, were published. This report highlighted the need to: (1) investigate barriers to adapting advanced care plans, (2) identify the demographic features of persons with HD and the types of professionals who care for them in order to develop recommendations for appropriate allocation of health care resources, including the need for LTC placement, (3) compare institutional versus home care settings to identify risk factors for placement in LTC facilities and resources required for persons with HD placed in LTC, and (4) develop evidence-based guidelines that clinicians can use to determine the need for LTC.

Dellefield and Ferrini (2011) found that the Robert Wood Johnson Foundation Promoting Excellence in End of Life Care HD Workgroup initiative provided a meaningful framework for setting clinical priorities and they used this framework to summarize the clinical lessons that

nursing staff and interdisciplinary team members learned about caring well for persons with advanced HD.

In summary, there is published information regarding the clinical characteristics of persons with HD in the LTC environment, the major predictors of LTC placement, and barriers to acquiring high quality care for the persons with HD. Future LTC research needs to focus on: (1) factors affecting access to LTC placement for persons with HD, (2) development of training/ education programs for health care providers in the LTC setting with respect to the more unique neurological and neuropsychiatric needs of persons with HD, and (3) more extensive health care utilization costs for persons with HD in the LTC setting.

GAP ANALYSIS

The Care Subcommittee performed a gap analysis primarily by contacting and interviewing key staff at seven LTC facilities with specialized HD units and two HD-focused group homes within the United States. It is estimated that of the approximately 1.5 million skilled nursing beds in the US, 250 or fewer comprise HD specialty beds. Therefore, most families dealing with HD are not relying on these specialty care units for nursing home placement.

Challenges to delivering care

Through the interview process, the Subcommittee identified the following as primary challenges to delivering care to persons with HD:

- High variability in the progression and severity of behavioral symptoms
- Frequent falls, though few with serious injuries

- Variable use of restraints, which are seen as a last resort
- Difficulty recognizing the movement disorder as distinct from “misbehavior”
- Environmental and equipment damage due to the movement disorder (walls, doors, toilets, etc.)
- Difficulty finding seating options and other equipment tailored to the special needs of the person with HD
- Provision of activity programs that appeal to residents with a “younger mindset”
- Difficulty with the constant need to be creative and try new approaches to care
- Challenges of providing support to LTC staff dealing with the ongoing decline of a person with HD
- Insufficient staff education

Barriers to care

The Subcommittee solicited feedback from a variety of clinicians who work with persons with Huntington’s disease in the home, outpatient clinics and LTC settings. This feedback was provided from colleagues in North America, Canada and Europe. The analysis focused on current perceived barriers to care for persons with HD, primary issues currently for persons with HD living in LTC settings, and secrets of success. The following were specifically identified:

- Overcoming myths and fears about HD, including beliefs that persons with HD are violent and require specialized care
- Fear of involuntary movements
- Fear of behavioral problems





- Lack of comfort with, and feeling unprepared, to deal with the younger age of a person with HD
- Fear that a psychiatric history is “too strong” (i.e. psychiatric symptoms have been challenging or difficult to manage)
- Previous difficult experience with a person with HD with the false belief that every person with HD acts the same way and has the same needs
- Feeling on the part of LTC staff that they aren’t valued for their contribution to caring for the person with HD
- Lack of time and focus on education so that staff can learn about HD and other diseases they are treating.

Primary concerns for current persons with HD residing in LTC settings

The Subcommittee identified the primary concerns for persons with HD in LTC settings. These include:

- Failure of family, especially young children, to visit
- Fear that rapid admission to the Emergency Room or Acute Psychiatric Unit will result in the inability to return to the LTC setting
- Disruption to families resulting from the need to travel a significant distance to visit the person with HD in LTC
- Lack of clear communication with HD experts (including family members) and direct care staff
- Need for HD research within LTC settings to define better quality of care
- Lack of age-appropriate activities so that there is nothing to do all day

- Need for patience and thinking “outside of the box” to provide care for persons with HD
- Lack of effective therapies for advanced stage HD
- Fear of a long distance placement which would make it difficult for family to visit frequently.

Secrets to success in LTC settings

Based upon the aggregated experience of providers of care in specialty HD LTC units, family members, experts in HD, and others suggests the following secrets to the successful care of persons with HD in LTC:

- Provide a variety of fun, unique, and age appropriate activities
- Recognize that activities and therapy should be available on weekends, as well as weekdays
- Let smokers smoke safely with help, as needed, and/or with adaptive equipment
- A team approach to decision-making and direct care, including incorporating ideas from front line staff
- Frequent, high-quality staff education to ensure a well supported staff with some working knowledge of HD
- Constant attempts to find ways to improve care
- Maintain a low staff turnover rate
- Pleasant interaction between staff and residents
- Maintain a staff that really cares about the person with HD residing in their facility.

RECOMMENDATIONS TO THE FIELD

Guidelines for the management of persons with HD residing in LTC

1. Improve understanding of the unique and complex needs of the HD family related to the LTC setting

A. The role of genetics and family dynamics

Huntington’s disease is an inherited neurological disorder with a late onset and long period of progression that is characterized by changes in motor function, cognitive deterioration and mood disturbances. Because it is an autosomal dominant disease, every biological child of a person born with the mutated HD gene, independent of sex, will have a 50% chance of inheriting the gene for HD and therefore developing HD. Predictive genetic testing for HD was introduced in 1993, but the majority of at risk individuals have opted not to undergo predictive testing. Those who decide to be tested generally choose to do so in preparation for a major life decision or profess discomfort not knowing something that can be determined about their genetic risk. The added complexity and “weight” that the inherited nature of this disease can place upon a family should come as no surprise. The atmosphere in families affected by HD is very different from a normal family. The presence of HD is associated with several types of losses experienced by the family: physical and personality changes of the affected family member, loss of the usual family structure, and potential loss through placement in a nursing home, and eventually death. Indeed the two most difficult things to deal with, as reported by families affected by HD, were identified as mental deterioration of the affected family member and fear of having

passed the HD gene onto their children. By the time families affected by HD consider LTC placement, the early years of living at risk, in some cases informed by a positive genetic test, followed by many years of progressive HD symptoms, have taken a toll on even the most resilient of families. For this reason, upon admittance to a LTC setting, many HD families bring the “baggage” of a chaotic upbringing, the genetic risk for their current family member, and many fears for themselves and the future generation.

B. Common emotional experiences of the HD family

It is not only in the genetic sense that HD is a family disease. With the average age at onset of symptoms between the ages of 30 and 50, HD begins during the time period when the family life-cycle is at its most complex, with childbearing, child-rearing and career development all important. Additionally, there may be many members in any one family, and almost certainly many members in the extended family, who are affected in some way by HD. Distress and grief may be directed at the ongoing genetic load, the losses experienced by affected relatives, the need to mourn those who have died, and suffering the anticipatory mourning for those who are symptomatic and being placed in a LTC setting. Family grief in the setting of a progressive illness or death exaggerates the existing structure and coping style of the family. Thus cohesive families may become more cohesive while disorganized ones may become even more chaotic.

C. Common stress points for HD families related to LTC placement

HD families have often faced either a long road to LTC placement or had to “wait for the crisis” to bring about the opportunity for placement. Regardless of how LTC placement has “come about,” the HD family has often been faced with some or all of the following barriers:

- False belief that all persons with HD need specialized care
- Concerns that the person with HD is younger than age 60
- Worry that LTC staff will know nothing about HD
- Fears that the person with HD will feel abandoned
- Guilt about placement
- Difficulty getting the person with HD to agree to placement, especially because inflexible thinking on the part of the person with HD doesn’t easily permit the vision of a new and better quality of life
- Financial considerations and pressures, with premature entry into LTC as a result of insurance coverage or limitation of options due to lack of coverage.

