Caregiver Guide for Mid to Late Stage Huntington's Disease:

For Long-Term Care Facilities and In-Home Care Agencies

Huntington's Disease Society of America

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Principal Authors

Joanna C. Burgess MA, CCC-SLP, Speech-Language Pathologist, UC San Diego Health Systems

Bob Davis, RN/FNP, Family Nurse Practitioner, HDSA Center of Excellence at University of Virginia

Eileen Fogarty, CTRS, Terrence Cardinal Cooke

Ann Gaba, Assistant Professor, CUNY School of Public Health at Hunter

Sally Gorski, MA, CCC, Manager, Speech-Language Pathology Hennepin County Medical Center

Amy Holmes, LGSW, HDSA Center of Excellence at University of Alabama, Birmingham

Anne Leserman, LCSW, HDSA Manager of Community Services

Deb Kegelmeyer PT, DPT, GCS Associate Professor Clinical Health and Rehabilitation Sciences, Ohio State University

Anne Kloos, PT, DPT, GCS, Associate Professor of Clinical Health and Rehabilitation Services, Ohio State University

K. Michelle Knewstep-Watkins, OTR/L, University of Virginia Health Systems

Martha Nance, MD, Director HDSA Center of Excellence at Hennepin County Medical Center

Contributing Authors

Jessica McDonough, OTR/L, RC, Good Samaritan-Specialty Care Community

Amy Chesire, LCSW-R, MSG, HDSA Center of Excellence at the University of Rochester

Editors

Martha Nance, MD

Debra Lovecky, MS

Anne Leserman, LCSW

Seth Meyer, LCSW



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Table of Contents

Late stage care in HD 8

Introduction 8

Diagnosis of HD 9

Stages of HD 11

Involuntary Movements 11

Cognitive and Behavioral Changes 13

Social Services in Long-Term Care 18

Role of the Social Worker 18

Cognitive Concerns 21

Assessing Behavioral Problems 21

End of Life 22

The Role of the Nurse in Late Stage HD 24

Care-defining Aspects of HD 25

Nursing Care Plans 29

Home Care Nursing 31

Hospice 31

Conclusions 32

Physical Therapy and Huntington's Disease 34

Role of Physical Therapy in Assessment and Treatment of Persons with HD 35

Goals of Physical Therapy 35

Physical Therapy Evaluation 35

Physical Therapy Treatment of the Person with Middle and Late Stage HD 39

Managing Cognitive and Behavioral Issues 44

Occupational Therapy for a Person with Huntington's Disease 52

The Role of the Occupational Therapist in the Healthcare Team 52

Occupational Therapy Interventions 55

Preparatory Methods and Tasks 68

Conclusion 69

Huntington's Disease and The Role of Speech-Language Pathologists 72

Communication Impairments and Huntington's Disease 72

Cognitive Changes in Persons with Huntington's Disease 73

Augmentative and Alternative Communication 75

Signs/Symptoms of Aspiration 76
Behaviors: 77
Huntington's Disease and the Role of Nutrition 82
Nutrition and Huntington's Disease 82
Effects of HD on Swallowing 86
Consultation with a Registered Dietitian 86
Meeting Calorie Needs 87
Feeding Tubes and Regimens 88
Summary Guidelines for Dental Care 89
The Role of Recreation Therapy and HD 96
Exercise Programs 96
Smoothie Socials 98
Horticulture 98
Animal-assisted Therapy 99
Music Therapy/Live Music 99
Arts & Crafts 99
Computers 100
Hospice Care in Huntington's Disease 102
Recognizing the Terminal Stages of HD 102
Goals of Hospice Care 103
Family Dynamics in the Terminal Stage of HD 105
What Hospice Provides 106
Brain Donation 107
Appendix 111

Chapter 1

Late stage care in Huntington's disease



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Late stage care in HD

Introduction

Huntington's disease (HD) is a slowly progressive neurodegenerative disorder, a degeneration of neurons in the brain, that leads to: gradual changes in a person's ability to control the movements of arms, legs, trunk, tongue, and lips; changes in mental processing, learning and memory; and varying degrees of mood and psychological disturbances. This combination of symptoms, combined with the uncommon (but not rare) nature of HD, presents unique challenges in long-term care facilities and to those who care for persons in the late stages of the disease. This guide will provide you with the information and resources that you need to provide the best care to persons with HD – to work through the difficult symptoms to reach the person behind the disease, and to allow the person to function as well as possible each step of the way. The goal of care for the person with Huntington's disease is to promote independence, freedom of choice, and ability to function within the confines of safety for the person, the family, and the care team.

In this chapter, we will provide an introduction to HD, describing the typical symptoms and stages, and common treatments for the disease. In following chapters, we will discuss the kinds of things that the nurse, physical therapist, speech-language pathologist, occupational therapist, recreational therapist, nutrition therapist, social worker, and hospice team should consider when working with a person who is in the late stages of HD.

Facts and Figures

HD is caused by a defect in a gene known as "huntingtin." Though everyone has two copies of the huntingtin gene, anyone who inherits the defect in one or both of these genes will develop HD if he or she lives long enough.

The disease was first described by Dr. George Huntington in 1872. For many years, it was referred to as "Huntington's chorea," because of the involuntary movements that can be quite striking in some people. However, we now recognize that it is the mixture of motor, cognitive, and behavioral features of the disease that together lead to the increasing need for assistance over the 15-20 year course of the disease. For that reason, we now use the terms "Huntington's disease" or "Huntington disease."

It is estimated that about 30,000 people in the United States have HD. Because it is an adult-onset genetic disease, passed on directly from parent to child, we recognize that, for every affected person, there are 3-4 (or more) persons "at-risk" of carrying the defective gene that causes the disease. For instance, an affected person may have two or three children, or a brother and two sisters, none of whom are diagnosed yet, but any or all of whom might carry the defective gene.

HD only affects the brain. Other organs, such as the heart, kidneys, and liver, are not directly affected by the disease. In comparison with Alzheimer's disease or Parkinson's disease, two other neurodegenerative disorders that are often grouped with HD, persons with HD are usually younger and have fewer medical problems besides the neurological disease. The typical onset of HD is usually around age 30-40 years of age.

Because Huntington's disease is genetic and because each child in the family has a 50% chance of developing this devastating disease, the entire family should be considered the "patient" in the larger sense. Excellent care for a person with HD will be remembered by the children and passed on to the grandchildren, and will reduce their fear as they confront this disease in other family members, or themselves.

In this guide, we will envision the care of the person with Huntington's as a flower, with the person who has a diagnosis of HD at the center, surrounded by the family and extended family. The "petals" surrounding this core include the health professionals: doctors, nurses, social workers, psychologists, rehabilitation therapists, hospice, and other allied health personnel, who provide care. The ideal care setting for persons with HD, however, also includes the involvement and support of others in the care facility: kitchen staff to prepare special diets; maintenance staff to address the challenges to the physical environment posed by HD; nursing administration, to support the direct-care staff in their care. And, it has been found helpful to enlist the support – or at least awareness – of other groups, such as local law enforcement, emergency medical services, and even the state Department of Health, whose services might be required at some time.

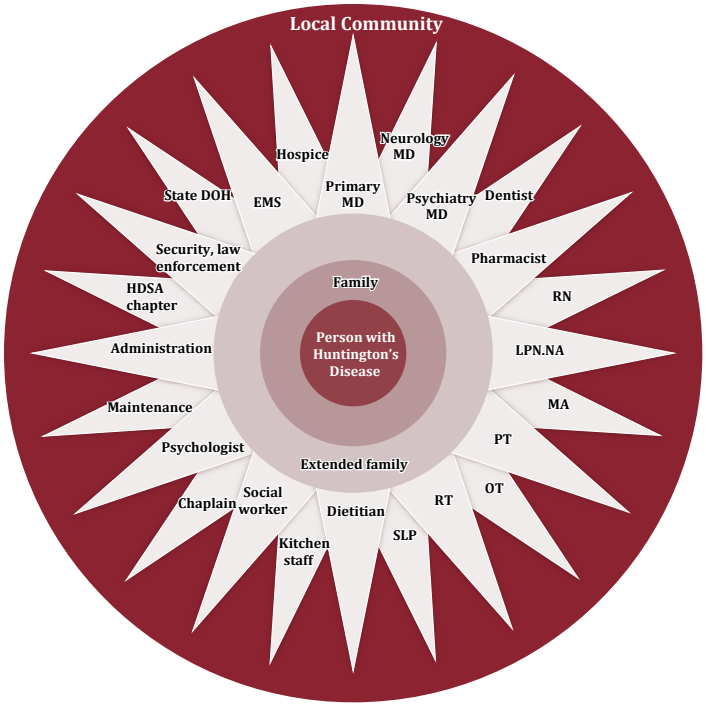


Diagram illustrating an ideal care community for a person with late-stage HD living in a long-term care setting, emphasizing the patient at the center, surrounded by family and extended family – whose experiences with a relative's course with HD have a great impact on their future attitude toward the disease and toward health care providers. The large variety of professionals who participate in the care of people with HD within and beyond the care facility are shown as "petals" around the patient/family core, and all fit within a larger (local) community. Abbreviations: MD=medical doctor; RN= registered nurse; LPN=licensed practical nurse; NA= nursing assistant; MA=medical assistant; PT=physical therapist; OT=occupational therapist; RT=recreational therapist; SLP=speech pathologist; HDSA=Huntington's Disease Society of America; DOH=Department of Health; EMS=emergency medical services

Diagnosis of HD

The diagnosis of HD is usually made by recognizing some or all of the symptoms described on page 1, in a person who is known to be at-risk for HD. Each child of a person who has HD has a 50% chance of inheriting the gene that causes the disease, and every person who has the gene abnormality will eventually develop symptoms of HD unless they die prematurely of something else. Although we said earlier that the average onset is 30-40 years old, about 5-10% of those at-risk develop symptoms before age 20, and an equal percent develop symptoms much later in life, as late as 60-70 years old. The disease often (but not always) seems to strike earlier in succeeding generations, a phenomenon called "anticipation." About 5-10% of persons with HD are not aware of an affected parent, either because of false paternity, adoption, or because they are truly the first person in the family to have developed the disease.

Symptoms

The symptoms of HD fall into three general categories.

Movement disorder. The most obvious symptoms have to do with movement. Loss of control of voluntary movements leads to clumsiness, dropping things, changes in handwriting, slurred speech, and

gait abnormalities. Persons with HD are often thought to be "walking like a drunk." Over time, there is increasing difficulty initiating and sequencing movements – at first, this includes complex activities such as a person might have to do at work, but later, there may be difficulty speaking, or performing movements that require more than one or two steps. There are no medications to improve coordination in HD, but it makes sense that continued physical activity will help a person to maintain function in the early or middle stages of the disease, and that a physical or occupational therapist could help a person or family to access the right equipment or develop strategies to optimize safety and quality of life throughout the course of the disease.



Obvious to others looking at a person with HD is the presence of involuntary movements, or chorea. The word "chorea" implies dance-like, flowing movements, but some people have more jerky or "tic-like" movements, while others have more writhing or twisting movements, which the neurologist might describe as "athetosis." Some persons with HD have little or no chorea, and some who have had moderate chorea experience substantial improvement of chorea with Xenazine™ (the only FDA-approved drug for chorea in HD) or other medications. Later in the course of the disease, many persons develop stiffness or abnormal postures or positions of the limbs or trunk, known as "dystonia."

It is important to know that persons who have HD may not be as bothered by their movements as are the people who look at them. The person with HD may lack awareness of their symptoms, called anosognosia. It is not always necessary to treat mild or moderate chorea if it is not bothering the person who has it.

Cognitive disorder. HD eventually leads to dementia, a loss of mental abilities. The earliest changes have to do with "executive functions" – making decisions, initiating activities, multitasking, completing tasks, and other complex behaviors. Learning new skills is difficult. Eventually persons with HD lack the cognitive skills to manage their own household affairs or even the tasks of daily living such as meal preparation, bathing, and dressing. Although speech may be very impaired in the late stages of HD, language function – the understanding of the meaning of words, and the ability to produce grammatical speech, is usually normal. Persons with HD typically recognize the people and things around them, even in the late stages of the disease. There are no medications known to improve the cognitive function in HD.

Psychological and psychiatric effects. The psychological and behavioral symptoms of HD range widely, and can often include depression, anxiety, irritability, obsessiveness and compulsiveness, perseveration, unawareness, impulsiveness, and apathy. Somewhat less common are substance abuse, sexual aggressiveness, premeditated violence, bipolar disorder, psychosis, or schizophrenic-like symptoms. Unlike the cognitive and motor symptoms of HD, the behavioral symptoms do not necessarily progress, but rather, may wax and wane during the course of the disease. These features of HD are treatable or manageable, more easily in some persons than others, using a wide variety of medications and nonpharmacologic strategies. You can read more about this and the diagnosis of HD in *A Physician's Guide to the Management of Huntington's Disease* available for downloading at:
http://www.hdsa.org/images/content/1/6/16692/HDSAPhysDeskRef_11_web.pdf

Stages of HD

Traditionally, Huntington's disease has been categorized into 5 stages based on symptoms and degree of disability. But this degree of detail is mostly used in HD research. Seldom do a person's symptoms fit neatly and precisely into a category. For the purposes of this chapter we will describe three phases of HD.

PHASES OF HUNTINGTON'S DISEASE			
	Stage 1- Early	**Stage 2-Middle**	**Stage 3-Late**
Symptoms	Subtle or mild symptoms, manageable with medications	Increasingly intrusive motor, cognitive, or behavioral symptoms	Poorly controlled movements
Location of Care	Home	Home with assistance or assisted living	Skilled nursing care
Activities of Daily Living	Independent	Not driving, managing finances, may need help with self-cares	Fully dependent on others for daily cares
Disability	No disability but may require work modifications	Unable to work	Unable to work
Emotional Cognitive	Depression, suicide risk	Mood changes, irritability, perseveration	Dementia, reduced ability to communicate

As with most neurological disorders, persons with HD will experience a unique progression of their disease. Some persons will have chorea early and cope well with it, showing few changes in their thinking, while others will have mood and memory problems early and uncontrollable movements later.

Involuntary Movements

Middle Stage: Chorea can be described as twitching or jerking movements of the head, arms, and legs. Early in the disease, affected persons are able to mask these movements into what might be considered an idiosyncrasy, like running a hand through their hair or repeatedly crossing and uncrossing the legs.

In the middle stages, walking becomes increasingly difficult. Walking requires the integration of vast amounts of information from our feet, legs, spine, eyes, and inner ear. This information is processed through areas of the brain that are impacted by HD. In the middle stage, the stance may be wider, and the walking speed slows. If the severity of the chorea increases and more muscle groups are involved, chorea itself can interfere with walking as the person's limbs flail and the torso heaves or twists.

A person with HD is often the youngest person in an assisted living or skilled nursing facility. Their activity, movie, and music tastes are likely to be quite different from those of the typical elderly nursing home population. They may be more interested in video games than in board games. Activities that require physical agility will need to be matched to the person's gradually changing ability to participate safely.

Falls and near-falls can lead to both minor and major injuries. The two most serious complications of falls are intracranial or subdural hemorrhage, and fractures. Traumatic brain hemorrhage can lead to delayed or subtle symptoms that can be mistaken for behavior changes in HD. Bone fractures can be difficult to heal when the person is moving constantly. Nursing care plans must balance the autonomy of the person, the care goals of the person and family, and the risk of injury from falls. The physical therapist can help design and encourage an exercise program that includes exercise and continued mobility, and assess the person for an assistive device. A walker may be a useful tool in reducing the risk of falling but must be used properly and continuously. Cognitive problems sometimes interfere with learning how to use a walker safely and when to employ it. You can read more about the types of walkers for HD in Chapter 4.

Late-Stage Disease: In the later stages, movements become increasingly difficult to control. Affected persons become fully dependent upon others for assistance with activities of daily living (ADLs), gradually lose the ability to walk, and eventually become wheelchair or bedbound. Those who are walking but fall frequently may benefit from using the same safety equipment used by skate boarders, such as knee, hip, and elbow pads as well as padded gloves. Helmets used by ice hockey players often come with partial face shields and provide some protection in the event of a fall. It may be a difficult to convince a person with HD to accept assistive or protective equipment; presenting options early, before their use is critical, sometimes helps.

Eventually walking becomes a hazard no matter what sorts of assistive devices are provided. The next step is some form of a wheelchair. Initially the person may have the coordination to self-propel the chair. One concern with the use of a wheelchair is the risk of tipping due to chorea. Anti-tip wheels help reduce the risk of tipping backwards.

Severe chorea may cause skin breakdown, bruising, or cause a person to fall out of bed. Securing the environment to avoid injury from pull cords, arm rests, bed rails, table and cabinet corners, and other furniture and equipment is important as the person moves into the late stages of the disease. Persons with HD may not be able to report pain or injury, so the health care team must be vigilant and anticipate problems. A queen-sized bed provides more space and can lessen the risk of falling out of bed. Reducing the height of the bed and padding the floor next to it can help to reduce injury from falls. Some facilities have used floor mattresses with cushioned surrounding bolsters. Foam pads on edges, corners, railings, and the wall can help reduce injury due to chorea. The occupational therapist, perhaps with help from the maintenance department, can think of creative solutions to these and other environmental issues.

Ensuring that the diet is high enough in protein to keep the skin supple and watching for signs of skin erosion and abrasion of prominent parts of the body such as the pelvis, elbows and feet, are the keys to prevention. Extra calories are needed since chorea expends energy. The more prominent the chorea, the higher the caloric requirements. Double portions of food and calorie-dense snacks are required to maintain weight. Likewise, ineffective self-feeding makes it difficult to determine how much the person actually consumes versus drops. Weighing the person regularly (weekly), providing extra portions, snacks and Ensure® or Boost® blended with ice cream to a milkshake consistency are good ways to maintain weight. Use the same scale each time you weigh the person with HD and note the weight of the wheelchair or other assistive device that the person uses.

As the disease progresses, chorea of the tongue, and difficulty controlling tongue movements, make moving food around in the mouth difficult. In addition, the gag and swallow reflexes are diminished making aspiration and pneumonia a risk. Persons with advanced HD will eventually need softened or puréed foods and thickened liquids. Liquids are thickened because regular “thin” liquids do not have a good “trigger” for safe swallowing as do more solid food stuffs. Persons with HD and their families need to make a decision on whether to have a feeding tube placed when safe swallowing becomes a problem.



Hopefully before this point, the person's wishes for a feeding tube are well known to the family and healthcare providers. Everyone must understand that a feeding tube does not guarantee protection from aspiration pneumonia. Bacteria and viruses live in everyone's mouth and upper throat. Through effective coughing and swallowing our normal bacteria and virus filled saliva is kept out of the lungs. If a feeding tube is considered, it should only be used when the person cannot take in sufficient calories to maintain their weight. Persons with excess chorea may be more likely to pull out their feeding tube.

Cognitive and Behavioral Changes

As is the case for other conditions that include dementia, the most challenging issues faced by the person with HD, the family, and the care team are often related to the disease's behavioral and cognitive effects. If a person is exhibiting challenging behaviors, it is important to document when and why the activity occurs. The care team can often develop environmental strategies to address difficult behaviors – rewarding good behavior, changing caregivers, scheduling the bath at a different time of day, or moving a resident to a different room – strategies such as these can sometimes be as helpful as sedating medications.

Middle Stage Disease: Not all persons with HD follow a precise sequence in the progression of their HD. For the purposes of this chapter, the middle stage for the cognitive and behavioral aspects of HD occurs when the person needs assistance from medications or their support network to support their deficits in memory, behavior, and thinking.

Persons with HD can become irritable and have little patience for waiting for something to happen or react out of proportion to an event. The irritation can erupt with an angry outburst. Safety is a priority for the person with HD and staff.

Due to mental slowing and apathy open ended questions should be avoided. "Do you want to change clothes?" should be replaced with: "Do you want to wear jeans or pants." "Do you want chocolate or vanilla pudding?" is a better alternative to the question, "Do you want a snack?"

Unawareness of symptoms or anosognosia is common in a person with HD. It is important for caregivers to learn to manage around the person's refusal to acknowledge their obvious HD symptoms.

There are methods to help decrease the likelihood of a confrontation which may cause anger and aggressive behavior. Keeping with a strict routine is best. Posting a schedule or a calendar of events and activities is a good idea. Maintaining one or two "key communicators" who work with the person decreases uncertainty and the amount of new information the person has to absorb. Limiting the number of people in the room trying to help the person with HD will ease their confusion. A person with Huntington's will likely be more comforted by the familiar and the routine. Most important is to give the person adequate time to absorb requests or questions. Slowed response time is common though the person with HD does understand what is being asked.

Depression and apathy are often difficult for mental health professionals to clearly sort out since they are often closely interrelated but one can distinctly exist without the other. Loss of interest in activities that the person once found fun is the hallmark of apathy. This apathy is a result of the changes in the brain and may not be related to depression. Though the person with Huntington's may no longer seem interested in previously enjoyed activities they should be encouraged to engage in activities, and sometimes brought to an activity.

Eventually, persons with HD go through a phase in which they become increasingly self-absorbed. They have trouble maintaining a sense of empathy and cooperation with those around them. Because of the ongoing changes in the brain, persons with HD lose their ability to control their emotions. Their wishes

evolve into demands. When these demands are not met promptly, the person with HD may become irritable and the irritability can escalate into aggression. Again, limiting stimuli in the environment, keeping to a rigid routine, and reducing the staff that interacts with the HD person to a few key communicators will foster and maintain trust. Persons with HD may have trouble processing stimuli and personal interactions. Keeping their living area as simple as possible, and the people with whom they deal to a minimum, may reduce agitation.

Unfortunately, medications will most likely be needed to keep the person and staff safe. While family members want to avoid sedating their loved ones, this must be weighed against personal and caregiver safety.

Late Stage Disease: Impaired judgment, unawareness of symptoms, apathy and withdrawal are all signs of increasing dementia.

In the later stages of the disease, the person with HD may change from someone whose demands must be dealt with immediately to a person who loses interest in the surrounding environment and people, and eventually with their own sense of self. It becomes hard to break through an increasingly hard shell of apathy. Irritability can evolve into aggression, eventually followed by despondency, then deteriorating into a need for total care. Persons who are unable to communicate may start to scream.

Initially, with prompting, the person with HD may be willing to take a bath or brush their teeth but over time they may be content to live in the same clothes for days on end and be indifferent to their own hygiene. Prompting by a key communicator may be helpful to have them accept a bath. The sedating quality of many of the medications used to treat chorea sometimes has the benefit of curbing aggression. Realize that tasks like changing clothes or toileting will take twice as long and require patience on the part of the caretaker.

In some cases the person with HD may become rigid as the disease progresses. This is called dystonia. Instead of chorea or “dance” with rolling movements, the person appears to make sudden jerking movements with stiff limbs. This makes dressing and bathing difficult.

Treatments

Many of the symptoms of HD can be treated with medications, even if the disease itself remains incurable at this time. Medications are primarily used to treat the psychiatric and behavioral aspects of HD, and the chorea. Medications that block or reduce dopamine levels in the brain (“neuroleptic” drugs and tetrabenazine) are commonly used to treat chorea. Depression can be relieved with any of a number of antidepressants, and anxiolytic medications can successfully reduce anxiety. Irritability, explosive behavior, psychosis, obsessive-compulsive symptoms, and apathy can often be reduced with medications. Sleep disturbances and pain are also treatable. On-site staff, therefore, should not hesitate to report these kinds of symptoms to the treating physician, so that appropriate medications can be prescribed. The physician will also initiate referrals to other specialists when there are particular challenges so timely reporting of new or changing symptoms can be addressed even when not treatable with medications.