D. Common issues HD families initially confront during LTC placement

Often times, LTC staff may wonder why a person with HD doesn’t receive a lot of visits from family members. In particular, for at risk family members, visiting a person with HD can sometimes be too emotionally taxing. It can feel like “looking into a mirror” of what may very

well be their own future. Hence, visiting for at risk members can become too challenging, thus leading to avoidance or denial as coping mechanisms. Often a person with HD may have younger children at home who may struggle to understand and visit a parent who is in a “funny and strange place with lots of old people.” Sometimes families become torn and stressed due to a lack of LTC options that are close to home. Indeed lack of proximity to good care can place an undue burden on families affected by HD. In many ways, the ability of the person with HD to adjust to placement appears tied to the family’s own ability to adjust. Families affected by HD that are overly anxious can tend to project feelings of neglect onto the LTC staff. Families can become “overly involved” in the care of their person with HD and optimal adjustment for both many never occur. Adult children and well spouses may feel that they have abandoned their person with HD or “broken” a promise that they would never placed their family member in a LTC facility.

HD families often report a struggle to understand the “long term care culture.” They may have distorted expectations of what a LTC facility is like. They feel uncertain in knowing how to respond to perceived deficiencies in care and their own role “ambiguity.” Indeed “well spouses,” in particular, may experience periods of depression and/or anxiety during the placement of their person with HD. They may have expected their stress levels to have lessened following placement, when, in reality, their stress has shifted from day to day daily care to managing their role as visitor, supporter, cheerleader and advocate. Due to the rare nature of HD, many families are much more knowledgeable about the disease than many





health care providers. Families affected by HD become used to being “experts” on HD and can often feel slighted or ignored by LTC staff that don’t appear to listen and respect their degree of knowledge. Alternatively, they may grow tired and resentful of this burden to be the experts.

E. Recommendations for and to help HD families in LTC settings

Family members should be urged to communicate with management as needed, to both express any concerns but also satisfaction and recognition of outstanding staff members. Typically the two senior managers in a LTC facility are the Administrator and the Director of Nursing. Families should be encouraged and supported to visit soon after placement, and often thereafter. The staff should be mindful of the unique issues that may impact the ability of HD families to visit, and should work with families to set reasonable expectations. No LTC facility will be able to deliver the same degree of care that was provided at home nor can a facility provide “perfect care.” It is reasonable to expect very good care that is provided on a consistent basis. Long term care staff, in particular social work staff, may need to gently remind a family caregiver what went into their decision for placement. Family members affected by HD may have a deep and profound regret that things got to the point of placement.

2. Medical Management of the Person with HD

The third and newest version of *A Physician's Guide to the Management of Huntington's Disease* serves as a resource to LTC Medical Directors and key staff for the medical management of Huntington's disease. It is the definitive work, and almost mandatory reading, for the latest

approaches to the management of late-stage HD. It offers specific detail on the staging, cataloging and respective approaches to the damage of chorea, oral-motor dysfunction, dementia, behavioral-psychological issues, and end-of-life issues. All are reviewed in the context of dignity, identity, safety, and overall quality of life, no matter how far along the disease takes the individual. Nance's "The HD Molecule" pictorially represents an elegant description of the gestalt of caring for families and networks affected by Huntington's disease. Dr. Ferrini's Huntington's Disease Plan from Edgemoor Hospital DPSNF importantly stresses the critical balance that must be struck between risk to self and others and freedom and choice for the person with HD and others. The need to develop relationships with persons with HD "to learn their habits, preferences, triggers, likes, dislikes, tendencies and responses and, through these relationships, attempt interventions which are effective for the individual and balance risks and benefits from the perspective of that individual," cannot be adequately stressed.

3. Maintain high quality nursing approaches to caring for the person with HD

A. Mobility

The person with HD brings a very important concern to the LTC settings with the high risk for falls with injury. The person with HD may have involuntary movements (chorea, dystonia, or myoclonus). These can vary from quite minimal, similar to twitches, to very severe thrashing movements.

In the early stages, the person with HD may have an unsteady, awkward gait but still be able to walk independently. Staff and observers may

be fearful of a fall and want to assist, but many persons with HD adapt their gait well and fall less than one would think. As the disease progresses, gait becomes more unsteady, and falls increase. Injuries often vary from none to minor abrasions, bumps and bruises, to more severe, such as fractures or subdural hematomas.

Persons with HD at this stage can benefit from adjustments to the environment. The maintenance department can apply some padding to the walls near the bed, pad the sharp corners, and add grab bars in bathrooms. It is best to try to reduce clutter and rearrange the room to allow more space, but caution should be taken as changes can cause agitation in the person with HD.

Occupational Therapy or Physical Therapy, or a Restorative Nursing program can assist with strengthening, balance and gait, or wheelchair training may be helpful. Some persons with HD can benefit from using a wheelchair or wearing a soft helmet, elbow or knee pads. Initial resistance to these strategies can be overcome by gradually introducing the wheelchair or protective equipment. For example, some persons with HD will begin to use a wheelchair or helmet that is left in their room. Wheelchairs can be modified with auto-lock brakes or anti-tip bars. Seatbelts may be helpful for positioning in the wheelchair. As with all restraints, the benefits versus risks of using a wheelchair seatbelt must be considered and documented. Specialized wheelchairs and other seating options are available, and include CareFoam™ chairs or Broda® chairs.

Bed side rails have the potential to harm persons with HD due to bumping and entanglement,

and persons with HD may try to climb over them when getting out of bed, increasing fall risk. Beds in low positions, with a mat next to the bed, may be a better solution if the person with HD is no longer walking independently. A wider bed, such as a full size or bariatric size, can also offer freedom of movement and possibly prevent injuries due to excessive movements. Putting the mattress on the floor is also an option, but should be used with caution due to the risk of staff injury from difficult ergonomics challenges.

It is important to include families, doctors and neurologists in the decision-making process as they are an important part of the care team. Medications may be helpful in controlling involuntary movements. Judicious review of medications can identify drugs with side effects that interfere with mobility or cognition.

B. Nutrition, eating and feeding

A person with HD, especially one with chorea, can require as many as 3,000 - 4,000 calories per day just to maintain body weight. Many require fortified foods and other nutritional supplements, such as Ensure® or Boost®, to help ensure adequate nutrition. Snacks should be offered for all diet orders (dysphagia diets, for example) and readily available at all hours with staff supervision. It may be necessary to have the refrigerator and cupboards locked so that the person with HD will not have free access to foods, as some will require assistance to consume snacks and others may have compulsive habits about food consumption or accumulation. “Food jags” or quirks are common where the person with HD seems to be stuck on a particular item, such as a certain food item or type of beverage. Whenever possible,

it is best to try to comply as redirection can be difficult for persons with HD.

A person with HD progressively requires assistance with feeding. It is best not to allow a person with HD to eat in their own room, as communal eating allows more supervision. Despite excessive spillage, persons with earlier HD may be resistant to staff assistance during meals. Adaptive equipment, such as plates with a ‘lip,’ and large handled spoons and forks, allow the person with HD to maintain independence. Nosey cups allow the person to drink without the need to tip the head back. Covered cups may also be useful. Clothing protectors with waterproof backing can help prevent wet, soiled & stained clothing. In later stage HD, feeding can take a long time. Staff members must be patient. While staff members may like to converse with the person with HD, they should be aware that distracting the person too much might lead to worse swallowing while eating.

Dysphagia is universal in HD. A speech therapy evaluation is invaluable in the management of dysphagia and most persons with HD require dietary adjustment. While they may resist this at first, most accommodate to alterations, such as thickened liquids and altered texture foods. If the person with HD refuses the altered diet, risks versus quality of life issues may need to be addressed. Tube feeding is an option that some may choose when swallowing and choking problems begin to affect the intake, and the person with HD is not able to eat enough calories to sustain body weight.

Tube feeding preference and advanced directives should be discussed with the person with HD and their families or guardian long before the need arises, and while the person can still

communicate their desires. This can take some of the pressure off the families/guardians to follow the wishes of the person with HD.

C. Communication

Communication in HD is hindered both by cognitive dysfunction (including language deficit and processing speed) and problems with phonation and articulation. It can be challenging to understand verbal communications; persons with HD are often aware of and frustrated by this inability to be understood. They may also have trouble comprehending information, particularly if it is complex. Persons with HD may be slow to process and respond. Because verbal fluency is reduced, it is often preferable to ask the person multiple-choice rather than open-ended questions. A simple communication board, with a few basic choices (such as hungry, thirsty, bathroom, soda, TV etc.) may be very helpful. A speech-language pathologist can often suggest strategies to improve communication.

D. Continence

With disease progression, persons with HD will begin to have problems with toileting and episodes of incontinence. Some may become overly preoccupied with toileting, spending long periods of time on the toilet or going into the bathroom numerous times when awake. This may initially start from the fear or anxiety of having an incontinent episode and then, for some, it may become an obsession.

A toileting program should then be attempted, first by gentle reminders, and then offering pull up protective undergarments which allow them to continue to be somewhat independent while protecting clothing from 'accidents'. A calm and gentle approach on the part of the staff can help





decrease some of the anxiety around toileting and incontinent issues.

Another challenge can occur when a person with HD is unable to transfer or sit on the toilet safely due to chorea or impulsiveness, putting both the person and the staff at risk for injury. Agitation and anxiety may increase and persist. Continue to gently redirect and reassure the person with HD. You might need to discuss with the physician whether medications that help reduce anxiety/agitation and obsessive behaviors are appropriate.

Loss of control of the bowel and bladder is a very challenging time for most persons with HD, causing anxiety and sometimes aggression. Staff approaches should be understanding, gentle and reassuring to help minimize the embarrassment and reduce anxiety.

E. Behavioral, Psychiatric and Cognitive Challenges typically seen in LTC

Behavior management of the person with HD, in the LTC setting, can be a challenging undertaking. Lifetime prevalence of psychiatric symptoms varies from 33% to 76% of persons with HD being affected. Treatment may improve overall functioning and quality of life.

Irritability and Aggression

Among the long list of behavioral anomalies exhibited by persons with HD, physical aggression appears to be the most problematic in the LTC setting, particularly because persons with HD may endanger staff and themselves if this behavior is not appropriately addressed. Irritability has not been shown to correlate with disease duration, cognitive or motor symptoms or functional impairment. From experience,

staff can develop a “fear based response,” as a fall out from difficult behavioral episodes that can become entrenched and difficult to overcome.

Initial strategies for the management of irritability and aggression should include identifying triggers for the behavior, such as hunger, thirst, fatigue, boredom, pain, frustration, changes in routine, and inability to perform certain tasks. It is important to understand that persons with HD may have difficulty regulating their responses to a given situation, leading to responses that are out of proportion to the stimulus. The person with HD may be remorseful following an irritable episode. Routine and consistency, along with appropriate pharmacological agents, can often diminish these types of issues.

Pharmacotherapy may be useful in controlling certain behaviors. Many HD experts use atypical anti-psychotics for urgent treatment of irritability or aggressive behaviors. Selective serotonin reuptake inhibitors and antiepileptic mood stabilizers can also be used. Benzodiazepines may be helpful adjunctive agents. Please refer to *A Physician's Guide to the Management of Huntington's Disease* (Third Edition) (available from HDSA) for a thorough review of pharmacotherapy.

Depression and apathy

Depression and apathy are distinct entities that often coexist in persons with HD and are significant issues within LTC settings. Depression is a mood disturbance, and the presence of sadness is the key identifying feature, while apathy is a disorder of initiation and motivation. Depression can occur at any stage of the illness, and is often coupled

with irritability. Appropriate recognition, and aggressive treatment of depression in a person with HD, can be critical and life enhancing. Like other behavioral manifestations, depression does not increase progressively over disease course, but may occur at any point. The assessment and diagnosis of depression may be difficult in HD, because persons with HD often lack insight into their mood state and it can be difficult to apply standardized DSM depression criteria. Changes in mood, appetite and sleep, or loss of interest in, or enjoyment of activities, or changes in personality may be more useful signs when evaluating depression. The motor manifestations of HD, including flexed posture, general slowing and loss of facial expression, can be confused with psychomotor retardation of depression. There is a poor evidence base to support clinical decisions for the treatment of behavior in HD. However, clinical case reports, and the expected side effect profile for a particular person with HD, can help guide the choice of drugs for HD-related depression.

Apathy is common in HD and presents unique challenges to caregivers. With disease progression, persons with HD often develop deficits in motivation, drive and the ability to self initiate conversation or activities. As in depression, postural and movement changes related to the disease can mimic apathy, providing additional challenges to diagnosis. There are no accepted pharmacological approaches to apathy. Environmental modifications and creative communication, combined with patience, can help overcome apathy. It is critical that staff remember that no response from a person with HD does not mean no.

Mental inflexibility, lack of awareness, perseveration

Mental inflexibility, rigidity in thinking, lack of awareness of deficit, and perseveration can also create unique challenges for LTC staff. It can be difficult for a person with HD to think flexibly and adapt to new situations or changes in routine. They often need to adhere to set patterns or routines to help compensate for these biological changes. These changes can hinder the ability of the person with HD to see another person's point of view or to become easily frustrated or irritated if their views or ideas are challenged. Persons with HD may suffer from a biologically based "lack of awareness" of their disease manifestations, and drawing attention to deficits can backfire. A proactive strategy that relies on structure, minimizes changes in routine, and reinforces the person's strengths can help to maintain a peaceful environment.

Persons with HD are prone to repetitive behaviors and can persevere or "get stuck" on ideas, topics, persons or situations. Gentle redirection, engagement in a new activity, and respectful use of humor can sometimes be enough to move the person with HD forward.

Executive dysfunction

Executive functions are sophisticated cognitive skills. They include the ability to manage complex tasks, shift gears, plan, organize, predict outcomes, learn from mistakes and multi-task. The inability to engage in complex cognitive tasks can be overwhelming for the person with HD. Daily structure and routine are critical to managing these deficits. It is important to recognize that comprehension abilities often remain intact long after the person with HD has lost the ability to speak. Hence, to avoid

isolation, it becomes critical to keep talking to the person. Persons with HD have difficulty waiting and controlling impulses. They may take longer to process information and therefore can benefit from more time and patience from their caregivers.

There are currently no proven treatments for the cognitive symptoms in HD. Caregivers must rely on creative strategies, including trying to respond quickly to requests, or provide a verbal cue for when they will be able to respond. Be patient and allow enough time for the person with HD to process what is being said to them.

4. Promote staff education

Staff education is the key to mitigation of harm. All experienced providers stress the need to educate staff through peer-support, literature review, and positive feedback in challenging situations, especially for persons with HD and nursing aides in closest contact with the issues. Preventing staff burnout, while caring for the person with HD, is a constant challenge that requires constant monitoring. Staff must be recognized, rewarded, and provided all supplies as needed. In-service trainings from HDSA are available to help provide ongoing education to staff.

5. Engage in care planning for the special needs of persons with HD

The long term care setting necessitates care planning. Good care planning effectively leverages each member of the caregiver team to facilitate meeting the heavy burden of need faced by those in care. The special focus in Huntington's disease is on safety; preservation of function; reduction of psychiatric morbidity,

joyful activity pursuits and meeting nutritional demands while avoiding aspiration. Problem and Response scenarios are offered in the Edgemoor references.

6. Develop liaisons with emergency departments and acute hospitals

It is critical in any LTC venue that the hospital to which the person with Huntington's disease will be brought in an emergency or in-patient care be familiar with symptom management and the manifestations of the disease. Acute illness exaggerates HD symptoms, such as chorea, depression, falls, and weight loss. Unless there is an informed reception, the likelihood of iatrogenic complications is high. Failing that, a possible ombudsman liaison, could fill the gap. Even in the case of large academic teaching settings, there is a major lack of knowledge in managing issues so familiar to veterans of HD care.

7. Facilitate the use of advance directives: when and what to consider

Advance directive discussions are different in HD than other neurodegenerative diseases. The clientele are younger, stronger, and may have support persons who view the slow mounting of infirmity as a battle that must be continued at all costs. Percutaneous endoscopic gastrostomy (PEG) tube feeding, for example, has a different prognosis in HD than with advanced Alzheimer's disease. The feeding tube may prolong life by many years. As HD special units have matured over the years, the number of residents with "do not resuscitate" (DNR)," do not intubate" (DNI), and even "do not hospitalize" (DNH) advance directives has increased greatly. It will be important to look





at the national picture of the Family Health Care Decision Acts to see in how many states surrogates are permitted to give medical orders for life sustaining therapy (MOLST) after capacity is lost.