The rest of this book will focus on nonpharmacologic strategies for managing HD in its late stages. It is important for all members of the health care team to have the attitude that HD is a treatable disease, and to communicate with each other if there is a problem. Often, the best solutions to a particular problem include both medications and a modification of the environment, a revised nursing care plan, an assistive device, or other nonpharmacologic treatment. On-site care providers also need to provide the physician and other more distant care providers with information about the response to a change in treatment.

HD is a condition best managed by a team of care providers, working together to improve the life of the person with HD. Remember that compassionate care in the late stages of this disease impacts not only on the person with HD, but on the person's direct and extended family. A good experience will go a long way to reducing fear about the disease in other family members, and conversely, a bad experience will be remembered for many generations.



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Chapter 2

Social Services in Long-Term Care



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Social Services in Long-Term Care

Role of the Social Worker

A social worker provides an important role in a long term care facility. A social worker is well trained to create a narrative history of the person and their family and work to improve the quality of life of the person along with assisting staff in the transition of the person entering a facility. Understanding the disease and the impact on other family members will help the social worker and inform the staff of the particular issues that confront family members with HD. The HD basics are explained in the introduction of this guide.



CASE STUDY

Cheryl had promised her husband that she would never consider nursing home placement like his father did to his mother when she was just 48 years old. Cheryl and Richard were married for 18 years when his symptoms started with increased irritability and irrational demands of their 2 children, then 10 and 11. Richard's behaviors worsened at home totally alienating his children and he soon lost his job due to disruptive behaviors with colleagues and not adequately managing his accounts. His episodes of rage resulted in several visits to the home from the police. Cheryl could no longer maintain the safety of herself and her children. Richard's falls were more constant and his incontinence a problem she could not manage. Richard needed care that Cheryl could no longer provide. The social worker at the care facility had multiple family members to assist in this difficult transition.

HD Family Dynamics

For a social worker, it is important to move beyond the basic explanation of a triad of symptoms challenging a person with HD. The impact on a family with a disease that is inherited and generational can be tremendous. The person with HD in your care has likely seen other family members progress and die from HD complications. They may feel guilty about the children they have that are at-risk for the disease. They may recall family members who committed suicide or spent time in jail or were in psychiatric facilities for treatment with unknown diagnoses. And because of their illness and their poor judgment, or inability to monitor their reactions to events, they may have alienated many or all of the people who once cared for them. All of this will have an impact on the person with HD as they enter long term care.

Some family members will be supportive and want to be involved in the care of their loved one. Others may not be able to face the progression of HD in their loved one or cope with their own "at-risk" status.

> "Sam used to visit his brother on a weekly basis but as the brother became more symptomatic, Sam's visits became less frequent and now he doesn't come at all and we cannot reach him by phone."

Family dynamics are complex and unique in an HD family. It is important to know who in the family is best able to be supportive at this time, knowing that adult children or siblings may not be able to help because of their own fears of developing symptoms, or because they are developing symptoms. For some family members, it can feel like "looking into a mirror" of what may very well be their own future. The best allies may be extended family, who are slightly more removed from the situation and possibly more able to be helpful. Knowing about the progression of symptoms in HD is important for the person in your care – but also being aware of possible early cognitive or behavioral changes for those at-risk may assist you in helping the family. In many ways, the person with HD's ability 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 nursing home staff. Families can become "overly involved" in the care of their person with HD and delay adjustment for both. Adult children and well spouses may feel that they have abandoned their loved one or "broken" a promise that they would never place their family member in a nursing home.



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 the "expert" on HD and can often feel slighted or ignored by nursing home staff who don't appear to listen and respect their degree of knowledge. Alternatively, they may grow tired and resentful of this burden to be the "HD expert."

As you are assessing the need for a Power Of Attorney, Advanced Directives or possible guardianship, it is vital to evaluate the various family members who may accept these roles. An "at-risk" family member (sibling or child of the affected person) may not be the right person to make future decisions for the person with HD.

Certainly all residents come to a facility with a past history, but families with HD likely will have a poignant story about the person with HD and the person they used to be. Take time to listen to the stories; they will make the person come alive, help you to understand the family, and give you the material you need to support both the person and the family through their course.

Care Team Members

All long-term care facilities have periodic meetings where they discuss issues about specific residents, and share information with each other. Some have care team meetings, others have behavioral committees, and others have special staff who do staff development. Whatever your facility calls these meetings, it is critical for all staff to be involved in understanding HD and helping the person make a good adjustment in what is a difficult transition.

Much has been written about providing a care team to assist in the care of a person with HD. Regular care plan meetings are vital to address a variety of issues.

The social worker's role will extend beyond the day to day management of symptoms and include a fuller picture to try to meet the multiple needs of a person with HD. A person with HD might require a nutritionist to address diet issues along with a speech-language pathologist to evaluate the swallowing mechanism of the person who may have excessive coughing or drooling that could lead to aspiration. Typically a neurologist might have seen the person to address movement issues. A psychiatrist or a psychiatric nurse practitioner may assist with medications for irritability or anger and help staff with orders for prn medications. OT and PT can assist the person with HD as they transition from being ambulatory to needing a walker to needing a wheelchair. These professionals can also offer suggestions about adequate bedding, proper toileting and safety in the environment where the person resides.

Are ancillary services available at your facility? What resources are at your disposal to get the best care for the person with HD without having to transport them outside of the facility? Like other persons with dementia, a change of environment, like transport to an outpatient doctor visit or a visit to the ER, could exacerbate their symptoms and challenge the ability of the person with HD to cope.

As the person with HD progresses in their disease, they will lose their ability to speak clearly and to let others know of their wants and likes. Some staff may be able to understand them or know them well enough to know their likes. Please read the segment in the Occupational Therapy section about a communication board. A simple laminated document can be made with bold pictures that a person can point to or nod at to get their needs met. It is important to not assume that if the person cannot communicate that they are not aware of their surroundings. Though a person with HD may be slow to respond (again related to changes in cognition), they still hear and understand what is going on around them.



It is important to provide a range of activities for a person with HD that are age appropriate and appropriate for the stage of illness of the person. Because of cognitive decline, the person may need extra encouragement to participate in activities. Initiation is often difficult for a person with HD though with encouragement the person can be convinced to participate. Working with staff and family to figure out the best motivators can lead to better results and picking activities of interest also add to the probability of participation.

Nutrition is a very important issue in the care of a person with HD. Swallowing issues must be regularly assessed. Making decisions about modifying diet should include family members. Persons with HD can have drastic weight loss and may need extra snacks to boost their calorie intake. Excessive weight loss may result from extra movements, metabolic changes and/or end of life issues. Discussion about the kinds of food that are easily swallowed versus foods that may cause choking should be considered. Are there arrangements that can be made if the family does not want to alter the diet, knowing the consequences of this decision? A discussion with family about the need for a feeding tube, along with knowing the wishes of the person with HD, will also likely be a point of conversation.

Persons with HD who enter long-term care facilities are likely to have falls. Independence is encouraged so continuing to walk is important to prevent muscle atrophy. But often the person with HD is unaware of their deficits (anosagnasia) and may forget to ask for assistance or be too impatient to wait for help to get up from their bed or a wheelchair. As falls are a concern at all facilities, it will be helpful to problem solve with the nursing staff to anticipate the fall issues and have an ongoing game plan. Understanding how the family has coped with this problem and what their expectations are will be helpful information as the staff approaches this issue.

CASE STUDY

Stephanie, age 44, lived at home with her husband and 2 teenage children. She was often left alone at home for many hours while her husband worked and the kids were in school. She was constantly reminded not to climb the steps while the family was away. She had trouble remembering the request from her family and wanted to remain the helpful wife and mother that she used to be. One day they found her at the bottom of the steps with a laundry basket by her side. She had attempted to carry the basket down the steps and start the laundry. It was time for more supervised care for Stephanie and a nursing home agreed to accept her. The family understood from their previous experiences with Stephanie's falls that the staff would need to work with her more intensely to monitor her activities and begin to use a rolling wheelchair.

Helping a person with HD learn to self-propel a wheelchair as part of the transition to a new environment can help to decrease falls while also taking advantage of helping the person learn how to use the chair, maybe before it becomes necessary.

Cognitive Concerns

Anosagnosia is a term meaning unawareness of symptoms. This is not a psychologically-based denial (although this can sometimes be seen, as well), but an "organic denial" that results from deterioration of brain cells. It often seems odd to notice a person with HD, who has obvious motor symptoms like gait imbalance or severe twitching of hands, feet and mouth, who repeatedly denies that they have trouble walking. They may even deny the existence of HD in themselves or of the disease in their family.

A common principle in social work literature is to "begin where the person is." Trying to convince someone with a brain disease of an issue can be frustrating and likely futile. It is important to work around issues like needing to bathe or change clothes rather than a direct approach that may cause lashing out or a verbal attack. This is a common problem in a busy facility where taking more time with a person with HD to accommodate their particular likes and dislikes can, in the end, create more calm and compliance than pushing an issue and having the person overreact. The response to the person's overreaction will likely be more time consuming and could put others at risk if the person lashes out.

Cognitive problems increase as the disease progresses and a person with HD may wander like other persons that have dementia. It is important to know if this is an issue for the person with HD so staff can be ready for these wanderings and not punishing of the person for something they cannot control. How does your facility deal with this issue?



Room placement for a person with HD must be considered. A person with HD will likely adjust to a quiet, less stimulating environment. Finding balance between a good roommate and the possible need to be close to the nurse's station for a quicker response to demands or fall issues needs to be considered. A room assignment should look at many factors other than just the age of the resident. A person with HD that prefers a quiet room may become agitated with a roommate who leaves the television on 24/7 even if the age of the resident is similar.

In most facilities that accept a person with HD, there are likely one or maybe two other persons with HD. It may also be important to educate other residents about HD and why a younger person may need nursing home care. Other residents may find sloppy eating and staggered gait as unusual and may have lost their own cognitive ability to be tactful and polite about someone else's disability.

Assessing Behavioral Problems

Behavioral disturbances are often the reason that long-term care is requested by the family or as a referral from a hospital after a short term inpatient stay (often psychiatric). As HD progresses, the person is not able to filter comments or emotions as they once could. Getting a clear picture of behaviors includes what was happening in the home. The family may have tolerated anger outbursts or inappropriate comments early in the disease but has less tolerance of these issues when other caregiving measures are required. Questions like, was the person on medication, were they taking their meds, did the family set boundaries on behaviors that were unacceptable, what measures were used to help the person be more stable psychiatrically, can help them ease into their next level of care. Expect that persons with HD that have behavioral problems will need to remain on a medication regimen. Altering their medications will likely

alter their behaviors and create disturbance in their environment. Learn to be a good advocate of the person with HD to urge staff not to reduce or increase medications for no apparent reason.

Another role of the social worker is to try to understand the full picture of the behavioral disturbances that may occur. What led up to a behavioral outburst? The person that shows preference to a caregiver that bathes him may overreact to a new person who tries to do the same task. It is important to not blame the person with HD for their behaviors but to try to understand the full context of the event. If a staff member feels threatened, it will be more difficult to have a conversation about the symptoms of the disease that has altered how this person typically responds.



CASE STUDY

A woman liked to go bed early (7-8 pm) in a very quiet and dark room. She was an avid pop drinker and went through multiple cans per day. The housecleaning staff came into her room to empty her trash in the early evening making lots of noise as her pop cans were transferred to a larger bag on the hard tile floor. The person with HD woke, very agitated by the sudden noises and attacked the worker with a dull butter knife resulting in several staff persons being needed to subdue her. She was sent to a psychiatric hospital and the nursing home refused to take her back. The facility failed to notice that persons with HD can react or overreact to stimulation (i.e. noise, light, activity) and that sudden changes in the environment can result in an increase in negative behaviors.

CASE STUDY

A woman's room was located near the kitchen of the nursing home. Around 10:00 am she would start yelling loudly from her room and was an annoyance to staff and residents. She had lost her ability to speak. It took some time for the staff to realize that the smells of the kitchen were making her hungry. Once they realized this, a mid-morning snack ended her screaming.

Involving a team approach when problems ensue will help all staff feel that they are being heard, as well as, understand who may have the best approach for the person with HD and how best to problem solve to move forward. Sometimes the least likely staff member has figured out a way to interact with the person with HD that will make sense to other staff. Sharing best practices with all front line staff will help to prepare them for future difficulties.

End of Life

End of life issues include timing for palliative or hospice care. Please read the hospice chapter for more detail. Hopefully the conversation about care at end of life happened prior to entering the facility or should be started as the person with HD becomes comfortable in their new surroundings.

References

HDSA Long Term Care Monograph, 2012

Chapter 3

The Role of a Nurse in Late Stage Huntington's Disease



Huntington's Disease Society of America

The Role of the Nurse in Late Stage HD

Nurses and nurse's aides play a pivotal role in the care of persons with HD in long-term care, because they are the eyes and ears of the medical team. A nurse who is knowledgeable about HD can ensure that care plans are written to ensure the safety and dignity of the person with HD, can identify medically important changes in the person's condition, relay information to the physician, and provide support to the person and family throughout the course of the disease.

The typical onset of HD is between age 30-50, and the course typically proceeds over 15-20 years. Thus, most persons with HD coming to long-term care will be somewhere between 40-70 years of age, younger than most residents in long-term care. Depending on the characteristics of the particular person and facility, persons with HD are sometimes better matched with a "young adult" or "head injury" population rather than with the general nursing home population or an "Alzheimer's unit."



Most persons with HD come to long-term care a decade or more after the onset of HD symptoms; neither they nor their families will be surprised by their diagnosis or the need for 24-hour care. Because this is an inherited disorder, the family may have had experience with the progression of the disease in other family members. They may, however, be exhausted after their years of managing at home alone, or in a strained household. The nursing team's first role is to assure the person with HD and the family that they will be safe and well-cared for, and that the family can remain involved in the life of their loved one. Of course, the nursing team can also learn much from the family about the person's likes and dislikes, and about their life prior to and after the diagnosis of HD.

If the nurse takes the time to talk with the person with HD and family, it will greatly enhance the relationship and the ability to care for that person over the coming years. Persons with HD often live in long-term care for several years, so the staff should anticipate a long relationship with the family. Persons with HD will show gradual changes over their time in the facility in their mobility, ability to feed themselves or be fed, continence, ability to communicate, and their ability to interact and participate in activities with others. By anticipating the changes that are coming, the nurse can ensure a smoother course for the person with HD.

The nurse can also serve as the on-site leader of a health care team that includes physicians (including the primary care physician, neurologist, psychiatrist, and palliative care/hospice physician), dentist, the floor nursing staff, rehabilitation therapists, support providers (including the psychologist, social worker, and chaplain), other health team providers (dietitian, pharmacy, laboratory), and other support services such as maintenance and administration. Each of these team members has a role in the care of a person with HD; the nurse is in the ideal position to know when their services are needed.

The overarching goals of nursing care are to optimize the person's comfort, safety, dignity, and autonomy. It is useful to consider those principles while designing care plans, and certainly as the care plans are implemented. Attending to a person's comfort includes addressing pain, emotional distress/depression, spiritual needs, facilitating leisure/pleasurable activities, and maintaining family involvement. Optimizing safety includes considerations of the physical environment – seating and sleeping arrangements, bathroom safety; safety while eating, and safety in human interactions with staff, other residents, and family. Dignity concerns include cleanliness, the ability to communicate, inclusiveness (for instance, including the person in decision-making and care plan activities), and preparing for the future, including death. And finally, autonomy means acknowledging and respecting the person's right to make choices regarding sleeping and eating habits and daily activities, managing the tension between autonomy and safety in areas such as smoking, sexual behaviors, and even the decision of when and how often to bathe. Conflicts between the affected person and the family can also arise. The person with HD should be encouraged to complete Advance Directives, so that their opinions and desires can be met even at a time when they are no longer able to speak for themselves.

Care-defining Aspects of HD

Several aspects of HD are important to the daily management of the affected person, and to the development of care plans. It is the combination of the motor, cognitive, and behavioral symptoms, in varying combinations, and progressing at different rates over time even in the same person, that most challenges the care team. Most of the individual problems in HD can be managed as they might be for a person who does not have HD, but there are some lessons to be learned from nurses who have cared for a large number of persons with HD.



Changes in Motor Function:

Involuntary movements (chorea) can lead to injury as a person hits the wall, falls out of bed, and bumps into furniture. Chorea is also the reason that a person with HD should never be subjected to a limb restraint; restraining a limb will not reduce involuntary movements, but rather, will likely cause agitation and increased movements, and will definitely cause injury. Extreme caution must also be used with any type of positioning or restraint device, including bedrails, lap or chest restraints, pommels, and seat belts. Chorea can cause a person to become unbalanced in a Hoyer lift, to be entangled in pull cords, or to be caught between the headboard and mattress in a bed. Involuntary movements can be misinterpreted as deliberate, with another resident or staff person saying "he hit me," when it was truly an involuntary movement.

CASE STUDY

The nurse manager on the Huntington's disease unit receives a call from a nurse at another facility. "We have never had a person with Huntington's disease before," she says, "and we need help! Mr. Johnson won't stay in his wheelchair. He gets up all the time, and then he falls. The aides are all afraid of him, because he lurches and swings his arms at them when they come to make him sit down. He falls out of bed, and once he got his arm caught between the mattress and the headboard and got a terrible bruise. He is always banging into things and getting bruises and scrapes on his skin. What can we do?"

The HD unit nurse manager replies, "Here are some suggestions. First, understand that swaying and moving his arms when he stands up is "normal" for persons with HD. Don't be afraid – those are involuntary movements, chorea. Persons with HD need a little more space. Consider getting him a wheelchair that he can maneuver himself, so he doesn't have to get up. Lower the bed closer to the floor, or put a wrestling mat next to the floor in case he does fall,

or get a queen-size mattress to give him more space. Pad the corners of his bedside stand or clothes chest, and consider padding the elbows, knees, or other parts that he injures when he falls. If he is really not able to walk at all, then look into a different seating arrangement, such as a Broda® chair or a heavy recliner, that might accommodate his chorea better than a regular wheelchair. Ask the doctor whether he might consider writing a prescription for Xenazine®, an FDA-approved drug for the treatment of chorea in HD."

Poor coordination of voluntary movements is universal in the late stages of HD. This can affect the person's lips and tongue, leading to difficulty with speech and communication, and to difficulty chewing and swallowing (dysphagia). It can affect the fingers, hands, and arms, so that the person has increasing difficulty with the tasks of daily living, such as dressing, bathing, brushing teeth, combing hair, attending to toilet hygiene, using utensils to eat – and also difficulty doing enjoyable activities such as writing, using the computer, performing music and art work, and many other activities. And finally, poor coordination impacts on gait and trunk control, so that the person has progressive difficulty walking, maintaining balance while standing, or controlling the trunk in a chair. Some persons with HD develop a "head drop." Movements often become ballistic in the late stages of HD, so that the person crashes onto the toilet seat or flings themselves out of bed when he rolls over, or springs out of the chair and loses his balance.



CASE STUDY

Sandy, a 52 year old woman who has had HD for 14 years, is having more difficulty with personal hygiene, speech, and walking. She hasn't fallen yet, but her family recalls another relative who died after a fall caused a blood clot on the brain. They want most to keep her safe, and to avoid choking and falls.

To address all these concerns, raised during a care conference, the care team does several things. The social worker meets with Sandy and her family to review her care goals and wishes, while her speech is still intelligible and her cognitive capacity good enough to make decisions about care. Because her chief desire is to avoid choking, she is provided with materials describing the placement and use of the gastrostomy feeding tube, and indicates that she would like a feeding tube if she reaches a point where choking is frequent or leading to medical complications. Physical therapy and occupational therapy train her to self-propel in a wheelchair, and she quickly adapts to scooting herself safely around the unit in the wheelchair. She refuses any assistance with personal hygiene despite obvious challenges until one day the aide offers to trim and paint her nails – if she will accept help with her shower. Excited about the offer, Sandy now showers with help 3 times a week, with a small reward each time (having her hair combed a special way, nails painted, a temporary tattoo, etc.)

Changes in Cognitive Function:

Persons with HD develop dementia, a progressive change in cognitive function. The cognitive change in HD is different from that of Alzheimer's disease, which nursing staff may be more accustomed to. Persons with HD generally are able to recognize others until very late in the disease, but they may have long processing times, so that the answer to a question may come a minute or two after the question is asked. Staff need to be patient and allow the person time to understand a question and formulate a response. By the end, a person with HD is often mute, and uses nonverbal cues to influence the world around him – often with behaviors that are viewed as disruptive or disturbing, such as screaming or kicking. Nursing staff who know the person well can make this stage much easier by anticipating likely needs or likes and dislikes.

CASE STUDY

Mr. B had become mute shortly before Alice began working at the long-term care facility. He had been in the facility for 5 years, and was nearing the end stages of his disease. After a week of caring for him, Alice was very frustrated. "He throws his food and grabs at me, she says. I wait until 8 to get him up, then bring him out to breakfast, where I have to feed him and I can't always get to him right away because I have other residents to help too. I tell him it will just be a minute, and then he grabs me and I think he deliberately knocks his tray over and throws his food. And then I ask him what's wrong and he just stares at me. I don't like this."

Mrs. B explains, "oh, my – didn't they tell you? He is really obsessed with getting his shower in the morning before he eats, always has been. He is an early riser, and they used to make sure he had his shower by 7 am, so he would be ready to eat. He can still take some finger foods, and so he doesn't like syrup and butter and ketchup on his pancakes and eggs and potatoes – just plain. And I think he is worried about our daughter. She is pregnant with her first baby, and there have been some problems with the pregnancy."

Alice changes her daily routine so that Mr. B can get his shower early in the morning, helps him with finger foods whenever possible, and schedules a surprise for him – a Skype visit with his daughter, who is on bedrest in her home in another state. Mr. B has a big smile on his face all week after that surprise!

Changes in Mood and Behavior

Some persons with HD have severe behavioral issues, while others do not. Depression and anxiety are common, and impulsiveness, perseveration, and loss of the sense of time are common and disruptive in the care facility. A minority of persons with HD have psychosis, hallucinations, or mania, but unawareness and denial of symptoms is fairly common. Irritability and aggressiveness are common, and may focus on a particular person (staff person or another resident) or activity (wanting cigarettes, or not wanting medications or baths). The nature and severity of behavior issues vary from person to person and also during the course of the disease, sometimes subsiding as the dementia progresses, but sometimes worsening as the person loses the ability to communicate verbally.



CASE STUDY

Sarah was one of the most difficult persons with HD that this facility had encountered. From morning to evening, she was either crying, yelling, interrupting, screaming, or repetitively asking the staff to get her something, put her back in bed, get her up in the chair, or call her mother on the telephone. A care conference was held with Sarah's family. The psychiatrist was asked to address her medications, and she started Sarah on a medication for anxiety and a mood stabilizer in addition to her antidepressant. The psychologist saw her weekly, and she was started on an afternoon exercise regimen off the unit in the physical therapy department. She was served first at mealtime. Nursing staff started her on a reward system, where she would get a small reward (tokens that could be added up to "buy" an extra can of soda or a special kind of soap or skin lotion) if she went an entire shift without yelling or crying. This approach worked reasonably well, although she still had days when the nurses resorted to the use of prn medications to reduce her anxiety and outbursts.