There is a critical need for proper training of physicians and physician extenders in the high art of discussing advance directives. Unless the discussions are accompanied by trust, reassurance, a promise of comfort and accompaniment throughout the hardest parts of the journey, the significant other will opt for higher levels of care. This is easy to say but difficult to exercise.

8. Be attentive to palliative care needs of persons with HD throughout their care and to hospice options near the end of life

Focus here is on preparation for the inevitable death and how best to prepare the family and the person with HD for symptom management, hydration issues, bereavement before, during and after with the expectation that predictions are often proven wrong mostly by the protraction of the late terminal phase. Facilities are encouraged to recognize and begin discussions of palliative or hospice care much earlier for the person with HD.

9. Address institutional “cultural issues” as they impact the person with HD

Culture here refers to the “institutionalization culture.” The challenge in overcoming this stereotype is to successfully mainstream the person with HD into the routines of the LTC setting. In most cases it is successful, with persons with HD welcoming the routines, activities, and security that come with the better settings. For many, the years in the

skilled nursing facility or in highly attuned settings in the home retain meaning, dignity, purpose, and place. Others cannot make the transition or cannot weather certain horrific stretches in their stay and fail. This outcome proves to be very difficult for all parties. Here, mutual support groups are key, as well as more pointed limitation of care.

10. Maximize the quality of life for the person with HD

A. Need for structure and routine

The unique cognitive features of HD foster a preference for sameness in day-to-day and moment-to-moment activity, and a compelling desire to maintain it. Arranging the greatest degree of predictability possible is the essential element in the management of the person with HD. Persons with HD find comfort in consistency and power in routine. Overseeing their days is an ongoing exercise in eliminating surprises and constantly reminding them, both passively and actively, about what's coming next. Engineering sameness and predictability encompasses minimizing the number of caregivers caring for the person with HD, as well as bathing and brushing teeth in the same manner.

B. Therapeutic recreation and coping with boredom

It has been said that hunger and boredom will often get persons with HD into the most trouble. Luckily, meeting hunger needs should be relatively easy, but coping with boredom is more challenging. Since many persons with HD lose their ability to initiate, it is up to the environment, and people in the environment, to provide the necessary jump start. The good news is, once started with an activity, more often than

not, the person with HD can keep going; it is the getting started that is so hard. Activity staff can be very helpful in directly asking the person with HD to attend activities. The challenge lies in offering activities that are interesting and tailored to the often younger person with HD. There is a need for activities that are truly fun, unique, engaging and age appropriate. It doesn't have to be a lot of work, but may require more creativity and thinking outside of the box. Many persons with HD just want to feel "normal," and any activity or way to help that can capture that feeling, creates the most opportunity for success.

C. Non-pharmacologic therapies

Many of these therapies tend to either be under utilized or used later than necessary in HD. Physical therapy can be helpful in several aspects, including gait and balance training to minimize falls and preserve mobility, and range of motion therapy. Therapists can assist with determining the need for walking aids, a Broda® chair or a wheelchair. As cognitive deficits worsen, it can become more challenging for persons with HD to learn new exercises, so programs are best started early and repeated often. The goals for treatment should be long-term maintenance rather than short-term improvement.

Occupational therapy can help develop strategies to minimize the impact of impaired spatial awareness. Persons with HD tend to benefit from "more open space" and it is often best to err on the side of having a more sparsely furnished room in order to help minimize injury potential. This type of therapist can often make excellent suggestions for adaptive equipment, whether in the case of creative seating, sleeping arrangements or mealtime aids. All of these measures can play a vital role in day to day living for the person with HD.

Speech therapy can have a variety of roles. These include evaluations for dysphagia, education about dietary modifications, and strategies to help minimize the burden and risks often associated with mealtimes. Dysarthria and dysphagia are often prominent features of HD and appear to have some degree of correlation, hence, as speech is becoming more difficult to understand, there is a greater likelihood that swallowing may also be worsening. Treatment should be individualized to the specific communication needs of each person with HD during each stage of the disease. Ideally it should be ongoing and focused on functional communication. Some common skills to address include metacognitive linguistic skills (short term memory, temporal orientation, working memory), dysarthric symptoms (breath support, phrase breaking/pausing, over articulation), prosodic patterns of speech and rate of speech, pragmatic language skills (initiation, commenting, topic maintenance), and musculoskeletal massage. Speech-language pathologists can make suggestions for simple/low technology communication boards, or medium to high technology approaches such as iPad like devices or other electronic methods of communication. Such interventions will benefit persons with HD all along the continuum of decline.

Music therapy is another under utilized modality that often can open up new ways of communication in particular for people in later stages of the disease. It can be another method to help improve quality of life for the person with HD. This can be as simple as depending on a person's preference for music that is played in their room or arranging for a music therapist to meet with a person with HD.

Finally, a daily exercise regimen is recommended to help with strength and mobility. Preclinical research, in mouse models of HD, suggest an enriched environment that provides opportunities for physical exercise can improve function, adding quality years to a person's life.

11. Maintain a working knowledge of HD for LTC staff

Helping nursing home staff to take the “mystery” out of dealing with a movement disorder like HD doesn't have to be difficult. Contacting HDSA or any of the HDSA Centers of Excellence across the US, to obtain resources that are closest to you, is one way to get general free educational support into a nursing facility. Providing the opportunity for the direct care staff to share what is really working with a person with HD and finding ways to support their efforts can go a long way in keeping both the person with HD and the staff satisfied. There are no cookbook approaches and, like many other neurological disorders, it is important to keep listening, keep trying and not give up. Often times, what worked last week might not work again this week but may work again the following week. Due to the potential for staff turnover, it is often helpful to video-record in-person training so the recordings can be shared with different shifts or newer staff members.

Lastly, and perhaps most importantly, is helping LTC staff to see the person as an individual and not just a person with HD. Since HD can often mask one's true expressions, it becomes very helpful and makes care easier if LTC staff have a idea of what the person was like before the onset of HD symptoms, what brought them both joy and purpose, and then find ways to keep those needs alive. ■■■

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SUBCOMMITTEE REPORTS:

Education Subcommittee Report

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REPORT ON THE STATE OF THE FIELD

Huntington's disease (HD) is manifest by a long trajectory ranging from 10-20 years from onset to death, and the later years of the illness are plagued by severe disability and dependency. Late-stage HD strains familial and societal resources, and the systems of LTC provision are often not attuned to individual need.

People with HD, and those who care for them, often find the choices in long term care bewildering, inadequate and, at times, ill equipped for the unique challenges of HD. Availability, cost and eligibility requirements complicate the process of choosing and arranging services.

The Education Subcommittee of the Huntington's Disease Society of America (HDSA) Long Term Care Workgroup reviewed the published literature from 1975 to the present using the search engines PUBMED, Google Scholar, and OVID. A total of 19 papers were identified in this review (see selected references). In general, the literature review revealed limited information regarding LTC in HD with most papers being descriptive in nature, and not including randomized controlled trials.

Two major themes from the literature review echoed personal experience of Subcommittee members. First, it is clear that caregivers for those with HD are looking for guidance on the best way to care for these individuals and are seeking guidelines, specialty consultation and assistance. Although the literature review revealed that care guidelines are needed, specifics on how these might be developed, and what they might include, were absent. Second, while a need for specialized LTC settings for those with HD was identified, the overwhelming majority of those with HD and their families do not live close to a facility that offers such care. If they do, those facilities are often

full. Overall, there were an inadequate number of specialized facilities and a lack of confidence in existing facilities among LTC stakeholders in regards to HD.

In addition to the literature reviewed, information was collected from the providers and establishments who served those with Huntington's disease. The existing HD organizations including the HDSA,

Huntington Study Group (HSG), International Huntington Association (IHA), European Huntington's Disease Network (EHDN), and two local Australian Huntington's disease associations were surveyed. It was noted that HDSA provides the most readily available resources regarding LTC in Huntington's disease for a larger, national audience. These resources are summarized in Table 1.

Table 1. National and international organizations with interest in HD and publications available to assist lay people and professionals with LTC issues

Organization	Publication(s)
Huntington's Disease Society of America (HDSA)	<ul style="list-style-type: none"> • Family Guide Series to Long Term Care • A Caregiver's Handbook to Advanced Stage HD (currently out of print – awaiting update/revision) • A Physician's Guide to the Management of Huntington's Disease (Third Edition) • The Marker magazine: article on respite care (Spring 2011) • HDSA Caregivers' Corner web series: Palliative Care (November 2010)
Huntington Study Group (HSG)	<ul style="list-style-type: none"> • The Palliative Care And Late-Stage Huntington's publication (2010) • The End Of Life Issues Of Huntington's Disease (2010)
Australian HD Association –New South Wales	<ul style="list-style-type: none"> • "Making a Decision on Residential Care" and "Making The Transition From Hospital To Home."
Australian HD Association –Victoria	<ul style="list-style-type: none"> • "Living with HD-Future Planning" (December 2010) and "Nursing Care In Late-Stage Huntington's disease."
European Huntington's Disease Association (EHDA)	<ul style="list-style-type: none"> • No written resources
International Huntington Association (IHA)	<ul style="list-style-type: none"> • No written resources

Table 1. Continued from page 39

Organization	Publication(s)
American Medical Director's Association for nursing home medical directors	<ul style="list-style-type: none"> • Caring For The Ages publication had two articles “A Place For People With Huntington’s Disease” (Ferrini, 2012) and “Huntington’s Disease Can Surprise Caregivers” (Kilgore, 2008), but no specific resources or educational options through these organizations.
HDSA Centers Of Excellence (CoEs)	<ul style="list-style-type: none"> • Variable site-specific resources available at different sites, but for the most part, these sites seem to primarily rely on and refer to HDSA resources provided on the national web site.
The HDSA Social Worker Network	<ul style="list-style-type: none"> • Relates to HDSA Centers of Excellence, relies primarily on HDSA, but some site-specific resources.
HDSA chapters in states, cities or regions	<ul style="list-style-type: none"> • Chapter-specific resources such as pamphlets or educational materials designed for those in their service area, but also rely primarily on national HDSA resources.

GAP ANALYSIS

After reviewing the state of the field, the Education Subcommittee of the HDSA Long Term Care Workgroup noted several educational needs impacting the quality of LTC provided to those with HD. These needs include lack of care standardization, insufficient availability and accessibility of resources targeted for stakeholders, and access, regulatory, and financial barriers limiting availability and quality of services:

- Barriers in availability, eligibility, insurability, and financing of services limit options. Choices are limited for those who are younger, only have Medicaid, have psychiatric or behavioral problems, need more supervision than other care, and live in areas where there are few physicians or other professionals knowledgeable about HD. Those with HD often have extremely limited alternatives for care.
- Existing scholarly resources do not identify content areas or priorities for guidelines or educational resource development.
- Regulations in state and federal arena governing LTC can further act to limit availability of these services for those with HD.
- Interstate and intrastate differences in the availability of services make the development of national materials challenging.
- Even for professionals, “keeping up” with the resources to assist persons with HD is a major challenge and there is a lack of knowledge, reimbursement and options for persons with HD thus putting a strain on both families and care providers.
- A lack of “standards of care” or “best practices” in caring for this diverse and challenging disease. This deficit challenges providers who

are coping with a complex and rare illness which has many manifestations without guidelines about what expectations and standards are appropriate.

- A deficiency in accessibility of resources. The resources examined were often out of date or not readily available and were not consistent.
- There is a failure to utilize principles of adult learning in the development of materials for LTC. These principles place adult learners in an active role in the learning process, providing materials that are relevant and practical and immediately applicable for the problem being faced. Simply put, adults seek and recall information based on their perceived need at the time; materials should be accessible and directed towards the needs and preferences of seekers at the time and location where they need the information the most. Adults like to be engaged in learning, interacting with the material, and enjoy discussions and stories in various methods of delivery (e.g. written, visual and auditory). Adults need feedback (e.g. self-evaluations or activities).
- The reading level of resources varies – one size does not always fit all and although lay people may wish to read physician materials, physicians may prefer materials more geared towards their professional needs.
- Resource quality, updating and customization are deficient. Materials may be out of date as the criteria for various programs change, reimbursement policies change or advances occur in thinking about an illness. Developing and printing a variety of resources can be very costly, requiring reimbursement of experts to develop and review materials, and establish consensus, as well as in the actual printing of brochures, pamphlets, and hand books. With so much variation geographically, clinically, financially, and even in what the experts recommend, it is nearly impossible to develop materials that meet the needs of all audiences at all times.
- No widely available curriculum has been developed for educating providers. It was noted that traditional in-service methods are not effective, particularly in settings of LTC where staff need less of a textbook knowledge about the biology of HD, and more applied information about how to care for this particular resident at this particular time.
- Education may be too general and not practical. Sometimes educational materials focused on the genetics of the illness and testing, which, although interesting and important, are not as important in the middle and later stages of the illness where staff are more often looking for practical solutions to day to day problems.
- Individuals, both lay and professional, were often searching for help with a particular problem involving a person with HD. There was also a paucity of education on how to change individual behavior related to caring for a person with HD, and a lack of approaches and strategies to enhance compliance and quality of life.
- There is a need for information to be available quickly when it is needed and with minimal searching in a format that is quickly readable. (e.g. too many steps to search through linked PDF files on a website).
- There is no clinical research base in HD middle and later stages in regards to what works and what does not. There is little knowledge and

collaboration among professionals in LTC or those dealing with other psychiatric-neurological diseases (such as Parkinson's disease or brain injury) about HD.

- Policy makers have a poor understanding of HD. It was noted that those who make decisions regarding reimbursement, LTC regulations, or service provision, often did not know or understand HD, and the special needs of this population.

RECOMMENDATIONS TO THE FIELD

Short Term Goals

The HDSA Long Term Care Workgroup Education Subcommittee established the following recommendations for short term goals:

- **Incorporate findings with other LTC Workgroup Subcommittee reports**

Incorporate the Guidelines for Management of HD Resident in LTC, developed by the HDSA LTC Workgroup Care Subcommittee, into the proposed resources stemming from the Education Subcommittee.

- **Establish ongoing LTC Workgroup**

Establish a workgroup comprised of representatives from the LTC Workgroup Subcommittees and include additional HD providers from the community. This newly established workgroup's purpose would be to revise existing resources and develop new ones, including web-based materials that integrate with print ones. The workgroup would establish priorities, timelines and milestones to measure project progress and seek out experts in HD and LTC.

- **Develop an online provider list**

Develop an online provider list of physicians

and other professionals who treat HD, including in LTC settings. Begin by using the list of physicians collected during the distribution of *A Physician Guide to the Management of Huntington's Disease* by HDSA. The online provider list should also include authors and providers who work with HD, such as clinicians from HDSA Centers of Excellence or other identified HD care facilities. Ideally, this group would also serve as a community advisory team to answer surveys, provide cases, review materials, support research, and help develop, review and assure that resource development is applicable to real world situations across the US in various settings. Eventually, the online provider list would be available for interactive, shared projects, and to pose and answer questions from primary care providers or family caregivers in the field. It might also be used to help find pockets of experience in various geographical areas to assist in referral of those with HD who need LTC services.

- **Develop and update printed resources**

Using the new Guidelines for Management of HD in LTC Settings, developed by the Care Subcommittee, the information found in the *Caregiver's Handbook for Advanced Stage Huntington's Disease* (Pollard, 2000), and the existing HDSA *Family Guide to LTC*, the new LTC workgroup would develop printed resources in English and Spanish on the following:

- *HDSA Healthcare Provider Guide on LTC*

This resource would be specifically for health care providers and designed to meet the needs of LTC professionals in residential settings, including medical doctors, nurses, psychologists, social workers, and administrators (see proposed content in table 2).