By the following year, her disease had progressed somewhat, so that she was no longer as disruptive. Medications were tapered down, and family became somewhat more involved, as visits were not as upsetting for them at this point. She died peacefully two years later.

Nutrition

Weight loss is common in HD, and has many reasons. It is important to have accurate weights on a person with HD, as weight loss can often be addressed by something that most people like to do – eating more! Careful attention to dental hygiene avoids additional dental complications that can make eating and communication even more difficult. The nurse can make life much better for a person with HD simply by ensuring that nursing assistants are obtaining accurate weights, and by notifying the physician, dietitian, or speech-language pathologist if weight is declining.

CASE STUDY

John had been homeless for several years before being admitted to the long-term care facility. He had no family involved in his care, and kept to himself, not interacting much with the staff or other residents. After a month or two, the aides became concerned. He was refusing food, and losing weight. He choked sometimes, but spent all his available money on Diet Coke, which he seemed to be able to drink without any difficulty.

John was seen by the speech-language pathologist, who found that he had swollen gums, multiple decayed teeth, and what she thought might be an abscess in one tooth. He choked on solid foods, but did well with softened foods. Dental consultation was obtained, and various teeth were extracted, capped, and filled. Nursing staff helped him attend to daily oral care. The dietitian saw him, and recommended high-calorie supplements. She also identified some food preferences and dislikes, which staff were then able to consider as they offered his meals. The chaplain met with him a few times and found that he had a strong religious background. Arrangements were made for him to address his religious needs, and after a while, he became more engaged in the activities of the unit. His weight rose to the low end of the normal range for his height, and he gradually reduced his excessive use of Diet Coke as he settled into the unit.



Family Support

Because HD is a genetic condition, each of the siblings and children of the affected person is at a 50% risk of developing the same condition. The nursing staff needs to be sensitive to this difficult aspect of HD, and nonjudgmental if at-risk family members are emotionally unable to participate in the care of the person with HD in the late stages. Helping these family members to obtain counseling, spiritual support, or medical care in the case of depression, may be beyond the nurse's job description – but could be terribly helpful to the person with HD, who likely needs the family's support in his declining days.

CASE STUDY

Lily had been in the care facility for 3 years. Her ex-husband visited rarely, and their two children were busy and in college. Nurses continued to provide updates to the ex-husband when he would call. One day, her daughter called. She wanted to visit and thought that perhaps they could plan a party for her mother's 50th birthday at the facility. The daughter had been too fearful to visit for many years, but had recently undergone genetic testing and now knew that she was free of the gene. She wanted to make up for lost time, now that she was free of worry about her own gene status.

The nurses provided some suggestions of things that Lily might like for her birthday (updated family pictures, some brightly colored socks, and a new comforter for her bed). They explained which food textures were safe for Lily to eat, and what time of day was best for her to have a party (in the evenings she was often tired and irritable; late morning was often her best time).

To surprise the family, the nursing staff made sure that Lily was dressed in her best clothes on the day of the party, and one of the aides helped her put on makeup and curl her hair. "This was my best day ever," Lily said at the end of the party. In the following months, her daughter came to visit every Sunday to read to her and work on a scrapbook of family pictures.

Nursing Care Plans

Understanding these aspects of HD can help the nurse to write an appropriate care plan for a person with HD, and to address, or at least watch for, any unusual needs or challenges. Table 1 shows some of the nursing care plans that may be relevant in HD, and point out how the nurse can include other members of the care team to optimize the quality of life for a person with HD. This is not an exhaustive list, as each person's situation is unique.

Table 1. Nursing Care Plans in Huntington's disease

Care plan	Special issues related to HD
Activity intolerance	Deconditioning in the late stages; inability to initiate activity due to apathy (related to dementia); involve physical therapy.
Alteration in bowel elimination-constipation	HD itself does not generally cause incontinence, but medications can.
Alteration in comfort: pain	Persons with HD may be unable to communicate that they are having pain; nursing staff must be attentive to nonverbal cues and to changes that should cause pain (bruises, skin tears, broken bones) even if the person is not complaining of pain.
Alteration in family processes	The genetic aspect of HD is particularly challenging; involve the social worker or chaplain.
Alteration in nutrition: less than required	Persons with HD may need an increased calorie intake, which may be difficult to achieve because of dysphagia or behavioral issues; involve speech pathology and dietitian.
Alteration in patterns of urinary (or bowel) elimination: incontinence	Persons with HD do eventually develop bowel and bladder incontinence.
Alteration in sensory perceptual	Persons with HD may have an unawareness of their own symptoms, and do often lose awareness of time, so need frequent reorientation.
Alteration in thought processes (dementia)	Dementia is universal in the late stages of HD and needs to be addressed.
Altered sexuality patterns	A plan needs to be in place for pregnancy prevention for women, and for safety and privacy for both men and women.
Anxiety	Anxiety is not universal, but may be severe in some persons with HD.
Behavior problem: refusing to eat/ bathe/other	Some may be fearful of eating certain foods or not like a particular staff member's approach; involve dietitian and SLP for eating issues.
Confined to chair with safety belt or positioning device	Restraints must be used with great caution; an occasional person welcomes a positioning or safety device.
Death and dying issues	Persons with HD and their families need to know if the end is likely coming soon (signs include weight loss or recurrent aspiration pneumonias in a person who is no longer walking or talking); involve hospice.
Dehydration due to reduced fluid intake	Dehydration is common in the terminal stages, particularly if the person needs thickened liquids; some can safely use a "free water protocol" prescribed by the SLP.
Difficulty swallowing	Nursing assistants are often called upon to feed a person in late stage, and need to be aware of the need for altered food textures, and the slowness of eating.



Disturbing to others: aggressive behavior	This is relevant in a minority of persons with HD, but is very disturbing when it occurs; utilize emergency services as needed, and ensure the safety of staff and other residents.
Grieving	Grieving is common in this disease, which produces a progressive loss of function; involve the chaplain or psychologist.
Hyperthermia	It may not be actual hyperthermia, but persons with HD are often hot, may be sweaty, may wear summer clothing and want the window open or air conditioning/fan on even in the winter.
Impaired adjustment/ family conflict	Demographic differences between HD and others in long term care, and family challenges may make adjustment to the facility difficult.
Impaired physical mobility/frequent falls/gait disturbance	Progressive changes in mobility are universal in HD, leading to potential injury from falls, and reducing independence; involve physical and occupational therapy, and introduce helmets, wheelchairs, or positioning devices before a crisis arises.
Impaired skin integrity	Persons with HD are subject to bruises and skin tears due to chorea, and to bedsores in the late stage when they are chair- or bed-bound; keep nails trimmed, pad corners and edges or body parts that tend to be injured, and treat existing injuries aggressively.
Impaired social interaction/ social isolation	Abandonment by the family is common, and progressive dementia and declining ability to communicate combine to reduce a person's ability to interact. Involve activities professionals, work to maintain family engagement.
Impaired verbal communication	This is a common issue as HD progresses; involve SLP, and take extra time to get to know the person before they become unable to communicate. Ensure that Advance Health Care Directives have been written before this time comes.
Ineffective individual coping/ spiritual distress	Persons may have emotional or spiritual crises at unexpected moments, related to personal or family anniversaries, specific functional challenges, or to interpersonal issues on the care unit; involve appropriate staff, depending on the details.
Knowledge deficit	Misunderstandings about certain aspects of HD persist even in the information/internet era. The Huntington Disease Society of America provides accurate information and support at a national, regional, and local level (www.hdsa.org).
Noncompliance	Some persons with HD are noncompliant with medications and other required activities such as bathing, and may need behavioral incentives to improve compliance; involve psychology and occupational therapy, physician or psychiatrist depending on the situation.
Polypharmacy	This is less common than in some older persons with multiple comorbidities, but it can happen; sometimes a medication important at one stage of HD can be discontinued later; involve physician and pharmacist.
Potential for infection	Aspiration pneumonia and bladder infections are common as dysphagia progresses and the person becomes immobile, and skin or joint injuries can become infected.
Potential for self-harm (suicide)	Remove medications from the room, sharp objects and weapons, consider 1:1 care, involve psychiatry.
Self care deficit: bathing, dressing, grooming	These kinds of self care deficits are universal by the late stages of HD; chorea and, in some persons, dystonia, can make it harder for staff to assist.
Sleep pattern disturbance	Sleep disturbances are common, and can sometimes be addressed by keeping the person more active during the day, and sometimes by adjustment of medications, reduction of medications, or addition of medications; involve activities professionals and/or the physician.



It should be evident that good care of a person with HD is complicated; by the same token, it is gratifying to the nurse or nursing assistant who can take a person with HD who is struggling at home, fearful, losing weight, perhaps abandoned by the family, and create a safe new home where the person can be clean, well-fed, socially supported, and engaged in the community. We will give some examples from a facility in Minnesota that has a 32-bed HD residential care unit.

CASE STUDY

A. After several years of frequent repairs to broken toilet fixtures caused by Sylvia's poor truncal control, with frequent falls and flinging herself onto a wall-mounted unit, after nursing staff reviewed with administration and maintenance staff their concerns, the facility installed floor-mounted toilet plumbing during its next renovation.

B. A nursing assistant took it upon herself to provide manicures to persons on the HD unit. Persons with well-trimmed and polished nails were less likely to scratch themselves or others, and took pride in their groomed appearance.

C. A person with HD went through a period where she was swearing and saying bad things about other residents frequently. The nurse contacted the physician, who determined that she was depressed and started treatment with an antidepressant. In the meantime, the nursing staff began an incentive process, in which she would get "points" if she went a whole shift without any disruptive behavior. At the end of the week, the points could be used for a reward (such as a small toiletry item).

D. A person with HD frequently screamed and resisted bathing, but occasionally seemed to be willing to have a bath. One particular nursing assistant had no trouble getting the person bathed, and it turned out that her secret to success was letting the person select between two different times for the bath, so that both she and the person had some control over the bathing process.

E. Nursing staff noted that a person with HD was losing weight despite their best efforts to feed her. They involved the dietitian, who determined that the calorie intake was as good as it could be, given how slowly the person was able to eat. They reviewed the person's Advance Care Directive with her mother, who confirmed that she did not want gastrostomy tube placement. The mother was encouraged to speak with a Hospice provider, which she did. The hospice team made sure that the family was able to visit, and helped nursing staff to ensure that the person was comfortable. She died peacefully during the night a few weeks later.



Home Care Nursing

Some persons with HD will be managed at home in the mid to late stages of their disease, generally by a devoted family care-partner. All of these family care-partners will need help at some point. Whether it is mobility issues, difficulty preparing high-quality altered-texture food, managing incontinence, communication problems, difficult behaviors, or the emotional stresses of late-stage HD that provides the greatest challenge, will vary from one family to the next. Support from a knowledgeable nurse can help the family to achieve its goal of maintaining the person at home.

Hospice

A nurse who has known the person with HD for a long time can help to recognize when a person is reaching the terminal stages of HD. In general, a person with HD who is no longer able to walk or talk is likely in the late stage of his or her disease. It is possible for a person to be stable in the late stage of HD. Clues that the person might be in a state of terminal decline include weight loss, recurrent aspiration pneumonias, or a serious hospitalization. In the terminal stages, hospice care can be very helpful for

the family who is managing HD at home or in the long-term care facility. Hospice has a strong focus on comfort and pain relief, engagement with the family, death planning, and support for the family after death.

Conclusions

In conclusion, the nursing care team is in a unique position to support a person with HD through the course of their disease. By recognizing the special features of HD that impact on a person's safety and quality of life, and creating care plans that address these issues, and by recognizing when other members of the medical care team should be involved, the nurse can allow a person in the late stages of HD to live with dignity and as much independence as possible.



Chapter 4

Physical Therapy and Huntington's Disease



Huntington's Disease Society of America

Physical Therapy and Huntington's Disease

Impact of Huntington's Disease (HD) on a Person's Function

- **Involuntary (chorea, dystonia) movements**
- **Decreased voluntary movement control**
- **Balance and falls**
- **Ability to perform ADLs**

Voluntary and involuntary motor impairments contribute to functional deficits and falls in persons with Huntington's disease (HD). Voluntary motor impairments include bradykinesia (slowness of movement), akinesia (delayed initiation of movement), apraxia, motor impersistence (inability to sustain a movement), diminished rapid alternating movements, and difficulties performing sequences of movements. Involuntary motor abnormalities in HD include chorea (brief, irregular movements) and dystonia (abnormal, sustained posturing of a part of the body). Chorea is typically seen first in the fingers, hands, and face muscles, and progresses to include all four extremities and the trunk. The most prevalent types of dystonia were reported to be internal shoulder rotation, sustained fist clenching, excessive knee flexion, and foot inversion. Younger age of onset is associated with relatively more dystonia and less chorea, whereas an older age at onset is associated with more chorea and less dystonia. Although chorea and dystonia are prominent features of HD, they do not predict the level of functional disability. Disability best correlates with voluntary motor impairments (e.g., bradykinesia, apraxia) and cognitive impairments.

Gait impairments begin early in HD and typically include slower gait speed, shorter and more variable stride length, a wider base of support, increased double support time, and increased trunk sway compared to healthy adults. Balance deficits usually occur in the early-mid stages of the disease and manifest as delayed motor responses to unexpected balance disturbances, and difficulties with tandem standing and walking and standing or walking with eyes closed.



As the disease progresses, falls occur frequently and often while the person is performing multiple tasks simultaneously, while maneuvering around obstacles on the floor, and while climbing stairs. Many factors may contribute to falls including balance problems, gait impairments (especially bradykinesia, stride variability, and excessive trunk sway), deficits in spatial perception, difficulties with saccadic (rapid) and smooth pursuit eye movements, cognitive deficits (decreased attention and ability to dual task), and possibly psychological changes (impulsiveness, impaired judgment leading to unsafe behaviors). The person with HD is likely to fall if he/she talks to family members while walking or if he/she carries something while walking or when climbing stairs. Impulsiveness leads to falls on stairs as persons with HD often approach stairs and begin climbing without stopping or slowing down. Family members will say that they appear to move rapidly onto the staircase as though propelling themselves over the edge of the stairs. This behavior may be due to impulsivity. The person with HD is then unable to safely manage the stairs due to motor and visual problems. Teaching the person with HD to "stop, hold the rail, step slowly" can help minimize impulsive behavior and allow them time to visually scan and optimally use remaining motor skills.

Balance and gait problems affect the person's ability to perform tasks such as reaching, getting up and down from a bed, chair, or toilet, stepping in and out of a bathtub, or going up and down stairs. Hand dexterity is often impaired and negatively impacts writing, dressing, personal grooming, and cutting food and using utensils for eating. In advanced stages, most persons will require assistance with all activities of daily living, relying fully on caregiver or nursing care.

CASE STUDY

Helen, a 50 year old woman diagnosed with HD 10 years ago, has fallen several times getting in and out of the bathtub. On one fall, she hit her head on the edge of the bathtub and sustained a laceration of her scalp that required medical attention. Physical therapy examination showed that she was unable to perform single leg stance. She reported that she fell when trying to step over the bathtub wall. The therapist recommended using bathtub transfer bench and instructed her to sit down on the seat, while both of her legs were outside of the tub, the same as if she was sitting down on a chair, and once seated, to lift each leg, one at a time, over the bathtub wall. After she started using the transfer bench, she no longer fell getting in and out of the bathtub.

Role of Physical Therapy in Assessment and Treatment of Persons with HD

Goals of Physical Therapy

- **Middle Stage**
- **Late Stage**

Physical therapy continues to be beneficial for persons with HD, even in the middle and late stages of the disease. In the late middle stage of the disease persons with HD typically remain able to transfer and walk but require assistance from caregivers and usually use an assistive device. Goals for therapy at this stage involve maintaining motor skills and any mobility that remains. (Box 1)

Persons in the late stages of HD are no longer able to support their weight without assistance and are not able to ambulate. They may bear weight during transfers but require maximum assistance to rise to their feet and return to a seated position. They are unable to purposefully change position in bed. Goals of physical therapy at this stage involve preventing negative consequences of inactivity. (Box 1)

Box 1: Goals of Physical Therapy

Middle Stage	Late Stage
Maintain the ability to: • bear weight, • take steps and • assist in rising from and returning to a seated position. • move in bed without falling out of the bed	• prevent decubiti, • maintain range of motion (ROM) and • encourage active movement.



Physical Therapy Evaluation

Before starting any assessment or treatment with a person with HD it is important to remember that his/her responses will be influenced by cognitive and behavioral issues as well as some unique motor problems. As with any client who has cognitive issues it is important to introduce yourself and explain each thing you are going to do. Persons with HD may be apathetic or agitated and delusional. Suggestions for dealing with these situations are addressed later in this chapter. Bradykinesia leads to a delay in initiating any movement including speech. If asked a question, the response by the person with HD will be delayed; you must wait at least 15-30 seconds to allow them time to make a response both motor and verbal. Additionally HD leads to a condition known as motor impersistence. Persons with motor impersistence are unable to maintain motor activity. If asked to close their eyes, they will not be able to keep them

closed for more than a few seconds during the middle and late stages of the disease. This impairment also leads to a propensity to drop things.

Body Function/Structure Impairments

As persons with HD become more and more immobile, it becomes increasingly important to assess range of motion and strength. Range of motion (ROM) can be difficult to measure in persons with severe chorea as the extraneous movements may prevent the tester from passively moving the limb to its full ROM. When possible, ROM should be measured with the person lying down and relaxed; in this way they do not have to focus on balancing in a chair. When moving the limb, be sure to fully explain what you are going to do before you start, then ask the person to actively move his/her limb. Follow the active movement with a slow, gentle passive movement. As in Parkinson disease, the major limitations tend to occur in the flexor muscles.



Strength is difficult to assess due to motor impersistence and cognitive issues. A lack of a strong isometric contraction could be due to either of these factors rather than any decline in strength. This prevents true manual muscle testing (MMT) at this stage of the disease. It is possible to assess overall strength by observing functional movements. Do they move their limbs through the ROM against gravity? If you place a cuff weight on the leg can they still use it to walk and do other activities? For the upper extremities, it is easiest to have the person try to pick up an object and put it on a shelf or hand it to you. By the later stages of the disease, persons with HD are unable to follow commands and strength assessment other than observation of spontaneous movement is not possible.

CASE STUDY

Lizzie, a 32 year old woman who became symptomatic with HD 6 years ago, is being seen for difficulty in doing housework and childcare. During MMT of her shoulder flexors, she initially demonstrates strong resistance and then suddenly gives way. This may be due to weakness or impersistence. When asked to place a gallon milk jug onto a shelf above shoulder level she is able to complete the task though her motion is jerky. Functionally she is exhibiting strength > 3+/5 while on the MMT she would be rated a 3/5. While she is able to lift the milk jug one time on the next repetition, she initially fails then releases it and has to try again. Motor impersistence would cause this inconsistent presentation of functional strength.

Chorea is an important impairment in HD and therapists should document when chorea is present, what limb it is present in and the size and speed of the movements. Chorea is rated in the trunk and each extremity and should be documented as intermittent or constant and rated as mild, moderate or marked (see scales for chorea in Box 2). Additionally note whether or not the chorea leads to safety issues or prevents functional movements. Chorea can interfere with the person's ability to feed, dress or groom himself/herself or participate in recreational activities. When chorea involves the trunk and lower extremities it can lead to falls from chairs, beds and during walking or transfers.

Eye movement is also impacted with HD and therapists should ask the person to look up, down and side to side to ascertain which motions are impaired or lacking. Deficits in eye motion should be considered when planning for mobility and activities of daily living. While visual acuity may be intact with or without corrective lenses, the inability to move the eyes to see and track objects severely limits independence in mobility and ADL's if it is not compensated. Ocular pursuit should be rated as to whether or not the person can complete the motion, only partially complete, or doesn't move the eye at all and should be tested in both the horizontal and vertical directions (see ocular pursuit scale in Box 2).

CASE STUDY

Richard, a 45 year old male with HD, became symptomatic 8 years ago and is complaining that he is constantly tripping over and bumping into things and shows evidence of multiple bruises and lacerations on his arms and legs. Physical therapy examination shows that his ocular pursuit is rated as partial movement in both vertical and horizontal directions. During physical therapy, the therapist asked Richard to slow down and to turn his head side to side and down to the floor and tell her what obstacles he saw that he either needed to step over or avoid when walking through the clinic. This strategy may help persons with HD to recognize the limits of their vision and improve their scanning. The therapist recommended that he apply this same strategy when walking in his home or out in the community. The therapist also advised Richard to reduce clutter, remove rugs or other objects that he might trip on, and maintain wide open spaces to allow safe walking in his home.

Suggested rating scale for impairments unique to HD

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For further information on the assessment scale, please turn to page 128.

STANDARD

[STUDY NAME]

0 0 UNIFIED HUNTINGTON'S DISEASE RATING SCALE '99 (MOTOR) 0 0

SUBJECT ID ____ VISIT NO ____

INITIALS ____ SITE NO ____ VISIT DATE ____ ____ ____ (MM DD YYYY)

I. MOTOR ASSESSMENT

1. OCULAR PURSUIT — Horizontal 1a. ____ Vertical 1b. ____
   0 = complete (normal)
   1 = jerky movement
   2 = interrupted pursuits/full range
   3 = incomplete range
   4 = cannot pursue
2. SACCADE INITIATION — Horizontal 2a. ____ Vertical 2b. ____
   0 = normal
   1 = increased latency only
   2 = suppressible blinks or head movements to initiate
   3 = unsuppressible head movements
   4 = cannot initiate saccades
3. SACCADE VELOCITY — Horizontal 3a. ____ Vertical 3b. ____
   0 = normal
   1 = mild slowing
   2 = moderate slowing
   3 = severely slow, full range
   4 = incomplete range
4. DYSARTHRIA — 4. ____
   0 = normal
   1 = unclear, no need to repeat
   2 = must repeat to be understood
   3 = mostly incomprehensible
   4 = anarthria
5. TONGUE PROTRUSION — 5. ____
   0 = can hold tongue fully protruded for 10 seconds
   1 = cannot keep fully protruded for 10 seconds
   2 = cannot keep fully protruded for 5 seconds
   3 = cannot fully protrude tongue
   4 = cannot protrude tongue beyond lips
6. FINGER TAPS — Right 6a. ____ Left 6b. ____
   0 = normal (≥ 15/5 sec.)
   1 = mild slowing and or reduction in amplitude (11-14/5 sec.)
   2 = moderately impaired. Definite and early fatiguing. May have occasional arrests in movement (7-10/5 sec.)
   3 = severely impaired. Frequent hesitation in initiating movements or arrests in ongoing movements (3-6/5 sec.)
   4 = can barely perform the task (0-2/5 sec.)