– *HDSA Family Guide to LTC*

This resource would be specifically designed to meet the needs of in-home and family care providers (see proposed content in Table 2).

• **Develop a Residential LTC HD Care Kit**

Develop an HD Care Kit that would be distributed by an HDSA social worker, HDSA chapter or affiliate representative, or family member, to LTC facilities receiving a new person with HD as a resident. Sections of the HD Care Kit would be developed as part of a tool kit or as stand-alone pamphlets enabling customization of the kit. The sections would include the practical clinical topics from the HDSA Healthcare Provider Guide on LTC, in written and DVD form, designed for direct caregiving staff. In addition to providing specific care information, the HD Care Kit would also include:

- DVD based in-service about HD and specific LTC care needs
- Sample care plans, notes, and risk management documentation
- Person with HD profile card (customizable by caregiver: photo of the person, list of likes/dislikes, hobbies, favorite things, problems at home, etc.) to give the staff a snapshot of who they are welcoming to their facility. Would also include advance directives and psychotropic consents
- Contact list of HD experts, including: neurologists, fellow LTC medical directors, psychologists, social workers, etc., that the facility could contact when they have questions about caring for the person with HD (especially helpful if not near an HDSA Center of Excellence or social worker)



- Magnet (or other promotional device) with HDSA information for staff referral.

- **Develop an In-Home HD Care Kit**

A shorter or modified version of the Residential HD Care Kit developed specifically for in-home service providers. The sections of the In-Home HD Care Kit can be determined based upon the in-home provider's area of expertise.

- **Market HD resources**

Work with HDSA to disseminate new information and materials, as well as updates as they are required through various communication vehicles including: email to the HDSA list serv directly, to HDSA Centers of Excellence, HDSA Social Workers, and Chapters/Affiliates, as well as to all identified HD providers from the Medical Resources/Physician Listing Directory, and social media platforms (i.e. Facebook, Twitter, etc.). This is an ongoing objective especially as new resources are developed and released.

Long Term Goals

- **Develop specific training for HDSA Social Workers and other social work providers working with HD**

Establish a workgroup including representatives from the LTC workgroup, additional HD stakeholders and HDSA to develop a standardized LTC training tool for social workers both connected with HDSA and those in other settings.

- Develop a standardized tool that could be used when conducting in-services at LTC facilities across the United States.

- The LTC tool would incorporate the elements developed in the one-year goals, including the HDSA Healthcare Provider Guide on LTC, the HD Care Kit and the HDSA Family Guide to LTC.

- The modules in the tool would be fairly general, given variability in regions, but with sections addressing issues such as insurance.

- After the standardized LTC training tool has been developed, the next training tool would be a module on LTC for HD that would be taught to HDSA social workers as part of their continuing education initiative.

- It would be recorded and archived so that new HDSA social workers could receive training as needed.
- It would be available to other social workers or health care providers in the HD community who may not be associated directly with HDSA through the HDSA website.

- **Develop an HD LTC website or section**

Develop a free, user-friendly and readily searchable web-based HD LTC resource inclusive of the content mentioned in the previous goals and accessible to all stakeholders. This web-based LTC resource should ideally:

- Incorporate a robust search feature
- Include information sections (see proposed content in Table 2) geared to various audiences, both lay and professional, with appropriate reading level material

- Provide downloadable and printable resources such as:
 - Sample Skilled Nursing Facility (SNF) care plan
 - Downloadable pamphlets and fliers
 - Sample SNF policies and procedures
 - Sample risk management notes
- Prompt users who are LTC professionals to join the list-serv
- Incorporate interactive components, discussion boards, and shared stories. These interactive elements could include, but not be limited to, the following:
 - “Classify the stage of disease” – questions can be presented asking about current problems, tendencies and where assistance is needed, and the program will come up with a “stage” of HD with common problems and risks and needs at that stage.
 - “What happens next? What can I look forward to?” – this feature could allow visitors to enter data related to what is happening now with the person with HD and the program will tell them what kinds of problems may come up in the future.
 - “Risk assessment” instrument – presenting standard risk assessment questions like:
 - Does the person with HD smoke?
 - Have they ever burned themselves?
 - Do they follow directions?
 - Do they walk?
 - Do they have a lot of chorea?
 - Are they aware of their abilities and inabilities?
 - How many falls have occurred in the last month?
- Provide links to Nursing Home Compare and other federal/Centers for Medicare and Medicaid Services (CMS) resources on insurance and LTC.
- Provide information about equipment, with the manufacturer’s URL.
- Offer an archive of resources, including: videos, presentations, webinars, sample in-services for staff, etc.
- Provide stories or examples of situations common in the LTC treatment of HD, including testimonials from care providers, families, caregivers and patients, include comments or discussion threads, an “Ask the Expert” feature, and panel discussions of possible problems and suggestions about addressing those issues, or a “What do I do about...?” Ideally this would include information targeted to family members, as well as a section developed for professionals, addressing their varied perspectives.
- **Develop a financial plan to both initiate and maintain the web-based LTC resource**
 - The workgroup would work with HDSA to develop an operating budget addressing both the development and ongoing maintenance for the web-based resource. Funding opportunities would be explored and recommended.



Table 2. Content Suggestions for the HDSA Health Care Provider Guide on LTC, the HDSA Family Guide on LTC, and ultimately the proposed web-based resource. Note: the reading level and approach will be geared towards the specific target audience, whether health care providers or families of those with middle to late stage HD.

Recommended Topic Areas	How HD is Different than other Neurological Diseases?	Definitions of LTC and Descriptions of Options
Specific issues within topic areas or questions leading to discussion threads	<ul style="list-style-type: none"> • Young age of onset • Need for age appropriate activities • Genetics and risk of other family members, etc. • Focus of care is palliative not curative and maximizing functional status and quality of life 	<ul style="list-style-type: none"> • Home based services <ul style="list-style-type: none"> – Family caregivers – Hired caregivers – Home health • Skilled Nursing Facilities (SNF) • Intermediate Care Facilities (ICF) • Medicaid waiver programs • PACE • Continuing Care Communities • Assisted living facilities • Group homes • Board and care facilities • Day care settings • Veteran Administration (VA) hospitals and facilities • Hospitals, medical and/or psychiatric, while awaiting placement • Hospices

Table 2. Continued

Recommended Topic Areas	Issues Related to Selecting a LTC Setting	Managing Motor Symptoms
<p>Specific issues within topic areas or questions leading to discussion threads</p>	<ul style="list-style-type: none"> • What are the reasons people select LTC services (i.e.: ADLs, supervision)? • When is it time to consider LTC? • How do I evaluate and select a LTC service provider? • Strengths and weaknesses of various alternatives • What can I expect from various professionals serving my loved one? • What can I do to improve my relationship with the caregiver or the nursing home? • How can I evaluate quality of care? • When do I need a second opinion? • What do I do if the care provided seems to not be “good enough”? • Funding and eligibility considerations • What is in-home care and how do I qualify? • What is residential care and how do I pay for it? • CMS resources: Medicare, Medicaid, SSI, SSDI • What is hospice and how is this different than LTC placement at end of life? 	<ul style="list-style-type: none"> • Excessive chorea • Gait disturbances • Falls • Fractures • Bruises • Head injuries • Proper use of adaptive equipment

Table 2. Continued

Recommended Topic Areas	Managing Eating & Nutrition	Managing Difficult Behaviors – Mood Symptoms	Managing Difficult Behaviors – Conduct
Specific issues within topic areas or questions leading to discussion threads	<ul style="list-style-type: none"> • Dysphagia • Altered diets • Dealing with those who refuse mechanically altered diets • Choking • Aspiration • Swallow strategies and equipment • Recipes • Weight loss • Tube feeding <ul style="list-style-type: none"> – Making the decision: to tube or not to tube – Making the decision in advance – Making the decision in an emergency – Tubes for medications only – When to stop tube feeding 	<ul style="list-style-type: none"> • Depression • Suicidality/self-harm • Isolation • Antisocial • Apathy/abulia • Anxiety • Psychosis • Aggression • Perseverative thoughts and behaviors 	<ul style="list-style-type: none"> • Cooking • Smoking • Wandering • Nudity • Disinhibition • Entanglement • Reaction to restraints • Elopement • Irritability • Injury to others