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(9/19/07) Page 1 of 3

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STANDARD

[STUDY NAME]

0 0 UNIFIED HUNTINGTON'S DISEASE RATING SCALE '99 (MOTOR) 0 0

SUBJECT ID VISIT NO

I. MOTOR ASSESSMENT (CONT)

		Right	Left
7.	PRONATE/SUPINATE-HANDS 0 = normal 1 = mild slowing and/or irregular 2 = moderate slowing and irregular 3 = severe slowing and irregular 4 = cannot perform	7a.	7b.
8.	LURIA (fist-hand palm test) 0 = ≥ 4 in 10 seconds, no cue 1 = < 4 in 10 seconds, no cue 2 = ≥ 4 in 10 seconds with cues 3 = < 4 in 10 seconds with cues 4 = cannot perform		8.
		Right	Left
9.	RIGIDITY - ARMS 0 = absent 1 = slight or present only with activation 2 = mild to moderate 3 = severe, full range of motion 4 = severe with limited range	9a.	9b.
10.	BRADYKINESIA - BODY 0 = normal 1 = minimally slow (? normal) 2 = mildly but clearly slow 3 = moderately slow, some hesitation 4 = markedly slow, long delays in initiation		10.
11.	MAXIMAL DYSTONIA 0 = absent 1 = slight/intermittent 2 = mild/common or moderate/intermittent 3 = moderate/common 4 = marked/prolonged		11a. TRUNK 11b. RUE 11c. LUE 11d. RLE 11e. LLE

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STANDARD

[STUDY NAME]

0 0 UNIFIED HUNTINGTON'S DISEASE RATING SCALE '99 (MOTOR) 0 0

SUBJECT ID VISIT NO

I. MOTOR ASSESSMENT (CONT)

12.	MAXIMAL CHOREA 0 = absent 1 = slight/intermittent 2 = mild/common or moderate/intermittent 3 = moderate/common 4 = marked/prolonged	12a. FACE 12b. BOL 12c. TRUNK 12d. RUE 12e. LUE 12f. RLE 12g. LLE
13.	GAIT 0 = normal gait, narrow base 1 = wide base and/or slow 2 = wide base and walks with difficulty 3 = walks only with assistance 4 = cannot attempt	13.
14.	TANDEM WALKING 0 = normal for 10 steps 1 = 1 to 3 deviations from straight line 2 = > 3 deviations 3 = cannot complete 4 = cannot attempt	14.
15.	RETROPULSION PULL TEST 0 = normal 1 = recovers spontaneously 2 = would fall if not caught 3 = tends to fall spontaneously 4 = cannot stand	15.
17.	DIAGNOSIS CONFIDENCE LEVEL To what degree are you confident that this person meets the operational definition of the unequivocal presence of an otherwise unexplained extrapyramidal movement disorder (e. g., chorea, dystonia, bradykinesia, rigidity) in a subject at risk for HD? 0 = normal (no abnormalities) 1 = non-specific motor abnormalities (less than 50% confidence) 2 = motor abnormalities that may be signs of HD (50% - 89% confidence) 3 = motor abnormalities that are likely signs of HD (90% - 98% confidence) 4 = motor abnormalities that are unequivocal signs of HD (≥ 99% confidence)	17.
18.	**Motor Examiner**	18. STAFF CODE

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Activity Limitations



In the middle stage of HD activity limitations are becoming more and more pronounced and a focus of therapy should be maintaining any independence in function. Useful tools in the middle and late stages of HD include the Timed Up and Go Test, the Physical Performance Test, the Tinetti Mobility Test and the Berg Balance Scale.

Persons with HD are at high risk for falls and typically require assistance for most activities. Use of the physical performance test can provide a good overall assessment of both UE and LE function within typical daily activities. The Berg Balance Scale, TUG and Tinetti mobility test are all recommended as measures of fall risk and mobility performance.

Physical Therapy Goal Setting and Treatment Planning

Clinical studies have suggested that exercise is beneficial for reducing impairments and maximizing function in persons with HD. Persons with HD can and do respond to physical therapy interventions. Studies have shown improvement in physical performance in middle stage persons. Psychological impairments such as apathy and depression can create challenges to engaging the person with HD in therapy, and cognitive issues can lengthen the time needed to teach exercise and mobility programs. Given these factors, goals should be set with longer durations to allow for time to engage the person with HD and work at a slower pace to compensate for cognitive issues. Appropriate goals focus on:

- maintaining function
- improving safety
- adapting the environment

Physical Therapy Treatment of the Person with Middle and Late Stage HD

Aerobic Exercise

Maintaining cardiovascular conditioning is important to the health of all persons, no matter their disability. Persons with HD should engage in aerobic activities for at least 150 minutes a week. One method is to encourage ambulation either with the assistance of staff or with an assistive device or a Merry Walker.® If ambulation is no longer feasible then self-propulsion of the wheelchair should be encouraged. If available, stationary bikes are an excellent means of providing aerobic activity. Care should be taken with bikes that require the rider to step over a bar and lift themselves up to the seat. Persons with HD are often unable to perform single leg stance and so are likely to fall if asked to mount this type of bike. A bike with a very low center bar and a seat at thigh height or a reclining bike with a standard chair seat that swivels is optimal for safe mounting.

Flexibility Exercises

Persons in the middle and late stages of HD are at high risk of muscle and joint contracture despite their choreic movements. They are at higher risk of decubiti due to the sheer forces that accompany the choreic movements while remaining in a stationary position for long periods of time. Flexibility exercises should be performed daily and should focus on moving the joints into extension and rotation. Trunk rotation and extension are key motions as they allow for upright posture, reaching, and stepping. Flexibility exercises should be done while supine or while seated in a supportive chair to ensure that the person with HD can maintain his/her balance with minimal effort while attempting to move his/her joints to end range. By the middle and late stages, active movements are often limited to very small ranges of motion, thus caregivers must provide active assistive or passive ROM exercises to maintain functional flexibility and prevent contractures and their sequelae.

Strengthening Exercises (postural, core)



Strength losses are evident in this phase of the disease and programs to maintain strength should be provided. The use of weights is typically not feasible given other impairments such as chorea and motor impersistence. Chorea can lead to the person with HD striking themselves with the weight or the weighted extremity. At this stage, a functional strength training program is most appropriate. For lower extremity (LE) strengthening, repetitive sit to stand transfers can be quite beneficial both for increasing muscle force and improving function. Persons with HD have high metabolic demands and difficulty maintaining their weight so exercise programs can help to improve appetite but should be kept at a level that won't lead to even more weight loss. Functional activities such as lifting objects into drawers and onto shelves help maintain upper extremity strength. Additionally, having the person with HD push and pull a wheelchair or other wheeled object can help work back and chest muscles. Adding weight to the chair (wheeled object) is a method to ensure that this activity provides enough resistance to be beneficial.

Balance Training

The ability to maintain a narrow stance and stand on one foot is impaired early in the disease process. By the middle stage trunk control is severely impaired. Additionally persons with HD lack insight and therefore do not understand that they are at high fall risk and don't modify their movements to ensure their safety. Balance work focuses on balancing with a narrowed base of support while doing dynamic movements such as turning the head and moving the arms. Additionally practice walking, with only the assistance that is necessary for safety, helps to improve balance during mobility.

Gait

Maintaining the ability to ambulate for as long as possible is absolutely essential to health and well-being. The occurrence of pneumonia, sepsis and death are directly correlated with the cessation of active movement, with physical inactivity and poor diet often being one of the leading causes of death. Persons with HD are at high risk for inactivity and poor diet. Therapists are the front line defense against inactivity and should carefully weigh risk versus benefit in making recommendations regarding ambulation and when to transition to wheelchair use. The presence of chorea leads caregivers to believe that the balance of the person with HD is worse than it is. Utilize accepted fall risk and balance outcome measures to gain an objective assessment of safety to ambulate. Some assessment tools that have been validated for use in Huntington's disease include the Timed Up and Go (TUG) test, the Berg Balance Scale (BBS), and the Tinetti Mobility Test (TMT). Additionally, observe ambulation in the person with HD's natural environment. If safety is an issue, consider the use of an appropriate assistive device. The device that has been shown to be safest for use with individuals with HD is the rollator or four-wheeled walker. Try to plan therapy such that you can observe the person with HD walking when they are not aware that you are there. Safety issues are more readily identified when the person is not trying to demonstrate their walking skills to a therapist. If walking with the assistive device is no longer possible, use of a Merry Walker® can prevent inactivity and maintain lower extremity strength and weight bearing for transfers. Strain on caregivers is minimized when the person with HD can bear weight through his/her lower extremities and provide some assistance during transfers. Walking safety reminders include slowing down when approaching obstacles, focusing attention on walking, and not talking and walking at the same time. In therapy, identify the key issue and try giving verbal cues such as "see that chair, slow down" and if the person with HD responds appropriately teach family and caregivers to use this cue. Simple, short cues work best. Constant repetition can lead to carry-over such that the person with HD learns to slow down without the cue. The ability to tandem walk and to stand on one leg becomes severely impaired by the middle to late stages of HD. In middle stage, have the person with HD practice balancing on one leg and with a narrowed base of support. Teach caregivers to use a gait belt for safety. Encourage use of appropriate assistive devices and continuation of gait even if assistance from staff is necessary. Ambulating 3 times per day until mild fatigue is an appropriate quantity to prevent further decline in health and function.



✔ **Please go to https://www.youtube.com/playlist?list=PLLQmMRDsNEY2_6crhI-z9yL9FCcOFLI-VD to view Clip #8 – Using a Gait Belt**

CASE STUDY

John, a 55 year old male with a 12 year history of HD symptoms, is experiencing 3-4 falls every day, mostly when he is walking. Physical therapy examination findings were a slow and wide-based ataxic gait with variable step lengths, path deviation to both sides, and moderate medial-lateral trunk sway. During 180 degree turns he was unsteady and once needed assistance from the therapist to maintain balance. He stated that when he held on to his wife's arm or pushed a shopping cart he felt steadier. The therapist instructed him on the safe and proper use of a rollator walker and observed significant improvements in his stability and gait speed. Three months later after obtaining and using a rollator walker, John reported that he had no falls.

✔ **Please go to https://www.youtube.com/playlist?list=PLLQmMRDsNEY2_6crhI-z9yL9FCcOFLI-VD to view Clip #7 – Use of a Four Wheeled Walker**

Key Points Gait Training

- Utilize accepted fall risk and balance outcome measures to gain an objective assessment of safety to ambulate.
- Have the person with HD practice balancing on one leg and with a narrowed base of support.
- Teach caregivers to use a gait belt for safety.
- Encourage use of appropriate assistive devices.
- Encourage ambulation to be performed at least 3 times per day; walking until the person with HD experiences mild fatigue.

Assistive Devices, Orthotics



ROLLATOR WALKER

A four-wheeled rollator walker is the safest ambulatory device to use for persons with HD compared to canes and other types of walkers. When using a rollator walker persons with HD ambulated with the fewest stumbles and had a gait pattern most closely matching a natural gait pattern. Standard walkers led to slow velocity, short step length and increased the risk of stumbles and falls while walking and turning. Use of the popular, two-wheeled walker also led to more stumbles and falls when turning and slowed gait speed and decrease in step length and step regularity. Use of a cane did not prevent stumbles and falls when turning but did not negatively impact gait for the group as a whole. It may be that the use of a cane is appropriate in early stages of the disease but insufficient to offset gait deficits in later stages. The majority of persons with HD can learn to use the rollator walker with minimal instruction. The use of the hand brake is not necessary for daily ambulation and does not have to be mastered by the user. Additionally rollator walkers are relatively inexpensive. They can be purchased for very close to the person's out of pocket cost when insurance covers a device. Prescription of an assistive device should be based on safety and well-being of the user and should not be dictated by third party financial pressures. An open access article on assistive devices and Huntington's disease is available at: http://www.plosone.org/article/info%3Adoi%2F10.1371%2Fjournal.pone.0030903

Orthotics are not commonly prescribed for persons with HD and should be considered on an individual basis. Arch supports and orthotics worn inside of a shoe that position the foot in subtalar neutral may improve ankle motion and lower limb stability. If ankle dystonia is a problem, a heel wedge and/or lateral wedge may improve ankle movement in the inversion/eversion direction and an ankle foot orthosis (AFO) may improve movement in the dorsiflexion/plantarflexion direction. A custom made shoe inlay may also be helpful for persons with clawing of toes during walking. Persons with HD may experience changes in balance or other motor skills when using a new orthosis. Therapists should perform careful assessment of ambulation and balance prior to and after prescribing orthotics to address and prevent safety issues related to the secondary impact of these devices on other areas of the body.

Footwear

Footwear can lead to poor walking patterns and falls. The ideal footwear provides support to the foot and

does not have a high heel. Shoes with a low heel can be worn as long as the heel is broad and provides a large area of support for the heel of the foot. Shoes should have contact all the way around the foot to ensure that the heel of the foot cannot slide off of the shoe. Mules, backless sandals and scuff slippers are not recommended.

Protective Techniques (helmets, knee and elbow pads)

Persons, who fall or are at high risk of falling, even when sitting or lying down, need to wear protective gear to help minimize or prevent injuries. Soft helmets can protect the head from lacerations and abrasions and provide some cushioning in the event of a fall. For persons who tend to land on their knees, pads can prevent abrasions and minimize bruising. Some persons bang their elbows on furniture due to their chorea and may benefit from elbow pads to protect from injury. Persons with HD tend to hit their heads when falling and are at lower risk for hip fracture than the elderly. If a person with HD is falling and lands on their buttocks, hip protector pads can significantly reduce the risk of fracture.

Wheelchairs and Seating Recommendations

Seating can be a challenge for persons who have chorea and who adopt poor postures. Persons with HD often like to sit with their legs over armrests or leaning off to one side. While these postures can lead to contractures and other issues, it is difficult to address them therapeutically if the person is determined to sit that way and unable to understand or remember instructions to adopt a better posture. Placing persons with HD into a posture and restricting them from changing position can lead to agitation and combativeness.

When a person is no longer able to walk independently, he/she can often maintain their mobility by learning to self-propel using their legs while seated in a hemi-height or drop seat wheelchair. Standard wheelchairs (either self-propelling or transit) can be modified with a small wedge cushion or lumbar roll to promote better sitting posture, and padding for armrests and footrests to prevent bruising. Reclining wheelchairs provide the option to change positions from upright, but may encourage a person to slide down in the chair. Specialized seating including side supports and tilt-in-space seating may be necessary to improve upright posture of the trunk and prevent sliding out of the chair. For persons who do not tolerate this type of seating it may be necessary to add padding or use specialized chairs such as the Broda Chair® or the CareFoam™ chair (Figure A). The CareFoam™ chair and others like it are particularly helpful with persons who tend to turn sideways in their chair and are best used in the later stages of the disease when self-propulsion is no longer possible.

Figure A. Specialized chairs for HD



Broda® chair



CareFoam™ chair

Task Specific Training

• Transfers

In later stages of the disease carry over and short term memory are poor. While the person with HD is unlikely to be able to verbally describe what they have learned about transfers or other motor skills, they can learn new habits and become safer. New movement patterns are learned through repetition of the functional activity. Ideally this repetition should occur every time the activity is performed and caregivers should cue the person to do the movement correctly and should not allow errors to occur. Through repetitive practice of a movement pattern implicit motor learning occurs despite the inability of the person to explicitly state what they have learned. It may take 2-3 weeks of constant reminders and practice to develop a new movement pattern and turn it into a habit. If it is not learned after this time, it may not be possible to change the movement pattern.

Sit to and from Stand Transfer

Persons with HD often lean backwards and/or to the side while rising from chairs and tend to fall backwards into chairs. Due to chorea and motor impersistence, using a walking device or railings may not provide consistent support. A method that has been successful with many persons with HD is to teach them to transfer using their hands on their thighs. Instruct the person to initiate rising from the chair with their hands on their knees. As they push through their hands for support they must lean forward and this puts their center of mass over their feet. They then keep their hands on their legs sliding them up their thighs as they come to standing. This provides for a smooth movement into extension and stabilizes the trunk and upper body during the transfer. Returning to sitting is accomplished by having the person place their hands on their thighs and slide their hands to their knees as they go to sitting. This provides for a smooth and controlled transition to sitting and prevents them from falling backwards into the chair while still in an extended posture.

✔ Please go to https://www.youtube.com/playlist?list=PLLQmMRDsNEY2_6crhI-z9yL9FCc0FLI-VD to view Clip #3 – Sit to Stand



An example of a treatment session would have the person with HD practice the transfer with hands on knees at least 3 times. At the same time have family and caregivers observe. Develop simple cues such as “hands on knees, now up 1, 2, 3” using the count to cue a smooth movement. The family would then be instructed to make sure that the person with HD is told to put their hands on their thighs every time they transfer for the next 2 to 3 weeks until the movement becomes established as a habit.

Bed Transfers

If upper extremity chorea is a problem turning in bed may be safer if persons are instructed to grab their elbows and then swing their arms to the side to initiate turning. This prevents flailing movements that can lead to bumping the arms on the bed or hitting themselves. If chorea of the upper extremities is impeding staff during a transfer have the person with HD “hug themselves” to minimize upper extremity chorea.

Bed Positioning and Safety

Persons in middle to late stages of HD may have problems with falling out of bed due to choreic movements, decreased spatial awareness that reduces their ability to sense the edge of the bed, and/or difficulties with force modulation causing them to “vault” out of bed when all they want to do is turn over or sit up. Possible solutions are to use padded bed rails or a lower bed height (3 to 5 inches from floor) if they are able to rise to standing. If these options are unsuccessful, a fully netted bed enclosure which fits over a standard hospital bed or a Craig bed composed of a foam mattress and four padded walls that form a

safe padded cubicle enclosure might be useful. While many people may initially feel confined or isolated in these enclosures, they often appreciate the ability to change positions without falling out of bed and may sleep better. For persons with sustained dystonic posturing that could lead to contractures, bed positioning devices such as padded rolls and supports may be useful.

Airway Clearance Techniques

Respiratory function is affected in persons with HD typically in the later stages of the disease. Breathing exercises such as diaphragmatic breathing and pursed lip breathing ("blow out a candle"), as well as postural exercises are recommended at all stages of the disease to increase the efficiency of breathing and coughing to prevent lung infections. At later stages, manually assisted coughing techniques or use of a mechanical Insufflator-Exsufflator machine may be helpful to clear lung secretions due to an ineffective ability to cough.

Managing Cognitive and Behavioral Issues

Cognitive problems in persons with HD occur early in the disease and include impaired attention, working memory, and executive functions such as planning, organizing, and sequencing. In later stages, the cognitive deficits progress to global dementia. These cognitive deficits can interfere with their abilities to learn new motor tasks. Persons with HD also report difficulty with "multi-tasking". To enhance the learning process, therapists can: encourage repetitive practice of tasks, allow ample time for cognitive processing of feedback, provide cueing (visual, verbal, or physical) to enable task completion, or break down complex tasks into simpler tasks and have persons attend to one task at a time. A quiet environment with few distractions can help a person with HD to concentrate.

CASE STUDY

Lisa, a person with HD, has had several falls when climbing stairs. She and her family are instructed that she is to stop before going up or down stairs and think about how she will climb the stairs. She is then to grab the hand rail and go up or down the stairs slowly. She is also told to be sure that she does not carry anything when climbing stairs and she is not to talk while on the stairs. Family are also instructed that they should not talk to her while she is on the stairs. They are also asked to cue her to "stop and think" before she goes up or down stairs.



Psychiatric symptoms exhibited by persons with HD in the middle to late stages are depression, apathy, perseveration (i.e. difficulty with shifting to new tasks), and impulsivity. Persons with HD who are depressed and/or apathetic may not be motivated to engage in or have difficulty initiating an exercise program or physical activity. Many times apathetic persons with HD will participate in exercise if someone just helps them get started and works along with them to keep them engaged. Strategies to promote inclusion of regular physical activity in the lives of persons with HD include educating them and/or their caregivers about the purpose and benefits of regular exercise, collaborating with persons with HD to determine their functional needs, identifying barriers to exercise (e.g. physical, environmental), and designing exercise programs that are acceptable and feasible to the person with HD. Some additional ideas for ways to motivate and facilitate engagement of persons with HD in exercise programs are the use of home-based exercise DVDs, group or gym-based exercise programs, or use of specific video games such as Dance Dance Revolution, as these interventions were all found to have high adherence rates in clinical trials with persons with HD. Setting a specific time during the day for persons with HD to exercise, and incorporating exercise into their daily routines, are also ways to improve adherence. For persons with perseverative problems (e.g. excessive hand washing), trying to redirect their attention to other tasks may be beneficial.

It is always best to give persons with HD specific time limits when prescribing exercise programs to avoid having them exercise in excess due to perseveration. Impulsivity and lack of awareness of deficits in persons with HD can result in unsafe behaviors. Talking with persons with HD about the "pros" and "cons" of their behaviors, and providing constructive ideas of other ways to behave may be helpful.

CASE STUDY – MIDDLE STAGE HD

Rob lives at home with his wife with additional caregiver assistance during the days when his wife is at work. Rob is 40 years old and was diagnosed with Huntington's disease 15 years ago. He can ambulate but has fallen repeatedly in the last year. He walks with his legs spread in a wide base of support and tends to clasp his hands behind him. When getting up from a chair he requires the use of his hands and has to rock back and forth several times. He tends to lean backwards during the transfer and has fallen backwards sometimes landing on the chair and sometimes missing the chair and falling to the floor. When sitting down in chairs Rob stops several feet away from the chair then falls backwards into the chair with a rigid trunk. He does not bend at the hips and chairs tend to slide backwards when he lands in them. He has broken two chairs when sitting down in them, both times the chair tipped over backwards.

Physical Therapy Evaluation



Appropriate measures for assessing Rob would be the Timed Up and Go Test and/or the Tinetti Mobility Test to assess gait and transfers as well the Berg Balance Scale to assess balance. In addition, the physical therapy evaluation should include an assessment of range of motion (including the trunk and hips) and of strength. Rob has significant motor impersistence and does not understand directions for a manual muscle test so the strength assessment is performed by having Rob perform functional movements against gravity such as a squat for knee and hip strength, reaching for an object on a shelf and lowering it to the counter for arm strength, and rolling in bed and transferring supine to sit for trunk strength. Eye movement and chorea are assessed using the UHDRS motor scale. If Rob has significant difficulty moving his eyes in the horizontal or vertical direction this will impact his ability to safely navigate and to accurately locate seating surfaces. Chorea cannot be treated by PT but the therapist needs to be aware of its severity, how it is changing over time and its potential impact on function. Chorea that is worsening and negatively impacting function should be brought to the neurologist's attention as there are medications that may help to dampen chorea.



Physical Therapy Treatment

Training Rob to transfer by first feeling the chair on the back of his leg and then by placing his hands on his thighs to encourage forward flexion and provide support throughout the transfer will improve safety during stand to sit transfers. Due to cognitive changes in the middle stage of HD, Rob will need to practice this new method of transferring many times before he will remember to do it. Visual and verbal cues are also

helpful in reminding him to do the transfer in a safe manner. To further improve safety during transfers, chairs in Rob's home should be heavy so that they don't tip over when he falls into them. There should be grab bars in the bathroom so that he has a safe, firm handhold for toilet transfers and when bathing.

Gait should be assessed with a rollator walker. These walkers encourage users to keep their arms in front of their bodies and provide constant support. The swivel wheels will allow Rob to walk and turn smoothly without having to lift or maneuver the walker. Most persons with HD are able to learn to safely use the rollator with little to no instruction. Due to cognitive issues, giving explicit verbal directions for walker use is of limited value. The brakes are not necessary for safety but family and caregivers, as well as Rob, should be trained to lock them before having Rob sit on the seat of the walker.

If Rob has been sedentary he would benefit from a program to improve his activity level while increasing strength and improving balance. Walking programs are highly recommended. Due to apathy related to HD, short daily walks are usually better tolerated. Encourage Rob to take on some chores/household activities that will require him to get up and walk multiple times a day. Some suggestions are that he do things like getting the mail, check that the porch light is turned on, carry his dishes and trash to the kitchen using the basket on the rollator walker. The chores and activities should be individualized based on Rob's living environment and his interests. Since many persons with HD spend a large amount of time watching television, it is recommended that persons with HD stand up and walk each time there is a break in the show for commercials. Rob could walk to the television, touch it and return to his chair or walk to the bathroom or any other appropriate target. Build in rewards that are individualized to help overcome the apathy that is present in HD.