Table 2. Continued

Recommended Topic Areas	Managing Dementia In HD	Advanced Planning/ End of Life Care & Important Decision Making	Legal Issues
Specific issues within topic areas or questions leading to discussion threads	<ul style="list-style-type: none"> • Assessing cognitive function • Assessing executive functioning • Apraxia • Memory loss • What does the person with HD understand? <ul style="list-style-type: none"> – What do I tell him? – How can I tell? – How should I talk to him? • What can she still do and what can't she do? • What decisions can she make for herself? • What contributes to poor decisions in HD? • What can I expect to happen next? 	<ul style="list-style-type: none"> • Assessing capacity • Making decisions about the future for yourself or someone you love • Ethical issues related to end of life decision making (judgment, prognosis, beneficence), • Advance directives, POLST and like directives • Speaking to the health care team about advance directives • Changing your mind 	<ul style="list-style-type: none"> • Rights and responsibilities • Surrogate decision making • Conservatorships • Guardianships • Power of attorney • Durable power of attorney • Representative payee • Practical considerations <ul style="list-style-type: none"> – When can I force something? – Deciding about tube feeding

Table 2. Continued

Recommended Topic Areas	The Role of Skilled Therapies in Mid to Late Stage HD	Adaptive Equipment	Dealing with Difficult Residents & Families
Specific issues within topic areas or questions leading to discussion threads	<ul style="list-style-type: none"> • Speech therapy • Physical therapy • Occupational therapy • Psychotherapy • Family therapy • Music therapy • Art therapy • Therapeutic recreation 	<ul style="list-style-type: none"> • Beds • Broda® chairs (pedal & lounge type with or without Y strap) • CareFoam™ chairs (with or without y strap) • Wheelchairs • Pommel cushion • Restraints • Walkers • Rooke boots • Merry walkers • Helmets • Padding • Adaptive feeding equipment, including various types of cups, plate guards and sticky mats 	<ul style="list-style-type: none"> • Cooking • Smoking • Wandering • Nudity • Disinhibition • Entanglement • Reaction to restraints • Elopement • Irritability • Injury to others

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Table 2. Continued

Recommended Topic Areas	Supporting the Caregivers	Special Handling & Expertise in Medications	Recreational, Social, Emotional & Spiritual Activities
Specific issues within topic areas or questions leading to discussion threads	<ul style="list-style-type: none"> • Self-care • Burnout-preventing, treating • Respite care, asking for help, support groups 	<ul style="list-style-type: none"> • Medication overviews • Behavioral recommendations 	<ul style="list-style-type: none"> • How do I engage the individual and fill the time?

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SUBCOMMITTEE REPORTS:

Policy Subcommittee Report

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REPORT ON THE STATE OF THE FIELD

The elderly and persons living with chronic illness and disability often need long term care (LTC). LTC includes medical, nursing, and allied health professional services, but it also involves personal assistance with basic and instrumental activities of daily living. Closely related to LTC are policies that pertain to employment leave for family members who are giving care, and respite care programs for them.

American LTC is underdeveloped and it is viewed primarily as a personal and family responsibility. In the United States, the provision of LTC is not considered an individual right or a societal responsibility. The lion's share of the responsibility for LTC provision falls on unpaid family members and other friends. Public sector services are considered to be part of a safety net. Access to publicly provided or funded LTC services is variable, and the services are not always comprehensive. They are meant to supplement family caregiving and tend to become available only as family resources are exhausted. In America, society is the LTC giver of last resort. This comports with American values to some extent, but it also results in a considerable burden on families, a substantial opportunity cost to the broader economy and society, and a situation of inconsistent access and poor continuity and quality of care in many cases. Few are satisfied with the status quo in the LTC system. It may, in the near future, become fiscally unsustainable and overwhelmed by the demand for services as the population ages and utilization of services increases. In the coming decade there will be policy changes designed to bring about significant restructuring of LTC financing and delivery. Whether these changes will benefit or burden persons with HD and their families remains to be seen.

The special needs of HD are often precisely those that the LTC system is least able to meet. Persons with HD are unusually vulnerable to the shortcomings of the current LTC system and current policy priorities. The characteristic trajectory and symptoms of HD call for the kinds of settings and services that are not always well supported by the current system. Government policy influences the LTC services that persons with HD receive in two main ways: (1) through payment for medical, nursing and other LTC services and (2) through regulation and standard setting for the conduct of health professionals and facilities. (These second aspects of policy will be addressed in the section on standards of care and professional education.)

THE POLICY LANDSCAPE

The policy dimension of LTC encompasses several facets and levels, including law (executive orders, legislative statutes, and judicial rulings), administrative regulations (HHS/CMS; state health departments), and implementation processes (e.g. nursing home surveys).

Policies affecting LTC come from all levels of government – federal, state, and local. The federal and state governments finance public health and disability insurance for eligible persons, but state government plays a very important policy role in the American health care system generally, and in the LTC system in particular.

Social Insurance

• Medicare

One major public insurance system is Medicare, covering those over age 65 and those receiving SSDI, mainly for acute and short term medical and hospital services. Medicare is financed by Federal taxes and subscriber premiums. It

covers hospital and rehabilitative services [Pt. A]; physician/medical services and professional services [Pt. B]; Medicare prescription drug benefit [Pt. D]; and the Medicare Hospice Benefit, which pays for comprehensive palliative care by certified providers for Medicare eligible persons with a life expectancy of 6 months or less. Medicare provides only a limited benefit for nursing home or home care, usually following a prior hospital admission and oriented toward rehabilitation following an acute illness. It is not designed to provide custodial care or many of the kinds of services that persons with HD require for a prolonged period of time.

• Medicaid

A second major public insurance system is Medicaid. It provides health coverage for the poor and those who are disabled and receive supplemental income benefits from the Social Security system. It is jointly funded by the federal and state governments. Eligibility for benefits under Medicaid is income and asset based, with each state determining the eligibility level. In most states an adult must be at or below the Federal poverty line to qualify, although the qualifying criteria for children is somewhat more generous. The 2010 Affordable Care Act (ACA) has provisions that will increase Medicaid eligibility above the poverty line beginning in 2014.

Like Medicare, Medicaid provides comprehensive coverage for acute care medical services.

However, since Medicaid reimbursement rates tend to be low, many providers will not accept Medicaid patients or do not practice in areas with high concentrations of Medicaid patients. Therefore, even though those eligible for Medicaid have insurance coverage, they often find it difficult to gain effective access to health

care services. On the other hand, unlike Medicare, Medicaid does pay for non-rehabilitative LTC services in the home, in community facilities, and in skilled nursing facilities (nursing homes). Indeed, Medicaid is the main source of insurance coverage for LTC for persons of all ages.

LTC is expensive. Many persons who are not eligible for Medicaid prior to needing paid LTC services eventually become eligible for Medicaid support after having exhausted their personal assets through a period of private payment for LTC services (this is often referred to as “spending down”). After these persons begin to receive Medicaid coverage, the LTC provider must accept the negotiated rate of payment for that patient. In virtually all cases, this is considerably less than the private pay rate. This pattern poses a hardship for many patients and families and serves as a financial barrier for many LTC providers. Persons with HD, like others who require prolonged LTC services, may experience this artifact of current federal and state Medicaid policy.

- **Social Security**

Health insurance pays providers directly, for the most part, or reimburses covered individuals for out-of-pocket expenses. But since access to health insurance coverage for LTC is limited, and benefits are not comprehensive, individuals often find it necessary to purchase LTC services on the private market. Hence income maintenance, particularly for those who are disabled, retired, or otherwise not able to realize an income through paid employment, is critical for receiving LTC. Private retirement funds, annuities, and savings factor in, but for elderly or disabled individuals,

the Federal social security system is a prime source of income maintenance support and hence impacts LTC in that way. Those with disabilities who qualify for income maintenance under Social Security also become eligible for Medicaid health insurance coverage. Recently, in response to educational and advocacy efforts undertaken by HDSA, the process for determining eligibility for Social Security Disability Income maintenance benefits has been improved relative to HD.

Trends in LTC policy

- **Institutional adaptation**

In recent years, LTC services have become more diverse and have improved in quality. Institutional care is more adaptive, flexible, and homelike. This is coming about both because of outcomes research which suggests that the quality of care resides in the built environment and the social atmosphere surrounding the patient, as well as in the competencies of the caregivers.

- **Rebalancing**

At both the federal and especially at the state levels, policy is attempting to shift the venue of LTC provision from residential institutions, such as nursing homes, to community-based walk-in services and home-based services.

- **Consolidation of LTC facilities and programs within a “managed care” (prospective payment) arrangement for Medicaid funding recipients.**

- **Emphasis on comprehensive case management approach for chronic illness and LTC.**

This is happening both in community-based LTC (e.g. the PACE program) and more adaptive and redesigned institutional settings (e.g. The Greenhouse model). But such

innovative models are currently financially threatened by state fiscal crises and the market pressures on private managed care plans.

- **Incrementalism and lack of comprehensive reform**

The LTC policy arena is failing to address universal and sustainable system financing for LTC. There is no insurance system for LTC for the middle class. This segment of the population continues to face tremendous financial risk.

GAP ANALYSIS

Policy in the context of HD

HD may be seen in the context of the broad array of severely debilitating conditions requiring prolonged periods of specialized and costly professional services. These conditions are increasingly prevalent in an aging society, and they are at the core of the challenge of LTC today. Alzheimer's, Parkinson's, traumatic brain injury, stroke, and many others fall into this category.

On the other hand, HD is also unique from the point of view of LTC, and there are several special LTC needs related to HD that are affected by policy. There are a number of key areas in which there is a significant "gap" between needs and available services or resources, or, in other words, there are a number of barriers to access to needed LTC that persons living with HD and their families face.

Priority LTC service needs in the context of HD

The following are among the array of services needed by persons with HD in LTC:

- Medical and nursing needs
- Rehabilitative needs
- Dietary





- Motor issues and assistance
- Special handling and expertise with medication
- Psychiatric needs and behavioral management
- Social needs and an age appropriate setting/activities
- Palliative care

Priority quality of life needs in the context of HD

Moreover, the quality of life and the quality of the LTC services persons with HD receive are affected by several additional values and considerations. Among these are:

- Access to services that are timely and medically, and psychologically or developmentally appropriate
- Equitable eligibility for benefits and services
- Admission and retention by facilities and service programs
- Geographical accessibility
- Adequate information and counseling
- Culturally appropriate care options
- Affordability
- Appropriate sharing of financial burden by individual, family, and society
- Reasonable out-of-pocket expenses: insurance co-pays and deductibles; non-covered services, drugs, equipment
- Quality of care (competence, appropriateness, timeliness, empathy)
- Quality of life (dignity, respect, agency, meaning, relationality)

- Individual rights (autonomy, informed consent, choice, control)

Social barriers to appropriate LTC services

Clearly, barriers to access experienced by persons with HD come from policies (broadly defined as above to include both financing and regulation) that give low priority to the settings and services most needed by persons with HD. But additional factors come into play as barriers to access as well, including:

- The person with HD or family's lack of financial assets
- Uneven geographical distribution of skilled and experienced HD care and therapy providers and services
- Lack of community awareness, support, and trust
- Stigma. Misinformation and aversive emotions
- Fear. Psychological and cultural ambivalence toward HD and to LTC services particularly in institutional residential settings
- Lack of adequate investment in the development of those rehabilitative and LTC technologies that meet the specific needs of persons with HD: special assistive devices to address motor and gait issues; or new behavior and affective management techniques to manage aggression or address issues of judgment and the like.

Implications of LTC trends for HD: a mixed forecast

The policy trend of moving LTC out of residential institutional facilities and into the home setting with community-based care and service delivery (rebalancing) offers benefits and burdens for

the HD community. The relatively young age profile of HD has made nursing home residence unwelcoming and undesirable socially and psychologically. However, there are many factors that make institutional residential care necessary at some point for persons with HD and their families. Appropriate housing and coordination of services can pose very serious obstacles in many community settings. The requirements of LTC for HD can exceed family capabilities at some point. Right now, the territory between inadequate in-home and community-based care, on the one side, and full residential institutional care, on the other, is large and unfilled. New kinds of service delivery models in the community setting that are affordable and cost-effective must be developed.

In part, this is due to the types of care that persons with HD require. HD care is relatively labor-intensive and behaviorally and psychiatrically oriented compared with some other conditions. These are not services given high priority in LTC policy generally speaking because they are both expensive and difficult to evaluate quantitatively and objectively.

And in part, this is due to financing systems. State governments are fiscally stressed by rising Medicaid costs. Institutional care providers are under tremendous pressure due to reductions in Medicaid reimbursement rates. The LTC experience of most persons with HD and families, at some point, will intersect with this situation. HD definitely presents a profile of need that is vulnerable to disadvantage in this financial and operational climate.

It is essential to the well-being of the HD community that a system be developed that reimburses LTC providers adequately to

support high quality of care, good quality of life, and respectful settings where the rights and dignity of persons with HD are honored.

Continuity of care, portability of benefits, and access to specialized LTC facilities are especially important in HD. Managed care systems, into which most LTC services will be placed, must be developed to address these dimensions of care. It is not entirely clear that the marketplace and regulatory finance policy will create incentives to orient managed LTC toward of persons with HD.

As noted above, institutional care is under tremendous pressure due to Medicare and Medicaid reimbursement rates. If providers have a case mix that is too inclusive of persons in need of non-rehabilitative (so-called custodial) care and services, they experience fiscal crisis. Policy makers are now gambling that capitated managed care arrangements will provide a market niche for the kind of services HD requires.

Outside of the catchment areas of specialized treatment centers, the access and quality of care that persons with HD experience will depend upon the fact that the types of services they need will also be needed by a large enough number of persons with other conditions so that a sufficient market demand will be created. Otherwise, persons who reach a stage of HD in which what they need is primarily custodial care in a residential facility will face a grim and uncertain future in the American LTC system.

RECOMMENDATIONS TO THE FIELD

- **Reduce barriers to access**

Advocate for rules and procedures that will remove access barriers for persons with HD to government insurance and social programs such as Medicaid and SSI disability

benefits. Work to eliminate discriminatory and medically inappropriate procedures that are barriers to HD access.

- **Develop comprehensive LTC reform**

As the next step in health policy, beyond the ACA, call for a concentration on comprehensive reform of LTC finance and delivery. A universal long-term insurance system should be developed so that neither severe disability nor personal spend down to impoverishment is required, as it now is for Medicaid.

- **Explore Medicaid waivers to improve HD care**

Advocate waiver provisions to shelter persons with HD from provisions unfair or harmful to their special situation.

- **Explore means of improving HD care through regional coordination**

Advocate for regional and even national coordination among state Medicaid programs to facilitate access to specialized programs and centers by persons with HD who come from out of state. Some conditions, like HD, often require care at such centers, which do not exist in every state. Medicaid reimbursement systems presently complicate such utilization.

- **Target HD needs through funding provisions in social insurance**

Advocate special funding provisions in Medicaid reimbursement policies to enable providers to give better HD care, such as special equipment and services (e.g. special psychiatric care, rehab, dietary care, equipment to improve quality of life).

- **Enhance private insurance options**

Develop HD specific guidelines for both policymakers and consumers relating to the creation of a private market for affordable LTC

insurance and commercial insurance products that would be affordable and beneficial to the HD community.

• **Realize the potential of the ACA to benefit the HD community**

Develop recommendations concerning the gradual implementation of provisions within the ACA that will affect LTC for HD. For example, provisions to restructure the health care delivery system that will impact LTC as well as preventive and acute care-chronic disease management; the Medical Home concept; and the creation of Accountable Care Organizations.



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