CASE STUDY

Sarah is a 54 year old woman diagnosed with Huntington's disease 20 years ago who lives in a nursing home. She is dependent in all activities of daily living and is able to follow simple commands. Sarah used a rollator walker in the past for ambulation but is no longer able to safely use one due to her motor planning and balance deficits. She is able to rise to standing and can walk short distances (e.g. from her bed to bathroom) with minimal to moderate assistance from one person before fatiguing. The staff at the nursing home are concerned because she is falling out of her bed and geri-chairs due to her severe chorea and frequently falls in her room when attempting to walk unassisted to the bathroom.



Physical Therapy Evaluation

The physical therapy evaluation should include assessment of body structure/functions such as range of motion and chorea, vital signs including respiration, and frequency and circumstances of falls in the past week and 3 months. Evaluation of her static and dynamic balance in sitting unsupported at the edge of bed or in a chair, and standing postures and performance of transitional movements (supine to sitting at edge of bed and back, sitting to standing and standing to sitting) should be performed. It would not be appropriate to use most standardized measures of balance and mobility such the Berg Balance Scale, Timed Up and Go, and Tinetti Mobility Test because she requires manual assistance. The six-minute walk test might be too difficult for her to perform, but the therapist might consider modifying the test to two minutes from six and document the amount of manual assistance that she requires.

Physical Therapy Treatment

To provide adequate physical activity for Sarah to maintain her present functional status, the therapist can recommend that Sarah be put on a scheduled walking program so that she has opportunities to walk with assistance from staff and/or trained family members at least 3-4 times a day. An alternative or additional way for her to increase her physical activity would be for her to use a recumbent bicycle in the physical therapy department on a daily basis if she is able. To ensure her safety during ambulation, the therapist can recommend that a gait belt be worn.

✔ Please go to https://www.youtube.com/playlist?list=PLLQmMRDsNEY2_6crhl-z9yL9FCc0FLI-VD to view Clip #11 – Use of an Exercise Bike

If there are railings in the hallways, she might be able to hold on to the railing on one side so that she requires less manual assistance from the staff. More frequent walking with assistance might decrease her attempts to walk alone in her room and might decrease her chorea due to fatigue.

✔ Please go to https://www.youtube.com/playlist?list=PLLQmMRDsNEY2_6crhl-z9yL9FCc0FLI-VD to view Clip #8 – Walking in the Hallway

If she continues to fall out of chairs because of her choreic movements, the therapist might recommend a trial of a Broda® or Carefoam™ chair. Based on the findings, the therapist should discuss with staff appropriate measures for preventing Sarah from falling out of bed that adhere to the policies of the nursing home.

Thing to Remember

- Falls may occur frequently and often while the person is performing multiple tasks simultaneously.
- One goal of therapy is to help the person maintain motor skills and mobility for as long as possible.
- Any assessment or treatment will be influenced by cognitive and behavioral issues.
- It is important to assess range of motion and strength as the person becomes less mobile.
- Goals of treatment focus on maintaining function, improving safety and adapting the environment.
- Therapists should carefully weigh risk versus benefit in making recommendations about ambulating and when to transition to a wheelchair.
- A four-wheeled rollator walker is the safest ambulatory device for persons with HD.
- Protective equipment (helmets, knee and elbow pads) may be useful for those at risk for falling.
- Specialty chairs like the Broda® or CareFoam™ chair work well for persons with HD because of special padding, side supports and increased versatility.
- For persons with HD that fall out of bed, padded rails, a lowered bed, or a Craig bed may offer some relief.
- Cognitive problems can interfere with the ability of a person with HD to learn new motor tasks.
- Persons with HD that are depressed or apathetic may not be motivated to engage in an exercise program. Sometimes it helps to get someone started to keep them engaged.



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Chapter 5

Occupational Therapy for a Person with Huntington's Disease



Huntington's Disease Society of America

Occupational Therapy for a Person with Huntington's Disease

The Role of the Occupational Therapist in the Healthcare Team

To promote maximal independence and safety in daily life!

Occupational therapy (OT), as defined by the American Occupational Therapy Association (AOTA), is "the therapeutic use of everyday life activities (occupations) with individuals or groups for the purpose of enhancing or enabling participation in roles, habits, and routines" within any environment of a person's daily life (2014, p. S1). Everyday life activities include personal self-care tasks, home management tasks, and community activities. Accordingly, OTs provide intervention in a myriad of settings including outpatient clinics, nursing facilities, mental health programs/facilities, or through home health for persons who are homebound. Individuals who receive home health OT may live in traditional homes, independent living facilities, or assisted living facilities.

As Huntington's disease (HD) typically results in a varied array of motor dysfunction, cognitive dysfunction, and psychological dysfunction, OTs are well-trained to provide holistic intervention that maximizes safety and independence to promote quality of life across the stages of HD, both in adult and juvenile onset HD. Multi-disciplinary clinics at a Huntington's Disease Society of America Center of Excellence (HDSA COE) will include an OT as part of the outpatient health care team. For more information on OT, please visit www.aota.org

The Guideline for Occupational Therapy Intervention for Persons with Middle to Late Stage Huntington's Disease

With a focus on occupational therapy intervention in home health and/or long term care for the person with middle to late stage Huntington's disease symptoms

The key to successful occupational therapy (OT) intervention for persons with Huntington's disease (HD) is client-centered care. The occupational therapist must integrate foundational concepts from the "Occupational Therapy Practice Framework: Domain and Process" with an understanding of the progression of how HD impacts a person's ability to function in daily occupations (2014, p. S1-S48). Due to the way HD progresses, persons with HD who require OT services through home health or in a facility are most commonly in the middle to late stages of the disease process. These stages can include a broad spectrum of HD signs and symptoms, and have variable impact on functional abilities. Many persons with HD experience similar trends in signs and symptoms, but there are powerful functional differences among individuals. This is why client-centered care is necessary for maximizing safety and independence for persons with HD through the OT evaluation and intervention.

Persons with HD present with varying degrees of dysfunction in three areas: motor dysfunction, cognitive dysfunction, and psychological dysfunction. Because of this variability, the OT evaluation must thoroughly assess all three within the context of occupations in preparation for initiating intervention strategies. It is crucial to recognize that true functional ability or inability results from the intersection of all of these components within a person's environment. Due to the interaction among the areas of dysfunction, when a person with HD makes lifestyle adjustments to reduce his/her stress then he/she may see other functional abilities improve. This can be seen when a caregiver retires and is now available to provide consistent supervision and assistance, or after a person with HD moves from independent living to a more supportive setting, such as an assisted living facility.

Initial Evaluation

We will describe the typical evaluation of a person with early stage HD, with the thought that subsequent evaluations will follow the same general principles. The initial evaluation begins by understanding the typical daily activities of the person with HD. To understand the person's current level of function or dysfunction, it is essential to have an understanding of the performance patterns of a person with HD, including habits, routines, rituals, and roles. The OT will evaluate the person's ability to safely and independently perform their daily occupations within their typical environment.

> A person's daily occupations include activities of daily living (ADLs), instrumental activities of daily living (IADLs), rest and sleep, education, work, play, leisure and social participation. ADLs include bathing, dressing, grooming, and hygiene; whereas, IADLs include meal preparation, child care, and shopping.

Often, the OT's emphasis for evaluation and intervention in home health and in facilities is mostly ADLs and IADLs. However, other occupations must not be neglected, especially sleep and rest. During the initial interview, the OT will ask about the quality of sleep and sleep routine of a person with HD.

It is requisite to evaluate the person's environment during the evaluation process. Accessibility concerns include whether there is adequate space for safe performance of ADLs, or if there are safety hazards within the environment. Considering the accessibility of pathways and doorways in the home is part of this process. In addition, the OT must also assess the vision and hearing of a person with HD, and be aware of any required vision or hearing aids.

The initial interview and evaluation is an ideal opportunity to determine what activities and goals are most meaningful to the person with HD and the family/caregivers, as well as to establish therapeutic rapport. Consistent and effective communication among the OT, the person with HD, and family/caregivers is essential to the success of OT intervention. This is particularly pertinent in facility settings where a person with HD has multiple caregivers.

The assessment of *motor abilities* should include gross motor control, fine motor control, dynamic balance, static balance, modulation of speed and force, and ability to sustain force (including grasp). The assessment of strength, range of motion, and endurance should also be included. It is helpful to consider sensory abilities, including proprioception (an awareness of posture, movement, and changes in equilibrium and the knowledge of position, weight, and resistance of objects in relation to the body), as this impacts motor function. Also, the OT should inquire if the person with HD has chronic pain. While the most frequently known form of motor dysfunction for persons with HD is chorea, not everyone with HD exhibits this movement disorder. Other or additional motoric manifestations of HD can include dysmetria (an inability to control range of movement), dystonia, rigidity, akinesia, and bradykinesia. An excellent resource for further details on these motor control impairments is *Motor Control: Translating Research into Clinical Practice (4th ed)* by Shumway-Cook and Woollacott. It is important to recognize that fluctuations in motor performance can occur as a result of stress, fatigue, medication effectiveness over the course of the day, and whether the functional activity is also cognitively challenging. Consequently, the OT should consider evaluating motor abilities over multiple sessions and situations. The team of healthcare providers should discuss their observations to note trends in the functional abilities of the person with HD. A great way to assess functional motor abilities is within the context of the person's ability to perform



toileting. The task of toileting includes functional ambulation, dynamic standing, bilateral upper extremity gross motor control (personal hygiene, hand washing, and garment management), and fine motor control (provided the pants have fasteners).

The assessment of *cognitive abilities* should evaluate orientation, attention, memory (especially immediate memory and short term memory), processing speed, initiation, sequencing, problem solving, insight, executive functioning, impulse control, and safety awareness. The OT should also consider both the person with HD's perception of his/her abilities and the family/caregiver's perspective on this; these responses can be starkly different. The activities used for functional cognitive assessment depend on a person's typical daily activities. Consider requesting that the person with HD follow a sequence of verbal and/or written directions to complete an appropriate multi-step task. Determination of an appropriate sequence of tasks should be based on the person's report of his/her normal daily activities. For example, in the home health setting, the OT may ask a person with HD to sort his pills for the week, perform simple hot meal preparation (e.g. assemble a lunch or snack using the microwave), or retrieve a cup of water. In a facility setting, part of the cognitive assessment could include completing a simple three step sequence of ADLs in the order requested. For instance, "Please put on your slippers, try to use the toilet, and then wash your hands." Another part of the assessment could include asking the person how he would respond in a hypothetical emergency situation. For instance, if the person with HD often uses the microwave for simple hot meal preparation, what would he do if he overheated the food by misdialing the time on the microwave, and then mildly burned himself? How would he respond to this unexpected situation and what might he do to prevent its recurrence? This example could be applicable in the home health or independent living settings. For a person in mid stage or late stage HD living in a facility, it is both practical and valuable to ask the person with HD to demonstrate how he would call for assistance in the facility.

The *psychosocial* assessment should emphasize observation of behavior when the person with HD is interacting with family/caregivers as well as in other groups. The latter can be very important when a person with HD is in a facility, especially if most meals occur in a group setting in a dining hall. Other valuable considerations include a person's level of emotional lability, level of motivation or apathy, as well as the person's values, beliefs, and spirituality. Also, consider any indications that the person with HD is dealing with depressive symptoms or stages of grief; these considerations are more common for persons with better levels of insight. The psychological symptoms in HD include all of those found in other medical settings. Mood disorders are particularly common, but persons with HD may have a variety of changes in mood or behavior. These are amenable to treatment with both medication and behavioral interventions. It is appropriate for the OT to advocate for the person with HD to have appropriate attention paid to their mental health needs, such as seeing a neuropsychologist or counselor.

Due to the genetic nature of the disease, most persons with HD have seen a family member or multiple family members progress through the stages of the disease. As a result, the person with HD may be sensitive about certain topics and/or therapy recommendations, which may result in an unwillingness to try compensatory strategies or medical equipment. For example, a person with HD may feel that using a shower chair leads to needing a wheelchair and, later, having to go to a nursing home. As a result, he may firmly avoid using a shower chair. A person with HD may express fear about having the same disease progression as a loved one. Consequently, the person with HD may need extra patience when beginning to use new equipment.

Finally, it is important to thoroughly interview the family and/or direct caregivers of the person with HD as part of the initial evaluation process. Either the impact of cognitive dysfunction on insight and/or the impact of denial can skew the self-report of a person with HD. A thorough interview includes communicating with any nurses, certified nursing assistants, and private care aides in both home and facility settings. OT

intervention is often most successful with support from the family/caregivers and effective communication with the entire healthcare team.

Occupational Therapy Interventions

Due to progression of the disease and potential temporary changes in functionality from a myriad of causes, such as infections, fatigue, or changes in medications, the OT intervention process must be flexible. The OT must meet the person with HD where he is *today*, and help him and his family/caregivers prepare for maximizing independence and safety on both good and bad days. It is particularly valuable to provide intervention suggestions that help prepare the person with HD and his family/caregivers for *bad days* due the progressive nature of the disease. In essence, today's *bad days* often become tomorrow's *every day*.

CASE STUDY

Mrs. Anderson, a 43 year old woman with mid stage HD, diminished balance, mild chorea, and decreased attention and memory, lives with her husband. The couple share that she has had some near falls in the shower in the last six months.

Mrs. Anderson's home health OT has recommended that she have grab bars installed in her shower. The OT emphasized to the Andersons that Mrs. Anderson must consistently use the grab bars for her shower transfers and while showering. Mrs. Anderson is able to comply with these recommendations with reminders from her husband, and this is effective at her current stage.

However, when Mrs. Anderson contracted a urinary tract infection, her functional abilities diminished, including her balance which resulted in a greater fall risk.

Mrs. Anderson's OT had anticipated that she would need a shower chair as her symptoms progressed and this became useful equipment to keep her safe from falls during her recovery from the infection.

Therapeutic OT approaches include remediation/restoration, maintenance of functional abilities, and more commonly in HD, compensatory/adaptive strategies. These types of interventions will be addressed in detail below including the different approaches employed. The following intervention information is organized according to the "Occupational Therapy Practice Framework: Domain and Process" (American Occupational Therapy Association, 2014).

Occupations and Occupation-Related Activities

Activities of Daily Living (ADLs)

➢Bathing/Showering

Bathing/Showering for Individuals with Cognitive Dysfunction

A person with HD with cognitive dysfunction may have significant difficulty remembering to take a shower, initiating taking a shower, and sequencing the process of washing (e.g. remembering if all areas of the body have been washed and rinsed). It is not uncommon for caregivers to report that when reminded to get in the shower, the person with HD gets in the shower, stands under the shower spray, turns off the water, and then gets out of the shower without actually washing.

In order to promote consistency with bathing/showering, it can be helpful to establish a **daily routine** that includes bathing/showering happening immediately after another consistent task. For example, it may work well for the person with HD to always bathe/shower immediately following dinner every day. This will help establish a routine that allows the person with HD to be able to expect the bath/shower, and possibly develop the habit of finishing dinner and then walking to the bathroom for the bath/shower.

CASE STUDY

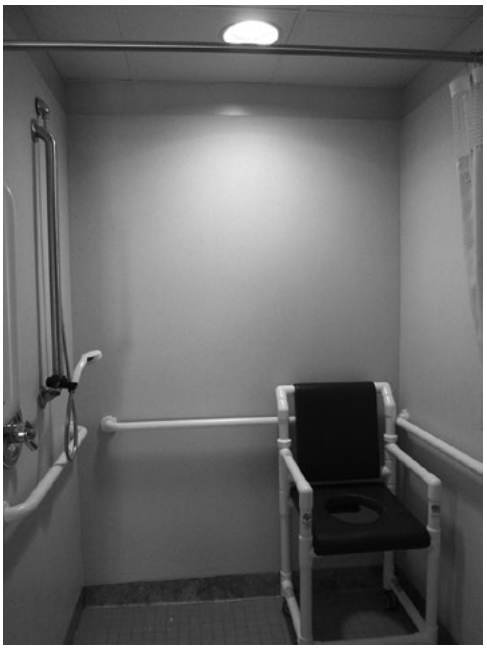
As noted earlier, Mrs. Anderson demonstrates diminished attention and memory in addition to her motor deficits. If her husband does not remind her, she occasionally neglects to shower despite intending to do so daily. This occurs because she is easily distracted within her home environment. Mrs. Anderson's home health OT recommended that she and her husband chose a convenient time daily for her to take her shower. The Andersons decided that since Mr. Anderson is always home in the evenings that Mrs. Anderson will always shower immediately after they have dinner. Consistent timing and sequencing of daily tasks help Mrs. Anderson develop a consistent routine. The establishment of a routine helps her to compensate for her diminished attention and memory. Using a similar compensatory concept, the OT also recommended that Mrs. Anderson bathe her body in the same sequence every time. This strategy help her remember to wash each area just once by starting with washing her hair first and then moving down in her body in the same order every time. Mrs. Anderson greatly benefits from using well-established habits and routines to compensate for her diminished attention and memory.

For the person with HD who has difficulty with sequencing bathing/showering tasks, it is valuable to establish the **habit of always scrubbing in the same order**. Even if a person cannot physically perform his/her bathing, using consistent daily routine and bathing/shower habits can establish expectations that foster cooperation; the regular routine will help minimize a person's refusals due to anxiety or stress.

A person HD with diminished attention, problem solving, and error recognition may be at risk for sustaining burns in the tub or shower if the water heater temperature is set too high. The OT can provide instruction to caregivers or maintenance staff to **adjust the water heater temperature setting** to avoid this risk. In facilities, staff may need to check the water temperature when a resident begins bathing if the water heater temperature cannot be adjusted centrally.

Bathing/Showering for Individuals with Motor Dysfunction

Impaired balance and/or coordination of movements can disrupt a person's ability to wash, rinse, or transfer in and out of the tub or shower.



Strategies to address these problems include no-slip adhesive applied to the shower floor, grab bars inside and/or outside of the tub or shower, a shower or tub seat, and the physical assistance of another person to help with bathing and/or transfers. To avoid injury to the person with involuntary or ballistic limb or trunk movements, it is important to use high-quality materials and install any grab bars or other devices securely into studs, walls, or other surfaces. A sturdy hard plastic shower seat may be less likely to bend or loosen than a shower chair with aluminum legs. An adjustable height or handheld showerhead and a wash mitt can help the bathing process for many persons.

Some persons with HD have severe motor control deficits, such as significant chorea, and may benefit from wearing a helmet for head protection. Given the inherent dangers of slipping or bumping into the wall of a small shower space, it can be appropriate to recommend a **vinyl helmet**, which can get wet, for wear that includes the shower.

Due to diminished balance, most people with mid/late stage HD require some compensatory plan or equipment in order to safely bathe/shower.

While bathing a person with late stage HD, the caregiver must use proper body mechanics for their own safety. OT education and training for caregivers should include guidance on **caregiver body mechanics for caregiver safety**.

CASE STUDY

Mr. Smith, a 33 year old man, lives with his mother and stepfather. He demonstrates late stage HD including significantly impaired motor function. Mr. Smith's primary motor deficits are bradykinesia and akinesia, which present as slowed movement and delayed initiation of movement. He also demonstrates impaired attention, memory, and executive function skills. His affect is generally flat.

Mr. Smith's caregivers are his parents, who are in their late 50's, and private caregivers. Given his notable progression in symptoms, his physician prescribed home health OT. This OT observed that Mr. Smith always relies on his caregivers to perform greater than 50% of the physical effort for him to perform functional transfers, and, often, he requires greater than 75% physical assistance. Mr. Smith's caregivers must transfer him frequently throughout each day to assist him with his personal needs. Due to the repetitive heavy lifting required from Mr. Smith's caregivers, the OT instructed the caregivers in using good body mechanics during caregiving tasks, which include the following key principles:

- Avoid repeatedly bending over while providing care when unnecessary. For example, elevate the hospital bed height when assisting a person with HD with dressing while he/she is lying in bed.
- Always use a wide base of support while providing physical assistance. Have feet staggered and at least shoulder-width apart.
- Avoid bending and twisting the back, but rather, bend with the hips and knees. The *power* for lifting must come from the strength of the legs rather than the strength of the back.
- The caregiver should keep the person who he/she is assisting close to his/her body while providing transfer assistance.
- If two caregivers are available to provide physical assistance, then work as a team to share the physical demand.
- The caregivers must communicate with the person with HD and one another while providing physical assistance.

As a result of the OT's caregiver training, Mr. Smith's caregivers use good body mechanics while assisting him from his bed to his rolling shower chair. The caregivers use a consistent method of elevating the hospital bed, using a gait belt, working as a team, and individually ensuring use of a good base of support and avoid bending or twisting their backs. Implementation of these principles helps Mr. Smith's caregivers provide physical assistance in a safer manner for all involved.

When the person with HD is bed-bound, it may be appropriate to use a supportive roll-in shower chair in a roll-in shower space. Some rolling shower chairs also provide tilt and/or reclining positioning. For example, BRODA® shower commode chair on wheels offers tilt and recline positions, as well as a padded toilet seat. Eventually, sponge baths may become necessary.

➢Toileting/Toilet Hygiene

Toileting/Toilet Hygiene for Persons with Cognitive Dysfunction

For persons with HD with diminished attention, one of the greatest difficulties regarding toileting can be awareness of urges and planning for toileting in a timely manner. Breakdown in these functional abilities can result in a relatively physically capable person with HD having incontinence issues or having falls/near-falls while rushing to the bathroom. These persons benefit greatly from the **establishment of regularly-scheduled bathroom breaks, or a toileting schedule, in their daily routine**.

For persons with HD, toileting hygiene can become a challenge. Remembering to flush the toilet can become difficult due to diminished attention and sequencing. As indicated above, **positive habit-formation training** can be effective. Otherwise, the person may need verbal cues from a caregiver in order to be successful.

Most persons with HD eventually require the use of incontinence **briefs** due to cognitive and motor difficulty with toileting. Whenever there is incontinence, both the person and caregiver need to employ strategies to maintain good perineal skin integrity. Successful maintenance of perineal skin integrity for a person in a facility requires collaboration with the nursing staff. Incontinence issues can be embarrassing for persons with HD, and can be the catalyst of significant safety concerns. For example, due to embarrassment from an episode of incontinence, a person with HD may try to independently doff clothing and take a shower. In order to hide incontinence issues, it is common for persons with HD to have falls or near falls as a result of trying to manage their own needs privately even when they are unable to do this safely.

Toileting/Toilet Hygiene for Individuals with Motor Dysfunction

Impaired balance and force modulation during toileting transfers can result in falls/near falls, damage to the toilet/plumbing, and/or injury to the person with HD. A first-line compensatory strategy for safety during toileting is habit-formation training to teach the person with HD to **consistently reach back** with one hand to the toilet seat for steadiness during the transfer. If the person with HD demonstrates further difficulty, then consider recommending **toilet grab bars** and/or **increasing the height of the toilet seat**. In particular, an increased toilet height makes the transition between standing to sitting on the toilet easier. Thus, it results in less *flopping* onto the toilet or *catapulting* off of the toilet, which often occurs with impaired force modulation.

CASE STUDY

Mr. Jones, a 42 year old man, lives at home with his wife and teenage children. He demonstrates Mid-Stage symptoms of HD, and his most limiting issues are significant chorea and impaired force modulation. He also demonstrates mild cognitive and psychological deficits. He and his family note that his impaired force modulation results in excessive force when placing items down on surfaces and when he transitions between sitting and standing. The Jones family says that there is often a loud banging noise when Mr. Jones sits down, especially when he sits down on the standard toilet. They also indicate that the toilet seat bolts are perpetually loose due to Mr. Jones' poor motor control.

Mr. Jones' home health OT recommended that the family try placing a bedside commode with a splash guard positioned at a comfort height over the standard toilet, as this will provide increased toilet seat height and convenient grab bars/armrests. The OT also strongly encouraged Mr. Jones to always reach for the bedside commode armrest to steady himself before beginning to lower himself on to the toilet seat. This environmental modification provides a compensation for Mr. Jones' poor force modulation and insures his safety while maintaining the condition of the family bathroom.



Wall – mounted toilet plumbing can crack or break off due to repeated damage from the person who flops onto the seat. A molded plastic 3-in-1 bedside commode installed over the toilet is cheaper to replace that than the entire toilet; some care facilities specializing in HD have installed floor-mounted stainless steel fixtures.

Men with HD who have diminished balance and/or diminished upper extremity motor control often have difficulty appropriately aiming to urinate in the toilet. Urine spills can be a safety hazard with regard to falls. To reduce fall risk and to improve cleanliness, **encourage men to sit while urinating**.

Persons with diminished upper extremity motor control may have difficulty physically performing hygiene. A person with HD may be more successful using **toilet wipes** rather than toilet paper. The texture of the toilet wipes provides increased sensory input, which may improve personal hygiene.

Encourage caregivers to have portable **urinals** available at night and for travelling to reduce issues with fall risk and urgency. During any travel, even to and from doctors' appointments, a urinal can be an easy alternative to managing in public restrooms.

Given the typical age of onset of HD, it is not uncommon for women with HD to still be premenopausal. The management of menstrual products and personal hygiene can be difficult for women with HD who have cognitive and/or motoric difficulties. If a woman with HD is having difficulty physically managing a tampon or cognitively remembering to change this product appropriately, it may be necessary to transition to using only pads. If a woman with HD has substantial difficulty managing her personal hygiene, the OT may consider advocating for a discussion with the woman with HD, the caregiver(s), and the physician about pharmacological options to reduce this burden.

➢Dressing

Dressing for Persons with Cognitive Dysfunction

Persons with cognitive deficits that include diminished attention and problem solving may have difficulty appropriately orienting garments when dressing and undressing. They also may be overwhelmed by the choices of what to wear. These individuals benefit from simplifying their everyday clothing options, such as by **hanging matching shirt and pants together** in the front of the closet. Other persons may need **extra time and/or verbal cues to complete their daily dressing**. As previously noted, the establishment of structured ADL routines and habits can assist a person with HD in appropriately selecting and donning garments.

CASE STUDY

Mrs. Williams, a 55 year old woman, lives with her husband. Her mid stage HD symptoms include her impaired attention, short term memory, and problem solving, as well as her increased anxiety and stress. She experiences minimal motor deficits. Due to her decline in function, she is seeing an OT. It is important to her to maintain a neat and tidy appearance, especially when she is out in the community, like her weekly attendance at church. On several occasions recently, she and her husband have intended to go to church but she has been too overwhelmed and anxious by her struggle to assemble an appropriate outfit. She reports that she will choose the skirt but then becomes overwhelmed and distracted attempting to find the rest of her outfit. She also indicates that she cannot consistently remember what she is planning to wear when she goes to her dresser to locate appropriate undergarments. Mrs. Williams shares that she was incredibly saddened that her planning and organizing difficulties prevent her from going to church, and Mr. Williams explains that he does not know how to help his wife.

The OT recommended that the couple hang two to three pre-arranged outfits, including appropriate undergarments and shoes in a grocery bag hung over the hanger, in an easily visible space in Mrs. Williams' dressing space. The OT strongly encouraged Mr. Williams to allow for ample time and support for Mrs. Williams to select the main pieces of her outfit options on Saturday at the latest. The unworn outfit option would be saved for the following week. This system of structured planning and simplification would allow Mrs. Williams to preassemble her outfits with maximal independence, allow her to be appropriately dressed in time, and decrease her stress.

Dressing for Persons with Motoric Dysfunction

By mid stage to late stage, persons with HD who have motor deficits have generally already **transitioned away from wearing clothing with fasteners** (buttons, zippers, etc.) or they may use Velcro fasteners. However, if garments have a zipper, it can be easier to manage when a **key ring or paperclip** has been threaded through the zipper pull. In these later stages, persons with HD typically wear loose-fitting t-shirts, shorts or pants with elastic waistbands, and shoes that can be slipped on/off. Supportive sneakers can become slip-on/off shoes using **elastic shoelaces**. Persons should **consistently sit down** for as many of the lower body dressing tasks as possible. Always encourage the person with HD to participate as much as possible in dressing activities, even if only to lift an arm. Dressing and bathing are opportune times for caregivers to integrate active and passive range of motion into the activity.

✔ Please go to https://www.youtube.com/playlist?list=PLLQmMRDsNEY2_6crhI-z9yL9FCcOFLI-VD to view Clip #5 – Footies

CASE STUDY

Mr. Johnson, a 70 year old man, with late stage HD lives in a long-term facility. He relies fully on the facility staff to manage his personal care although he can participate minimally. The facility OT was consulted to assist the caregivers in maximizing Mr. Johnson's safety, comfort, and quality of life. The OT encouraged the caregivers to use the tasks of personal care to encourage Mr. Johnson to perform active range of motion (AROM) and/or passive range of motion (PROM). The OT provided the example of assisting Mr. Johnson with bathing *and* AROM by coaxing him to lift each arm as much as he is able while the caregiver bathed and rinsed under each arm. Another example would be to hold up the pants leg for Mr. Johnson at attempt to lift his foot toward the pants. The caregivers indicated that these strategies encouraged Mr. Johnson to move or more easily tolerate being moved because it was within a functional context.

When it becomes difficult for a person with HD to stand, it is safer to put underwear and pants on while lying supine in the bed or in a chair that fully reclines.

➢Feeding

Feeding for Persons with Cognitive Dysfunction

Persons with HD who have diminished functional cognition may require **reminders** about when it is time to eat. Sometimes, these persons will recognize hunger and eat a snack, but they may not be able to initiate getting a meal when it is time to eat. Maintaining good caloric intake can be critical for persons with chorea.

Due to diminished attention, they may also require **reduced distractions** during meal times. A person with HD may better focus on controlling his/her utensils and persisting with the task of eating in a less distracting environment. Avoid having the TV or radio turned on during meals. Likewise, in a facility dining room, consider having the person with HD who has cognitive deficits eat with minimal distractions. For example, consider having the person with HD face away from other people in a facility dining room to diminish distractions during mealtime.

Feeding for Persons with Motor Dysfunction

Motor dysfunction results in greater self-feeding issues for persons with mid stage to late stage HD including spillage of food. A person with HD with impaired motor control in his/her upper extremity may have difficulty sustaining a persistent grasp on the utensil, difficulty effectively scooping or spearing food, and/or difficulty with smoothly bringing the utensil to the mouth.

Persons with HD should generally **eat their meals at a table** rather than in a recliner or on the couch. Typically, the most effective position for persons with HD for self-feeding is sitting in a sturdy chair scooted up against a sturdy table. The mid-abdomen of the person with HD should be in contact with the edge of the table. If the person with HD is perpetually sliding his/her chair back and away from the table while eating, put the back of the **chair against a wall** and then slide the table up to him/her. Additionally, encourage the person with HD to **prop his/her elbows or forearms on the table** to increase stability and minimize extraneous arm movements.

CASE STUDY

The staff at a long-term care facility complained to the OT that Mr. Scott, a 53 year old man with late stage HD, always had substantial food spills during meals. The OT observed that Mr. Scott experienced excessive chorea especially during mealtime. The OT ensured that Mr. Scott was in a very supportive chair, positioned the back of his chair against a wall, and then slid the table up to him. The OT also recommended that the caregivers place Mr. Scott's utensil in a universal cuff on his hand because many of the spills occurred when he was repeatedly dropping his utensil. The OT additionally provided Mr. Scott with a weighted plastic cup with a lid to reduce drink spills. The person with HD and the caregivers were provided with information on proper positioning and posture during mealtime.

Many persons with HD who have difficulty with self-feeding benefit from using adaptive utensils, dishes, and cups. There are many different styles and brands of adaptive utensils, and what works for a particular person may change as the disease progresses. Thus the OT should be prepared to re-evaluate whenever the person, family, or care staff have concerns or notice a change. Examples of the kinds of adaptations that can be helpful include: unbreakable plastic dishware, cups with lids, handles, and a narrow spout; weighted cups; large handled silverware (or silverware with foam rubber tubing wrapped around the

handle); weighted utensils; and a universal cuff. Other adaptations include plates or bowls with a lip, a plate guard and using Dycem® or other non-slip shelf liner on the table to keep bowls and dishes from shifting. The OT should work closely with the speech-language pathologist on recommendations about food texture and the use of straws, thickened liquids, or other texture changes.

➢Functional Mobility (including wheelchair positioning)

Functional mobility can be extensively impacted by both motor and cognitive dysfunction for persons with HD. Many persons with HD fall due to being distracted during functional ambulation. A person demonstrating their walking to a healthcare provider often does not walk the same way as they do while engaging in functional tasks, especially with regard to safety. One does not exhibit the same version of ambulation when walking down a straight hallway while being watched as one does when urgently rushing to the bathroom. It is crucial that the OT observe the person with HD performing functional ambulation within the context of typical ADLs and IADLs.

CASE STUDY

Pam, a 46 year old woman with late stage HD, lives in a care facility. Pam continues to self-ambulate to the bathroom but was experiencing an increase in near falls. The OT recommended that Pam's room be rearranged to reduce the distance between her bed and bathroom and to maximize open pathways for functional ambulation within her living area. The OT emphasized to Pam the importance of keeping clutter out of her pathways for safety during functional ambulation, which included establishing new storage areas for her slippers and a stool. The OT also reinforced the importance of keeping these pathways well-lighted, especially at night; Pam and the OT decided to put a nightlight in the bathroom and a touch lamp on her bedside table.

The OT should speak with the physical therapist in order to determine which assistive device(s) may be most appropriate. For example, it may be that the person with HD needs to use a rollator or 4 wheeled walker to participate in meal preparation when there are significant distractions or a need to carry ingredients. However, he/she may not need to use a rollator in order to ambulate to the dining room of the facility. In essence, the person with HD may require *different tools* for *different jobs* of living. These recommendations for safe functional ambulation must be consistently supported and encouraged by healthcare providers and caregivers to foster habit-training and compliance.

✔ Please go to https://www.youtube.com/playlist?list=PLLQmMRDsNEY2_6crhI-z9yL9FCc0FLI-VD to view Clip #7 – Use of a Four Wheeled Walker

CASE STUDY

James, a 57 year old man with late stage HD, has been using a standard manual wheelchair in the facility where he resides but due to multiple falls he required a new device for safety and to provide functional mobility. A Broda® wheelchair was recommended to help James remain upright and allow him continued independence with functional mobility rather than risk propulsion from a standard chair.



Broda® chair

In general, persons with HD often benefit from using a **gait belt or rollator when ambulating, or a hemi-height wheelchair** when ambulation is no longer safe or practical. Persons with HD are typically less likely to benefit from using a cane and a front wheeled walker, as these items are less accommodating of cognitive deficits and chorea. A cane or front wheeled walker may increase the risk for falls/injury as motor and cognitive dysfunction progress.

✔ Please go to https://www.youtube.com/playlist?list=PLLQmMRDsNEY2_6crhI-z9yL9FCc0FLI-VD to view Clip #8 – Walking in a Hallway

Reducing the risk of injury becomes increasingly more important as the disease progresses. For some persons, safety equipment recommendations may include hip bolsters and helmets to reduce the risk of injury when falls occur. Soft and hard shell helmets are available for head and facial protection. These products may not meet with early compliance, but continued encouragement and education may result in better usage, especially when injuries begin to occur with more frequency.

Should the therapist determine that a wheelchair is necessary due to an increase in falls, decreasing the person with HD's reluctance to using a wheelchair may require time and education. Sitting the wheelchair in the room or area the person most occupies may be a way to introduce the device. The OT and physical therapist will work together to determine the type of wheelchair needed and assess for any wheelchair modifications that may be necessary. Anti-tip bars and hill-holders greatly improve the safety of wheelchairs during transfers. Hill-holders prevent the wheelchair from rolling backward even if the user forgets to lock the brakes. It is often noticed that the magnitude of chorea diminishes when a person with HD uses a chair that provides adequate support, good surface contact and good positioning.

Manufacturers have produced a multitude of seating and positioning devices touted to provide safe and effective positioning for persons with motor dysfunction. Due to the complex progressive nature of HD symptoms, it is imperative that the therapist who assesses seating and positioning for the person with HD understands the complexity of both cognitive and motor dysfunction in the person. Often, it is safer to permit the person with HD to ambulate unsteadily with supervision or assistance than to attempt to significantly restrict movement in a confining wheelchair.

CASE STUDY

Joan, a 67 year old woman with late stage HD, has had multiple falls in her long term care facility. Several staff members met as a team to review where and when the falls were occurring. It was determined that Joan was falling during transfers from her wheelchair to her bed. The OT recommended adding hill holders to the manual chair to prevent it from moving as Joan, due to her cognitive decline, was not able to remember to lock the brakes. This addition of equipment reduced the frequency of her falls.

Should restricted movement become necessary, the therapist may consider some of the following options:

Cushions:

Persons with HD may have problems with pelvic thrusting and posterior pelvic sitting positions. Pommel cushions and anti thrust cushions may be appropriate positioning devices. Anti-thrust cushions put the pelvis in a neutral position in the back of the wheelchair resulting in a more upright posture.

Seat Angles:

Another suggestion can be to statically tilt the wheelchair to the back by changing the axles on the manual wheelchair. This allows gravity to assist in keeping the pelvis in the back of the wheelchair creating more upright functional posture.

Lap trays, lap buddies, or positioning belts:

Documentation should clearly reflect the need for these positioning devices and the failure of all alternatives previously tried. These devices are sometimes used in conjunction with a pommel or anti-thrust cushion to provide upright functional posture in a wheelchair. Positioning or safety belts may, at times, be used to help keep a person with pelvic thrust in a more upright posture. Progression of positioning belts, from front-latching to back latching depends on the needs of the person with HD. For those with poor head/neck/trunk control, shoulder harnesses may also provide assistance but must be thoroughly assessed for safety and functional improvement. When seat belts that cannot be removed by a client are not permitted, consider a Velcro seat belt with connected chair alarm.

✔ **Please go to https://www.youtube.com/playlist?list=PLLQmMRDsNEY2_6crhI-z9yL9FCc0FLI-VD to view Clip #6 – Fastening the Seat Belt**

The person with HD must have freedom of movement or they will be considered a restraint. Although restraints are at times medically necessary, their need must be assessed, monitored, and documented appropriately.

✔ **Please go to https://www.youtube.com/playlist?list=PLLQmMRDsNEY2_6crhI-z9yL9FCc0FLI-VD to view Clip #1 – Adjusting the Seat Belt**

Specialized Wheelchairs:

Wheelchairs and wheelchair modifications may progress with the person's needs. High back or reclining wheelchairs may provide the back support needed when extensor tone or chorea occurs. If the person begins to exhibit a lateral lean, lateral supports can be added to most wheelchairs to provide upright positioning. As needs increase, personal wheelchairs can be ordered from wheelchair vendors to help therapists meet the needs of the person with HD.

Tilt-n-space wheelchairs may provide pressure relief to persons and also use gravity to keep the pelvis positioned to the back of the wheelchair. Broda® specializes in high back wheelchairs with bilateral supports and has developed an HD package for one of their chairs. Bigger wheels and wider bases of support provide stability while allowing the person to propel the wheelchair if they are still able to do so. Carefoam Chairs® use a concave style to their wheelchairs, chairs, and beds to reduce falls and assist in positioning, and their padding and sheepskin covers provide comfort and support when the person is upright in the wheelchair.

✔ **Please go to https://www.youtube.com/playlist?list=PLLQmMRDsNEY2_6crhI-z9yL9FCc0FLI-VD to view Clip #7 – Use of Four Wheeled Walker**

*Please see the chapter on physical therapy for further advice on ambulation and appropriate assistive devices.

➢Personal Device Care

Persons with HD with diminished motor control can have notable difficulty managing their glasses and hearing aids and often require assistance. Applying a **strap to glasses** minimizes the chance of the

glasses falling off. Encourage the person with HD and the caregivers to try to select glasses with durable frames.

➢Personal Hygiene and Grooming

Personal Hygiene and Grooming for Persons with Cognitive Dysfunction

It is common for persons with HD who have diminished cognitive function to have difficulty remembering to perform their daily grooming tasks, and some also have difficulty with sequencing these tasks. **Establishing a consistent daily routine** and keeping a **posted written list** can provide a visual reminder of the tasks to be completed. However, some persons with Mid Stage HD require **verbal reminders** from their caregivers, even though they may be physically capable of performing their grooming tasks. Occasionally, some persons with HD can be obsessive and compulsive about performing select grooming tasks. This should be addressed from a psychological behavioral perspective if it is causing harm (e.g. brushing teeth so vigorously that the gums bleed).

CASE STUDY

Mrs. Morris, a 60 year old woman, with late stage HD is compulsive about brushing her hair. She repeatedly goes in to the bathroom throughout the day to brush her hair. However, she is having frequent falls in the bathroom because her rollator is difficult to maneuver in the small bathroom space. She has diminished attention to her balance and safety while she is focused on brushing her hair. Brushing her hair is not directly causing her any harm. So, the OT works with Mrs. Morris to create an open space outside the bathroom with an easy-to-see mirror, sturdy chair, and easy storage for all of her hair brushes and combs. Initially, Mrs. Morris required reminders and encouragement about using her new hair-brushing space to alter her habit but ultimately she adjusted and experienced fewer falls due to these changes.

CASE STUDY

Sam, a 44 year old man, resides in a nursing facility. What began as a gentle reminder of washing his hands has become an obsession. He stands at whatever sink he can find and washes and rewashes his hands throughout the day. His vigorous washing causes water to pool on counters and floors increasing his risk for falls. Staff worked at providing time limits on Sam's washing focusing on before and after mealtimes at one particular sink. They also added a non- slip mat in front of the sink where he stands to wash his hands to decrease the risk of falls.

Personal Hygiene and Grooming for Persons with Motor Dysfunction

Persons with mid stage and late stage HD often demonstrate difficulties with effective fine motor control, making grooming tasks challenging. Initially, it is helpful for the person with HD to **lean against a nearby wall** and/or **prop their elbow** on the countertop while performing grooming tasks at the sink. If this positioning does not provide adequate stability, then it is beneficial to perform the grooming tasks while **seated**.

Using an **electric toothbrush** can aid in the effectiveness of oral care when motor control is limited. If a person finds it difficult to apply toothpaste to the toothbrush, they can squirt the toothpaste directly into the mouth. If a person with HD has difficulty spitting, they can **dip the toothbrush in mouthwash**, rather than using toothpaste, to reduce the need to spit.

➢Sexual Activity

Persons with HD with cognitive deficits and psychological impairments sometimes develop issues with promiscuity. This concern is often associated with diminished safety awareness and diminished impulse control. It can be helpful to guide the person with HD and their caregivers in **basic behavior management strategies**. It is also beneficial to encourage the person with HD, the caregiver(s), and physician to have a discussion regarding any birth control preferences.

Sexual activity may also be impacted by motor dysfunction and/or medications. The OT needs to be comfortable talking with the person with HD, the significant other, and the physician about safe positioning and the impact of medications on sexual activity.

Sleep



A person who has disrupted sleep will have difficulty performing other occupations safely and successfully. Persons with HD often have difficulty adhering to a consistent routine of sleep and wakefulness. Also, HD medications may impact sleep patterns. As noted above, it is best for a person with HD to have a consistent routine to aid in compensating for diminished cognitive function. Many persons with HD have a tendency to stay up very late at night and sleep late in the morning. However, a person with HD who is a "night owl" is likely to have less readily available supervision and assistance. Accordingly, it is extremely beneficial for a person with HD to be on a **consistent sleep/wake** routine in order to allow caregivers to be most available for helping with other occupations. Encourage the person with HD to wake at nearly the same time daily and to avoid shifting the sleep/wake schedule day-to-day.

Some persons with HD exhibit increased motor activity during sleep and some have issues with rolling out of bed. Various *soft* methods can be tried to reduce risk of falling out of bed. Tucking a rolled blanket or a swimming pool noodle under the fitted sheet near the edge of the bed creates a version of a concave mattress, which may reduce the risk of falls out of bed. Commercially available concave mattresses can also be used. It is critical that rigid bedrails, if they become necessary, be covered with soft material, such as foam tubing, as they can result in additional injury for active sleepers.

For persons who roll or maneuver out of bed, mats may be helpful to soften their landing on the floor. Keeping the bed in the lowest possible position will also decrease the risk of injury if the person with HD does fall out of bed. Low bed frames may work best in a facility. Furniture, such as dressers and headboards, can also be padded at the edges and corners with adhesive foam, to prevent injury.

If there is concern about the person with HD trying to get out of bed alone when they are not able to safely do so *and* their cognitive deficits reduce the reliability of a timely call for help, implement use of a **bed alarm**. Bed or chair alarm systems are available for purchase or rental, such as through Patterson Medical (http://www.pattersonmedical.com/). A facility might consider having the room of the person with HD near the nurses' station, as would be done for others at risk for falling at night.

Instrumental Activities of Daily Living (IADLs)

The following select IADLs were chosen given the likelihood of applicability to persons who are receiving home health OT or facility-based OT. For further information on other IADLs, please contact the OT at the nearest HDSA COE (www.hdsa.org).

➢Communication Management

If a person with HD does not require consistent direct supervision, it is critical that the OT address how the person with HD can effectively communicate to call for help using a telephone or call bell. The ability to use a phone is important for emergency situations as well as for social interactions with loved ones. Persons with HD with diminished fine motor control can have difficulty using some styles of phones, especially cell phones. There are cell phones with a 911 button and cell phones with large buttons and speed dial numbers, which can be helpful. If motoric function is especially compromised, voice command systems are now standard in many cell phones. This may involve acquisition of a new skill for a person with HD, which may be difficult to learn.

For persons with HD with diminished functional cognition but fair to good motor control, consider the complexity of the operations of contemporary smartphones and ensure that they are able to effectively use their phone. Smartphones can serve as great cognitive aids for those persons who are able to use the reminder or calendar features, the maps and directions features, or the videos to reinforce task progression.

➢Health Management and Maintenance

Medications

As medication compliance is a critical component of symptom management for persons with HD, it is essential that they and their caregivers establish reliable medication management strategies. In order to avoid dropping pills, the person with HD can be more successful when **sitting down while taking pill(s)** and using the recommendations provided in the section on self-feeding. Also, pill(s) can be **placed in a firm, unbreakable cup**, and then the person with HD can use the cup to bring the pill(s) to the mouth (like drinking from the cup). Similarly, some persons with HD may do better when the **pills are placed in a sticky food**, such as applesauce or oatmeal. When considering this last option, it is helpful to review the speech-language pathologist's (SLP's) recommendations or consult with a SLP regarding the most appropriate strategies for oral management of nutrition.

Exercise

Involvement in an appropriate exercise routine is very valuable to one's overall health. Due to impairments, persons with HD often have difficulty participating in sports or vigorous activities that they previously enjoyed. As a result, it can be difficult to find an appropriate form of exercise. The other, perhaps greater, challenge for many persons with HD is apathy. Therefore, in order for a person to be successful with an **exercise routine, it must be well-incorporated into the daily routine** and must be an appropriate challenge in order to promote compliance. For example, many persons with HD would be unsafe riding a bicycle, due to diminished motor control and safety awareness. In contrast, using a stationary, recumbent-style bike could be an appropriate form of exercise. Input from the physical therapist and/or a recreational therapist can be helpful when trying to determine an appropriate form of exercise to add to a person's daily routine.

✔ **Please go to https://www.youtube.com/playlist?list=PLLQmMRDsNEY2_6crhI-z9yL9FCc0FLI-VD to view Clip #9 – Occupational Therapy**

✔ **Please go to https://www.youtube.com/playlist?list=PLLQmMRDsNEY2_6crhI-z9yL9FCc0FLI-VD to view Clip #11 – Using an Exercise Bike**

Smoking

Some persons with HD smoke cigarettes regularly. Ideally, it is best to encourage the person with HD to quit smoking, which requires ample support from caregivers and may require a physician's prescription for smoking cessation aids. It can be more difficult for a person who smokes to successfully quit smoking if other people in the home continue to smoke. In order for the person with HD to quit successfully, it may require that the family quit smoking.

For persons with HD who have decided to continue smoking, there are strategies for helping them remain safe while smoking. A person with HD with diminished motor control risks dropping cigarettes and causing a fire. It can be especially challenging to sustain a functional pinch on cigarettes because they are such small, light objects. The first line for safety is to have the person with HD **sit down at a table while smoking**. Sitting increases stability and reduces distractions. Leaning over a table, as was recommended for self-feeding, increases stability and reduces how far a cigarette could fall. It is also critical that any designated smoking areas are **free from flammable materials**. Thus, it is ideal if the smoking area is outside and has a type of flooring or ground that is unlikely to catch on fire (i.e. free from debris or dry leaves). Products that can help reduce smoking-related fires include using **electronic cigarettes, a smoker's robot, and/or a smoker's vest.**

➢Religious and Spiritual Activities and Expression

For some persons with HD and their families, it is important to remain involved in their religious organizations or religious practices, especially given the stress involved in facing the disease process. As homebound status does not prevent a person from attending religious services, a home health OT can and should address any barriers or determine alternatives for persons with HD and their families who wish to be involved in their religious organizations. Considerations may involve addressing physical accessibility of the religious center, collaborating with the person's facility leadership and the religious organization's leadership to determine if it is possible to have religious events in the resident's facility, and/or determining if technology allows for an alternative avenue of participation (for example, using a video-streaming application to watch services or activities).

➢Safety and Emergency Maintenance

It is critical to assess whether a person with HD is able to safely be alone for *any* amount of time. In order for a person with HD to be safely alone even briefly, the person must be able to do the following: notice that a situation is occurring, recognize that the situation is unsafe, be able to initiate a timely and appropriate emergency response, and be able to communicate the situation on a 911 call. It is crucial to ensure that anyone with HD who will be alone participate in habit training to know how and who to call for help. Generally, encourage a person to **immediately remove themselves from the unsafe situation and call 911**. Accordingly, it is strongly recommended that persons with HD always keep a cell phone in their pocket so that they always have a way to call for help. Keep in mind that the person with HD must be able to use the cell phone appropriately. As noted previously, some cell phones have a quick-dial 911 bar.

Preparatory Methods and Tasks

Splints and Contracture Management

Contractures are the condition of shortening and hardening of muscles, tendons, or other tissue, often leading to deformity of joints. Persons in the advanced stages of HD may develop contractures, especially

as independent mobility decreases. OTs can evaluate the need for interventions to both prevent contractures and assist with contracture management. Splints, positioners, and the associated skin care must be individually evaluated and monitored by the OT. The use of commercially produced splints and positioners must be used judiciously after thorough assessment. Caregiver education is paramount for successful contracture management for persons with HD. Therapists should provide clear recommendations for splint schedules, as well as staff and patient education on how to apply the splints. Consider taking a photograph of the splint positioned correctly and then post a large copy in the person with HD's room. Skin integrity should be frequently monitored whenever splints are used. Redness and skin breakdown may occur if the fit is not proper or if the person's choreic movements impact the fit. Many medical supply companies have vendors that will assist in recommendations of splinting equipment and most splinting devices can be covered under the person's insurance policy.

Good documentation presenting the trial and usage of least restrictive devices provides support to the recommendations made by the therapists.

Conclusion

In conclusion, persons with HD can experience meaningful improvements in function and quality of life through OT intervention. Furthermore, the caregivers of persons with HD can gain valuable insight and skills to safely and effectively provide the necessary care. In each stage of HD, effective OT intervention requires a client-centered focus on the person, their meaningful occupations, and their environment.

Things to Remember

- There is a need for a client-centered approach to care that includes assessing motor, cognitive and psychological symptoms.
- It is important to understand a person's patterns, habits and routines with their ADLs in order to make recommendations.
- Involve caregivers in the assessment of the person.
- Accommodations for cognitive and motor deficits should be assessed in making recommendations in the following activities:

  Bathing issues:
    - Cognitive deficits – maintain routine and same order of washing
    - Motor problems – help with shower transfer and use shower seat

  Toileting
    - Cognitive deficits – regularly scheduled bathroom breaks
    - Motor- toilet grab bar, higher than standard toilet

  Dressing
    - Cognitive deficits – person will need extra time, verbal cues
    - Motor – consistently sit down to dress

  Feeding
    - Cognitive deficits- reduce distractions
    - Motor- prop elbows on the table
- When assessing comfort in a wheelchair, it is important to think about cushioning, seat angles, and positioning belts.
- Sleep disturbance is common for a person with HD so try to create a consistent sleep/wake routine, think about lowering the bed, softening bed rails, softening landing on the floor if person rolls out.
- An OT is an important team member to evaluate the need for splints and contractures in late stage disease.

Resources:

American Occupational Therapy Association. (2014). Occupational therapy practice framework: Domain and process (3rd ed.). *American Journal of Occupational Therapy, 68*(Suppl. 1) S1-S48. http://dx.doi.org/10.5014/ajot.2014.682006

Huntington's Disease Society of America. (2014). *Stages of HD.* Retrieved from http://www.hdsa.org/living-with-huntingtons/family-care/stages-of-hd.html

Nance, M. A. (2012). Therapy in Huntington's Disease: Where Are We? *Current Neurology and Neuroscience Report, 12*, 359-366.

Shumway-Cook, A. & Woollacott, M. H. (2012). *Motor Control: Translating Research into Clinical Practice* (4th ed). Baltimore, MD: Lippincott Williams & Wilkins.

Chapter 6

Huntington's Disease and The Role of Speech-Language Pathologists



Huntington's Disease Society of America

Huntington's Disease and The Role of Speech-Language Pathologists

Introduction

The goal of this chapter is to provide caregivers with a background of knowledge relating to the challenges of speech/communication, swallowing and cognition and to provide tips to optimize function within the long-term care setting. The caregiver should be armed with the knowledge necessary to make informed decisions regarding care for the person with HD. When properly educated on the basic anatomy and physiology of swallowing, symptoms associated with swallowing difficulties, common communication impairments as well as the cognitive impairments associated with HD, the health care team will be able to provide the best care. In any patient care setting, continuity of care is imperative – however, communication across disciplines and caregivers is particularly important when assisting a person with HD, and relies heavily on the education of every team member about each aspect of the care plan. In this chapter you will find all of the necessary information needed to understand and successfully follow through with the Speech-Language Pathologist's (SLP) recommendations. The American Speech and Hearing Association (asha.org) has excellent basic educational materials on its website about common symptoms seen by the speech-language pathologist, as well as common approaches to management of these symptoms.

Communication Impairments and Huntington's Disease

At some point during the progression of the disease, persons with Huntington's disease will develop communication and cognitive impairments. Most commonly observed may be motor speech impairments. A *motor speech impairment* refers to speech disorders resulting from neurologic damage impacting motor control of speech musculature or motor planning/programming of speech movements. Common motor speech impairments are:

Dysarthria: a motor speech disorder in which the muscles of the mouth, face and respiratory system become weak, move more slowly or not at all. There are various types of dysarthria and the severity and type depends on the area of the brain affected. Symptoms of dysarthria include: "slurred" speech; speaking softly or whispering; slowed rate of speech; rapid rate of speech or mumbling; reduced tongue, lip and jaw movement; abnormal intonation and rhythm; changes in nasality when speaking; hoarseness/ breathiness; and drooling or poor saliva control.



Apraxia of Speech: Also commonly known as verbal apraxia, this motor speech disorder involves difficulty with sequencing sounds in syllables and words. Apraxia can be seen in conjunction with dysarthria. Signs/symptoms include approximations of words (knowing what they want to say but the word comes out wrong). For example, a person may want to say "table" but may instead say "biple." The brain has difficulty coordinating the muscle movements necessary to complete the word and therefore the person may try again, only

to have a completely different word come out, such as “terple.” Due to the variety of errors presented, it can be challenging to understand a person with apraxia of speech and very frustrating for the person who is attempting to communicate and producing multiple errors. Persons with HD may have a particularly difficult time as well, due to poorly coordinated jaw, lip and tongue movements, which can further impair communication. This reduced coordination also makes it difficult for those with apraxia to repeat words or even single sounds correctly.

Other communication challenges that persons with Huntington’s disease face include difficulty with word-finding, stuttering, reduced voice quality (hoarse, breathy or harsh) and poor coordination of breathing and speech which impacts voice quality and volume.

Cognitive Changes in Persons with Huntington’s Disease

Cognitive changes may make it difficult for persons with HD to complete activities of daily living and may present in the following ways:

- Memory impairment (immediate and short-term)
- Poor reasoning/judgment – lack of awareness of deficits, reduced safety awareness
- Reduced problem-solving ability
- Difficulty sequencing/organizing thoughts or ideas
- Easily distractible/short attention span
- Poor concentration
- Reduced ability to learn new things
- Problems with numbers/mathematic computation

These kinds of cognitive changes make it difficult for a person with HD to complete simple tasks throughout the day, and therefore present challenges related to their care. Learning to develop clear, appropriate communication strategies to assist the person through activities is imperative in allowing them to maintain independence for as long as possible.

Cognitive impairments can cause communication breakdown in a variety of ways. A person with HD who demonstrates impairment in cognition may repeat themselves (or others), may get “stuck” on certain words or may not be able to monitor pragmatics or turn taking within a conversation and therefore may be seen as rude when they interrupt their conversation partner or change topics before allowing the conversation partner to complete a statement or thought. It is important for others to recognize that the pattern of communication is not indicative of personality, but rather impairment, and take extra care to be patient and understanding.

Cognitive Strategies:

Persons with HD may have difficulty learning new information. Therefore, the caregivers’ use of compensatory strategies during interactions with the person may aid in better communication and continued maintenance of cognition. Some common compensatory strategies for cognition include:

- Problem Solving – Caregivers in the facility should frequently talk to the person with HD about possible problems they may encounter and how to react. (e.g. What happens if you need to talk to the nurse?” – answer should be use call light, not attempt to get out of bed). Utilize situations within the facility to find ways to work on problem solving in functional ways before the problems become unmanageable.

- Orientation – Use a calendar (preferably large, visible) as well as signs (to indicate room/location) and clocks. Throughout the day, re-orient to month, date, year, time, place, etc.
- Memory – Use a schedule (and communicate about the schedule throughout the day), establish a routine specific to the person. Although staff will change frequently, it is important to have as much routine as possible. Speak with the team about having a scripted way of introducing each care team member, such as "Hi, Mary, I'm your nurse, I'll be with you for the day/night shift."

CASE STUDY

Mary, Amy's caregiver, posted a calendar in Amy's room and throughout the day directed Amy to use the calendar. In directing attention to the memory device (such as a calendar or "all about me" board with identifying information) Mary has an opportunity to communicate with Amy in the hope of improving behavior or maintaining a calm demeanor. Amy is able to read and recognize basic symbols and, with repeated direction, is aware of the signs around the room regarding safety and problem solving (using her call light, remaining in bed, etc.).

Tips to Improve Communication

Communication is how all human beings relay their thoughts, feelings, needs and wants to those around them and it is important that a person continue to communicate for both quality of care and quality of life. People with communication challenges are quite often left without a proper means to sufficiently relay this information and therefore it is imperative that all members of the care team involved with persons with HD have the tools necessary to optimize communication for all involved.

Tips for the Listener:

- Reduce environmental distractions (TV, radio, cell phones, computers, close door)
- Ask Yes/No Questions – do not use open-ended questions.
- Ask questions with only two or three options (e.g. "Do you want water or coffee?")
- Allow adequate response time – do NOT rush the speaker.
- Keep it simple! – Do not make communication too complex. Remember that cognitive impairments/ memory may play a role.
- Have a schedule – Let the person know what's expected. If memory is a problem, recognize that they may forget to do a task if it's not on the schedule.
- Only ask ONE question at a time – Wait for the answer, THEN move to the next question.
- Do NOT pretend you know what was said! – Ask clarification questions, rephrase, but do NOT give up. What they say is important and needs to be understood!

Tips to give the Speaker/Person with HD:

- Encourage the person to speak clearly/exaggerate the sounds.
- Encourage the person to speak slightly louder (especially if volume is reduced).
- Encourage the person to take a deep breath before speaking.
- Ask them to speak one word at a time.
- Encourage them to use gestures to assist with communicating.
- If he/she can't think of the word, ask to "describe it."

Augmentative and Alternative Communication

When persons with HD have significant verbal communication impairments an SLP can look into the use of another system of communication called Augmentative and Alternative Communication, or AAC. According to the American Speech-Language and Hearing Association (ASHA) website, AAC includes "all forms of communication (other than oral speech) that are used to express thoughts, needs, wants and ideas."

There are many types of AAC systems, from pointing and gesturing, to blinking and high-tech computers. Two basic types of aided communication systems can be considered for those with HD; *a static communication board* and a *dynamic display/speech generating device*.

Static Display – Static refers to items that are fixed to a particular location and do not change. Frequently, static communication boards will involve photos of commonly used objects or needs related to daily care. The person with HD can either point to the photo or with some low-tech voice output devices a caregiver or the person can record a message for each button.

✔ Please go to https://www.youtube.com/playlist?list=PLLQmMRDsNEY2_6crhI-z9yL9FCc0FLI-VD to view Clip #10 – Communication Tools

Dynamic Display Communication Devices – Dynamic display communication devices refer to devices in which the display changes to show other pages which may be linked by topic or other categorization techniques. Commonly, dynamic display communication devices are computer based and have synthesized speech for verbal output. Dynamic display communication devices can allow for the person to customize buttons or pages specifically to their needs and can change easily depending on the need or environment.

It is important to recognize that not all persons with HD can successfully use an AAC system. Cognitive function heavily impacts the successful use of communication devices or systems and therefore, if there is decline in function or inability to learn new information, AAC may not be appropriate. It is also important to consider the need for AAC and the implementation across disciplines. If a person receives a high-tech computerized speech device, many people involved in the care of the person may need to be trained on that specific system. A low-tech option like a static communication board, including an alphabet board for spelling and picture/symbol based options representing common items associated with activities of daily living, may be most appropriate and easily used by all involved in the person's care.

Dysphagia

The term dysphagia refers to difficulty swallowing. Dysphagia is very common in persons with HD, and is a major contributor to aspiration pneumonias and weight loss in persons with late stage HD. It is imperative that all involved in the persons' care are aware of signs and symptoms of dysphagia, as well as strategies to optimize oral intake and reduce the risk of aspiration. Laryngeal aspiration refers to any material entering the airway and passing below the level of the vocal folds. Laryngeal aspiration can lead to pulmonary infection or pneumonia.

There are multiple phases to the swallow mechanism, and persons can present with dysphagia involving any or all of the phases. Below are some descriptions of some of the phases related to swallow and symptoms of dysphagia associated with these phases.

- Oral Phase – Involves moving food throughout the mouth and preparing the bolus to be swallowed. Persons with HD may notice difficulty chewing or manipulating food or liquid within the mouth.

The tongue, lips and jaw may also be uncoordinated and therefore result in food being held in the mouth (delay in initiation of the swallow). Persons with HD may also have a poor rate/bolus control and take too large a bite. This is worsened with impulsivity. With oral dysphagia, you may also see food or liquid falling out of the mouth.

- Pharyngeal Phase – refers to the passage of boluses through the pharynx (throat). Problems in this phase include a delay in the swallow reflex, meaning that food and/or liquid spill over the base of the tongue and into the pharynx before the brain initiates the reflex to swallow. This can result in premature spillage of boluses before the swallow is initiated leading to aspiration of food or liquid prior to the swallow. Due to pharyngeal weakness and muscles being uncoordinated, it is also common that persons with pharyngeal dysphagia will demonstrate residue in the pharynx after the swallow. This makes it necessary for persons with HD to swallow multiple times per bite/sip. One hallmark of pharyngeal dysphagia is a wet voice or cough immediately before, during or after the swallow.

  The severity of dysphagia increases as HD progresses. Symptoms will progressively worsen and a persons' choreic movements may make it difficult for them to feed themselves or be fed by others.

Signs/Symptoms of Aspiration

As mentioned above, an early sign of difficulty swallowing will be a cough or throat clearing. Detailed information is important for the physician or the speech-language pathologist in order to develop an appropriate treatment plan. Does coughing or choking occur once or more per meal? Once per day? Only when in a reclined position? Only with liquids? Only with certain foods? The cough is the body's way of removing any material from the airway. Warning signs of dysphagia and/or aspiration include:

- Increased coughing episodes, especially immediately after swallowing.
- Wet sounding voice.
- Slower rate of eating or drinking.

When to consult with a healthcare provider:

- Wet sounding breathing.
- Frequent congestion.
- Fever, clear evidence of an upper respiratory infection.
- Continued weight loss.

CASE STUDY

Susie's aide, Camilla, noticed that Susie was having more difficulty at mealtime. Camilla reported to the charge nurse that Susie was coughing more frequently during meals and sounded congested after eating. Although Susie was recently changed to thickened liquids, she continued to have difficulty drinking liquids and the congestion persisted. She now has intermittent fevers, and refuses to eat her full meal resulting in weight loss. The charge nurse responded to Camilla's concerns and consulted the doctor to determine if Susie may have an aspiration pneumonia or if there is any underlying reason she may have had a change in status.

As symptoms progress, a person's hand to mouth coordination may deteriorate, directly impacting their ability to feed themselves. Modifications can be made to their food, such as offering more finger foods or preparing foods that are softer and easier to pick up with a spoon or fork. An Occupational Therapist can recommend adaptive or assistive equipment such as a built up handle for utensils or a mat that

stabilizes a plate or bowl. A sippy cup with a straw may help to prevent spillage when drinking liquids. The use of a straw can also limit the size of the sip, thus increasing the chance for a safer swallow. It's important to encourage the person with HD to maintain a neutral head position when drinking liquids rather than throwing their head back to drain the liquid from the can, bottle or cup.

The person with HD who has dysphagia is also in danger of not getting enough calories during a single meal or across the day, if they struggle with spilling food either from the utensil or from their mouth. Caregivers may be reluctant to assist the person with HD and most persons with HD want to maintain their independence and may not accept help.

Behaviors:

Impulsivity – Some persons with HD stuff food in their mouth and continue to do so without obvious or visible swallowing. This could lead to a serious episode of choking. The sequence of bite-chew-swallow before the next bite may be lost as cognitive decline continues. Verbal reminders or a light touch on the person's arm may be appropriate tactics to prevent impulsivity. Caution with liquids is also very important. Persons with HD may have a tendency to guzzle or gulp liquids without stopping for a breath. One sip at a time may be difficult to "enforce" but is a strategy that could work, especially if the person is experiencing coughing/choking during or after the swallow, or if they demonstrate a wet, gurgly voice quality when they're done with their beverage. Adult "Sippy" cups or a straw may be an option.

Excessive sleeping may impact the ability to meet caloric needs as meals are missed. It's important to encourage a regular schedule of meals and snacks.

Write out a schedule on sticky notes, or on the calendar in the kitchen. For example:

- ***Lunch at Noon – sandwich/pudding/milk are in the fridge***
- ***Snack at 2pm – ice cream bar in the freezer or Ensure® in the fridge***

Consult the person's medical provider for possible sleep aids to help regulate sleeping patterns to allow for more daytime wakefulness.

Food refusal is a behavior that may be related to the person's realization that the texture may be difficult to chew or swallow. Perhaps he/she had a previous choking episode related to that type of food. Food likes and dislikes may vary as the disease progresses or may vary from one day to the next for no discernable reason. Medication side effects may also affect how food tastes.

Excessive consumption of anything can be problematic (caffeine, smoking, alcohol). For example: During a recent clinic visit, a person reported that she typically drinks 2-3 diet sodas per day while her sister held up 6 fingers, indicating the actual number. Discussions around excessive consumption can become problematic when persons exercise poor judgment or are unaware of their excesses. Families and care facilities have reported coping with this behavior in a variety of ways, including a lock on the refrigerator or freezer, setting up a rigid schedule for the next caffeinated beverage during the day, or creating a reward system for proper eating.

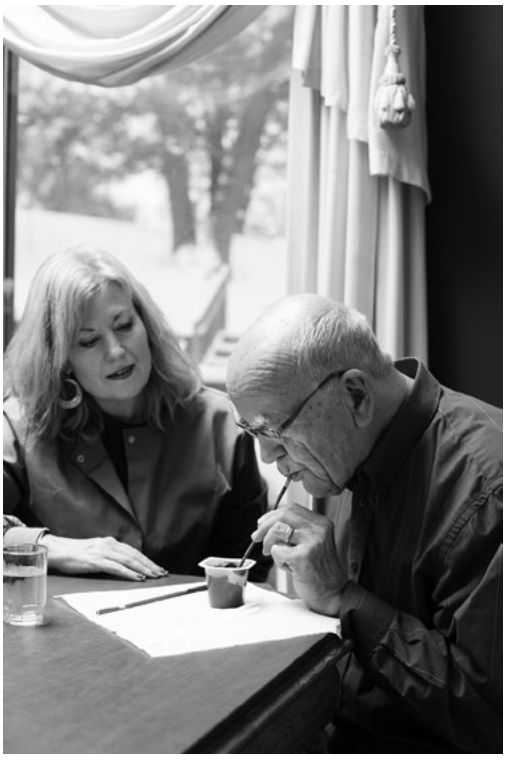
CASE STUDY

In an effort to prevent their loved one from binge eating during the day, Steve's family members were able to lock the door to the kitchen overnight while leaving snacks and beverages easily accessible when Steve was hungry. At the same time, they were dealing with Steve's excessive eating during meals and his rapid rate of drinking liquids and eating solids. One strategy, to remove the food from the table, did not work as Steve would then become angry and aggressive.

The family found that with a consistent 1:1 approach they were able to help Steve slow down during meals thus reducing the frequency of coughing/choking episodes. They also provided additional snacks after dinner such as ice cream or a high calorie smoothie to help prevent hunger during the night.

Dangerous food texture choices – While the person's chewing/swallowing coordination deteriorates even further, their judgment is also declining. Even though caregivers may carefully prepare balanced meals with texture modifications, the person's snack choices may be not be aligned. Here are examples of food textures that create "red flags."

- ***Particulate food:*** Nuts, popcorn, trail mix, dry rice, peas, corn. Placing a few nuts in the mouth at one time and chewing immediately breaks them into small particles – similar to placing a spoonful of rice, peas or corn in the mouth. If there is any tongue or jaw weakness or lack of coordination, this texture will be very challenging. While the person is chewing and coordinating the small particles into a single bolus, a small piece could fall into the airway causing a choking episode while particles are still in the mouth.
- ***Dense, chewy food:*** Meats like pork chops and steak, dense breads or bagels. Without intact chewing and coordination, this texture is especially challenging and often the first texture to require modification (i.e., chopping or grinding) or to be eliminated from the diet altogether.
- ***Dry, hard, crunchy food:*** Crackers, chips, raw carrots, apples. These types of foods, like the particulates, can also break into little pieces making coordination in the mouth difficult. The dry texture can make it difficult to move the bolus from the front of the mouth to the back where it can be swallowed. Soft, canned fruit and cooked vegetables are ways to provide important nutrients in a more manageable way.
- ***Stringy food:*** Lettuce, celery, pea pods. With this texture there's a sensation that it sticks in the throat making it difficult to swallow. It's easiest to eliminate these textures from the diet and substitute cooked vegetables as mentioned above.
- ***Some candy:*** Candy can be difficult to chew and/or swallow. Textures such as nuts and caramel can combine to create a challenging combination to chew. Choose softer, easy to chew candy bars or pieces of candy. Avoid types of candy that are sticky, crunchy, hard or chewy.
- **Holding food in the mouth** – This can be problematic not only for the person with HD, but also for the caregiver, especially if they're involved in feeding the person. The person may have difficulty moving the food or liquid from the front of the mouth to the back. This may be caused by difficulty chewing and manipulating the bolus secondary to chorea affecting the lips, tongue, jaw, vocal cords, and respiratory system, *or* there is deterioration in cognitive function that affects the oral phase of the swallow – the purposeful phase. A gentle reminder to swallow, and careful observation of the person's larynx (Adam's apple) to see the rise and fall that indicates the person has swallowed, will help to determine when it's safe to present the next bite or sip.



Developing strategies for modifying the texture of solid foods, and considering the possibility that liquids may be safer if thickened, will be vitally important as HD progresses. The change in swallow function happens gradually over time and family members often have first-hand, up close knowledge of this as they have witnessed a similar decline in other family members.

CASE STUDY

Tony's family members detected a pattern of coughing or choking with difficult to chew foods such as nuts or raw carrots. The Speech-Language Pathologist recommended elimination of specific textures of food. Foods like raw carrots or celery, salads, nuts or trail mix can be modified or completely eliminated from the person's diet. Vegetables can be cooked and salads can be finely chopped but items like nuts or trail mix may have to be eliminated.

A common scenario is that although the care provider is aware of changes that need to be made to food texture, the person adamantly refuses the changes – unaware of any difficulty or simply not wanting to be singled out as different.

Some strategies for the person with HD at meal times include:

- Sit upright at the table for all oral intake including medications. Avoid sitting in an easy chair in front of the TV.
- Avoid distractions when food or liquid is in the mouth. Caregiver should wait to ask a question until the person has swallowed.
- Encourage small bites and single sips. Remind the person to maintain a neutral head position instead of throwing the head back to drain the liquid from the cup. Introduce the use of a straw if the person seems to be coughing when drinking from a cup.
- Encourage a second swallow to clear food from the mouth.
- Have the person with HD take a sip of liquid frequently throughout the meal to clear any residue in the mouth.
- For medications: Advise the person with HD to take one pill at a time with a sip of liquid. If coughing occurs, try placing the pill in yogurt, applesauce or pudding. Crushing the pill and placing the powder in applesauce may also be a safe option.

It is important to supplement what is eaten during meal times with snacks. Keep healthy snack options easily accessible such as single serving cups of diced peaches, pears, yogurt or applesauce. Other options such as sport shakes, individual cans of tomato/vegetable juice, smoothies, and string cheese can be stocked in the fridge for easy access. If weight loss is a concern, consider snacks higher in calories such as ice cream, milkshakes, pudding or full fat yogurt. Drinking water throughout the day is important to maintain hydration but it may be appropriate to substitute beverages with calories like fruit juice or milk (at least 2% or whole). Supplements, such as Ensure® or Boost®, may be recommended by the physician or dietitian.

When is it appropriate to thicken liquids? What are the implications? If the person is coughing with regular thin liquids, first try introducing a smaller sip or a single sip at a time or try a straw. Notice if there's a pattern that results in coughing like talking with liquid in the mouth, distracted drinking, too large of a swallow or reclined in a chair in the living room. Next try naturally thicker liquids such as tomato juice or a smoothie. Most physicians and speech-language pathologists agree that a formal swallowing study is indicated prior to thickening liquids on a "permanent" basis. (See Modified Barium Swallow Study(MBSS) description on next page.)

CASE STUDY

Carol, a 56 year old female, was noted to have a documented 30 pound weight loss during the previous 9 months. It was important to evaluate the types of food textures that were the safest or to determine if dysphagia had caused recent and significant weight loss. Results of the MBSS revealed no evidence of aspiration but significant difficulty chewing and manipulating the food in her mouth. The recorded results of the swallowing "x-ray" were reviewed with Carol and her family, and the decision was made to provide softer foods with added moisture to allow for easier chewing and movement of the food in her mouth.

Indications that may lead to an alternative feeding method such as a gastrostomy tube occur when the person with HD continues to lose weight, is fearful of eating or drinking, or is malnourished or recently hospitalized for pneumonia. This is a discussion that the person with HD, along with their family members/caregivers, should ideally engage in early in the course of the disease. The physician, speech-language pathologist and/or dietitian are all professionals that can ask the questions and engage in discussion with the person/family.

Speech-Language Pathologists are professionals who are trained to evaluate swallowing and determine the most appropriate diet for the person with the goal to swallow safely, maintain nutrition and hydration and avoid aspiration. In a clinic setting, their involvement typically begins with a swallow exam in the office that includes an oral mechanism examination, a motor speech examination and the swallowing assessment. Further evaluation can include a Modified Barium Swallow Study (MBSS), a test that takes place in the Radiology department. During this test, the person is seated in a chair and fed various textures of barium to represent different food textures and liquid consistencies. This allows the speech-language pathologist and the radiologist to accurately evaluate the type and severity of dysphagia, and observe any aspiration of material.

FOR MORE INFORMATION

American Speech and Hearing Association website, asha.org, has useful information for health professionals and patients/families.
http://www.asha.org/public/speech/disorders/AAC/#sthash.LzxCVm31.dpuf)

Things to Remember

- Communication impairments/changes will occur with all persons with HD.
- There is a need to develop clear communication strategies to deal with a person's cognitive impairment and not just their changes in speech.
- A simple communication board with pictures and a few words may be the best option to assist a person with HD
- It is important to be aware of the signs of dysphagia (excessive coughing or throat clearing) so modifications in diet can be addressed.
- Be aware of a person's eating style of stuffing their mouth with food that may increase choking episodes. Attention should be made to aid the person to sip and take one bite at a time.
- Some textures of food are difficult for a person with HD to manage safely. Use caution with dense meats and breads, dry crunchy foods, stringy foods and some hard candies.

Chapter 7

Huntington's Disease and the Role of Nutrition



Huntington's Disease Society of America

Huntington's Disease and the Role of Nutrition

Introduction

While the guidelines about the importance of good nutrition are appropriate for the general public, nutritional issues and eating with HD introduce additional considerations. Diet cannot prevent or cure HD, but it contributes to maintaining quality of life and maximum functional ability which is paramount in care.

Nutrition and Huntington's Disease

Research has shown that Huntington's disease often manifests as a lower than average body weight for height and may produce higher than average calorie needs. This may be due to chorea, metabolic changes, or some other factor yet undiscovered. There is also anecdotal evidence that maintaining a body weight slightly above "desirable" weight (about 110% DBW) will facilitate control of the disease. Any significant weight change, whether increase or decrease, should be reported to the physician. Along with adequate calories, maintaining a sufficient intake of all essential nutrients is important. Meeting the DRI (Dietary Reference Intake) levels for vitamins and minerals can easily be accomplished with a little planning, attention to a varied intake, and appropriate use of supplements.

In disorders resulting in increased calorie needs, it is often necessary to encourage eating even in the absence of hunger. However this is usually not true with HD. More often persons with HD have excellent appetites and sometimes eat very quickly. Frustration at being unable to get sufficient food down quickly enough without choking can exacerbate the psychological problems associated with food and eating in Huntington's disease.



As the disease progresses many activities become more difficult or impossible to perform. Maintenance of independence in self-feeding becomes increasingly important at this time. Foods should be selected with this in mind. Items that are easier to manipulate, such as finger foods, may be appropriate. Also, with diminished capabilities in other areas, catering to food preferences provides an important psychological boost.

When it is no longer possible to meet nutritional needs with an oral diet, enteral feedings (tube feeding) may be initiated. Tube feedings may be given as a supplement to an oral diet, to provide extra fluids in case of swallowing difficulty, or as a sole means of nutritional support.

All of these issues will be discussed in the later sections.

Nutrition and Brain Health

Nutrient Dense Foods / *Calorie* Dense Foods

According to the 2010 US Dietary Guidelines, "Nutrient-dense foods provide vitamins, minerals, and other substances that may have positive health effects, with relatively few calories. They are lean or low in solid fats, and minimize or exclude added solid fats, added sugars, and added refined starches, as these add calories but few essential nutrients or dietary fiber. Nutrient-dense foods also minimize or exclude added salt or other compounds high in sodium. Ideally, they are in forms that retain naturally occurring components such as dietary fiber. All vegetables, fruits, whole grains, fat-free or low-fat milk and milk products, seafood, lean meats and poultry, eggs, beans and peas (legumes), and nuts and seeds that are prepared without added solid fats, sugars, starches, and sodium are nutrient-dense." Foods that are low in nutrient density are foods that supply calories (energy) but relatively small amounts of micronutrients, or none at all (empty calories). Efforts have been made to quantify these general guidelines in order to plan and/or evaluate diets on the basis of nutrient density.



The concept of ***nutrient density*** is beneficial advice for the general US population, where weight and obesity are substantial health issues. However, since maintaining appropriate weight can be a challenge for persons with HD, ***calorie-density*** is also an important consideration. Calorie-dense foods have relatively high calorie content per serving. Foods that are high in both nutrients and calories (for example: avocados, eggs, or nut-butters) are recommended to help maintain adequate intakes.

"Brain-healthy foods"

Based on research literature for the general population and some of the more common neurological conditions, such as dementia, there are some dietary factors considered to be "brain healthy". Two of the most important dietary factors are anti-inflammatory agents and anti-oxidants.

Healthy Fats

Some mono and poly-unsaturated fats have been found to have anti-inflammatory properties, and have been shown to be of benefit in a number of chronic degenerative diseases. Sources include: olive oil, fish oil, avocado oil, walnut oil, evening primrose oil, borage seed oil, and flax seed oil which all contain proportionately higher amounts of omega-3 fatty acids. These can be incorporated into the diet, or taken as supplements.

Several studies have looked at the potential benefits of supplementation with unsaturated fatty acids and eicosapentaenoic acid (EPA) in HD, with positive results. However, a 6-month randomized placebo-controlled clinical trial by the Huntington Study Group – TREND-HD (2008) found no differences between EPA treated and placebo groups.

Antioxidants

There are numerous compounds known to have antioxidant properties. A number of these are "phyto-nutrients," found in fruits and vegetables. While no one specific antioxidant is recommended, a diet rich in a variety of fruits and vegetables is appropriate for anyone. Color is important when choosing foods with antioxidant properties – foods with deep, rich color tend to be higher in antioxidants. The table on the next page lists some examples of food sources of various phyto-nutrients. Supplements of several different antioxidants have been studied or are being studied in HD animal models; to date, none have any proven benefits with HD. Vitamin E and Coenzyme Q10 have been proven to be ineffective in large clinical trials.

Color Code Sources of Phyto-nutrients in Vegetables and Fruits

Color	Phyto-nutrients	Sources
Red	Lycopene	Tomatoes, pink grapefruit, watermelon, guava
Purple/Red	Anthocyanins, polyphenols	Berries, grapes, red wine, prunes
Orange	α-, β-carotene	Carrots, mangoes, pumpkin
Yellow/Orange	β-cryptoxanthin, flavonoids	Cantaloupe, peaches, oranges, papaya, nectarines
Green/Yellow	Lutein, zeaxanthin	Spinach, avocado, honeydew, collard and turnip greens
Green	Sulforaphanes, indoles	Cabbage, broccoli, Brussels sprouts, cauliflower
White/Green	Allyl sulphides	Leeks, onion, garlic, chives

Data from: Heber D. Vegetables, fruits and phytoestrogens in the prevention of diseases. Prostgrad Med 50:145, 2004.

Vitamin D

Vitamin D deficiency and insufficiency are common in older institutionalized people. Symptoms include muscle weakness, increased body sway, and impaired balance, which puts these individuals at higher risk of falling. Since Vitamin D deficiency is also associated with osteoporosis, the risk of fractures is multiplied.

Chel et. al (2013) examined serum 25(OH)D levels in 28 persons with manifest HD. 89% (n=25) of these were found to be deficient in Vitamin D. Furthermore, a positive association was found between serum 25(OH)D levels and the UHDRS Functional Ambulation Classification Score. This suggests that Vitamin D deficiency may be common and functionally important in HD.

Vitamin B12

Vitamin B12 is found in animal foods (meat, dairy, eggs, poultry, etc.). Prolonged deficiency of this vitamin may have neurological effects on the brain and can cause nerve damage, although these effects are not specific to HD. Most people in the US get enough B12 if they include animal foods in their diet, only a small amount of this vitamin is needed, so eating a variety of foods containing B12 should be sufficient to meet the needs of most individuals. An exception would be individuals with decreased gastric secretion of intrinsic factor, needed for absorption of this nutrient. There is no specific relationship between HD and problems with absorption or handling of B12 by the body.

Caffeine intake

Although caffeine intake has been shown to be of potential benefit in other neurological disorders, a recent study by Simonin et al. (2013) showed a link between caffeine consumption of greater than 190 mg/day and earlier age of onset of HD symptoms by as much as 4 years.



Calorie Supplements

Supplemental shakes or puddings can be added to the diet in order to boost calorie and protein intake. Like many food products, persons with HD often have their preferences as to brand and flavor, although

nutritional differences may only be minor. It is convenient to have pre-mixed cans available as they have a fairly long shelf life. Most such products are lactose and gluten free, and usually also Kosher. Some persons with HD like to use these as a base for their own calorie enhanced beverages. A home-made smoothie can provide easy-to-drink calories, and many recipes are available. *(See references at the end of this chapter)*

Effects of HD on Appetite



It appears that lowered levels of appetite-suppressing substances in the brain may be a cause of increased appetite in HD. Increased appetite, combined with impulsive behavior, may cause a person with HD to eat more rapidly, and increase the risk of choking on food.

Increased thirst may also play a role in HD. In one study, persons with HD were significantly more likely to report problems related to dry mouth such as difficulty in talking, chewing, swallowing, and sleeping. Saliva stimulants (such as Pilocarpine) may relieve these symptoms. Artificial saliva products may also be helpful.

Effects of HD on GI Function

Various gastrointestinal symptoms, such as GERD, regurgitation, constipation and diarrhea have all been reported with HD. There have been few studies looking specifically at GI symptoms, because unlike Parkinson's disease, which routinely affects intestinal motility, this does not appear to be a major or common issue in HD. However one study found that 44% of persons in various stages of HD tested positive for gliadin antibodies, reinforcing the importance of ruling out celiac disease, lactose or soy intolerance, or other common conditions unrelated to HD. GI symptoms can also be secondary to medications commonly prescribed. It is especially important to review the medication list closely in a person who is either gaining or losing weight unexpectedly. A number of medications commonly used for mood or behavioral issues in HD can cause weight gain, such as olanzapine, risperidone, haloperidol, valproic acid, and mirtazapine. Others, like methylphenidate or citalopram, can cause weight loss. Dry mouth can be a side effect of medications that have anticholinergic properties. Many drugs can also cause mild degrees of nausea.

CASE STUDY

Mary, a 48 year old woman living at home, presented at an HD clinic with weight loss and complained of a chronic metallic taste in her mouth, possibly related to some of her medications. Her appetite was good, but she would only eat foods with a sour taste, since that was all that was appealing to her. She also experienced mild dysphagia. She was referred to the RD for assistance with a diet plan. Per discussion with Mary and her family, foods were to be made with thick citrus-flavored sauces, vinegar, and other tart ingredients added to other foods to create her preferred taste profile. A brand of lemon pudding was also identified that Mary was willing to try. On her next clinic visit, Mary's family reported her intake was better and her weight had stabilized.

Constipation is a common problem in the late stages of HD for several reasons. A late-stage person may be bed-bound or wheelchair-bound, and therefore less active. Inability to drink sufficient fluids, or the need to use thickened fluids, reduces the moisture in the stool. The use of narcotics and other sedatives and muscle relaxants may reduce intestinal motility as well. Constipation can, in turn, lead to reduced appetite and weight loss. Initiation of a constipation protocol may help to increase comfort and improve appetite.

Adaptive Equipment – Role of OT

When self-feeding becomes difficult, an occupational therapist can make recommendations for adaptive equipment. This may include weighted or wrap-around utensils, utensils with specialized grips, bowls and plates with angled or built up sides, or other items. The goal is to promote independence in self-feeding as much as is possible. *(See Occupational Therapy chapter)*



Effects of HD on Swallowing

Dysphagia is a common complication of HD, with choking and aspiration being potentially lethal. Abnormalities of self-feeding, chewing, bolus formation, and the pharyngeal phase of swallowing may all be present.

Role of the Speech-Language Pathologist

Speech and language therapy has an important role in the management of Huntington's disease. The speech-language pathologist helps with the assessment of swallowing, screening for difficulty with certain textures of solids or liquids, as well as changes in the act of eating, that can lead to coughing, choking and ultimately to weight loss or aspiration pneumonia. The speech-language pathologist works closely with the dietitian to identify and implement a safe, healthy, and pleasurable diet.

The speech-language pathologist and the dietitian should be part of the discussion regarding non-oral feeding/supported feeding. This discussion should be introduced early in the disease course, while cognitive abilities still allow full understanding and participation in the decision making process.

(Also see Coping with Speech and Swallowing Difficulties in Huntington's Disease by Karen Bryant, PhD, CCC-SLP, HDSA 2014 (www.hdsa.org) as well as Speech-Language chapter).

The National Dysphagia Diet (NDD) is the standard of care for adjusting diets for persons with swallowing problems. A Registered Dietitian (RD) can use the NDD guidelines to help to create a menu that incorporates favorite foods in a way that reduces the risk of coughing or choking.

Thickened liquids are an important part of a diet for safer swallowing. Standard descriptive terms for thickened liquid consistencies that are used in the NDD include: "nectar thick", "honey thick", and "spoon thick." Nectar thick liquids have slightly more body than thin liquids, but still can pour easily. Some examples include tomato juice, pear nectar, peach nectar, apricot nectar, and some bottled fruit smoothies. Honey thick liquids are liquids that pour slowly, like honey or molasses. An example would be a very thick milkshake. Spoon thick liquids could be eaten with a spoon, like pudding or custard. Spoon thick liquids are too thick to drink with a straw.

Consultation with a Registered Dietitian

A dietitian should evaluate every person with HD on admission to the care facility, and again at any time if there is weight loss, progressing dysphagia, difficulty with the mechanics of eating, or the need to initiate enteral feedings.

A comprehensive nutrition assessment should include evaluation of:

- Appetite, ability to eat with or without assistance
- Any episodes of choking or problems with swallowing

- Any recent changes in intake or eating habits
- Weight history, especially recent weight changes
- Alcohol use
- Arrangements regarding food shopping and cooking
- Any previous diet counseling/education

There is some evidence that total daily intake becomes more irregular with advancing disease. Additionally, dementia may contribute to inaccuracies in reporting intake. It is therefore important to include family/caregivers as informants in obtaining intake data and to inquire about intake over at least several days to try and capture the variability of daily intake.



For men:
B.E.E. = 66.5 + (13.75 x kg) + (5.003 x cm) – (6.775 x age)

For women:
B.E.E. = 655.1 + (9.563 x kg) + (1.850 x cm) – (4.676 x age)

Total Caloric Requirements equal the B.E.E. multiplied by the sum of the stress and activity factors. Stress plus activity factors range from 1.2 to over 2.

Harris J, Benedict F. A biometric study of basal metabolism in man. Washington D.C. Carnegie Institute of Washington. 1919.

Calculations for energy needs

When calculating estimated calorie needs, adjustments must be made to the stress/activity factors to account for the impact of HD. When using the Harris-Benedict equation to estimate energy needs, one study used an adjustment factor of 1.3. Another indicated an activity factor of 1.4 to 2.1. Comparable adjustments should be made with other estimates of energy expenditure.

Meeting Calorie Needs

Eventually eating begins to require so much energy and concentration that meals result in fatigue and frustration before adequate amounts of food can be consumed. Significant weight loss or difficulty using utensils or handling food are other reasons to discontinue self-feeding. The risk of choking also tends to become less frequent when self-feeding is stopped, since the caregiver can regulate the size of bites and rate of eating. Before providing another bite, the caregiver should continue to remind the person to slow down, refrain from breathing when swallowing, and to search the mouth for food or provide a sip of water before introducing another bite.

Finally, the transition to assisted eating at mealtimes does not have to be an “all or none” decision; the person may be able to continue eating some things unassisted, while being fed when necessary.

If the person is in the right frame of mind and setting (properly positioned in a calm, quiet room, and assisted by a known caregiver), he or she can eat more than one might expect, and perhaps be fed by mouth longer into the illness.

Emphasis on a quiet room with few to no distractions is critical especially in late stage illness, so the focus is only on eating.

Feeding Tubes and Regimens



Although a nasogastric tube may be used temporarily, for longer term nutritional maintenance a percutaneous endoscopic gastrostomy (PEG) tube is placed directly into the stomach. Formula flows through the PEG tube (also called a gastrostomy tube or G-tube) into the stomach. Since the tube does not pass through the throat, it is possible to receive both tube feedings and an oral diet. It is usually desirable to continue oral feeding for as long as possible, even if the quantities given are not nutritionally significant, since this can provide significant psychological benefits. As an alternative to the standard PEG tube, a "button" tube may be used. This is a very short tube attached to the stomach with a longer "snap on" tube for use during feedings. When the tube is not in use, a plastic cap covers the opening. These can be useful for persons who do not receive a 24-hour feeding.

Another option is a jejunostomy tube (J-tube), which is implanted below the stomach, directly into the small intestine. It functions similarly to the G-tube, but with several differences. The advantage of a J-tube is that it reduces the risk that formula will come back up the esophagus into the trachea and lungs. For someone at high risk of aspiration, the J-tube might be the preferred option. Increased probability of diarrhea and increased probability of the very narrow tube getting clogged are some potential disadvantages of this kind of tube.

Feedings are usually prescribed as either "continuous" or "bolus" servings. As the name suggests, continuous feedings run down the feeding tube at a fixed rate throughout the day and/or night. Tolerance problems are usually minimized when feedings are given this way. A very weak or debilitated person may need to be fed continuously in order to tolerate enough intake to meet his/her needs. High risk for regurgitation because of limited stomach capacity is also an indication of need for continuous feeding.

Most persons with HD who are just starting out on tube feedings are given continuous feedings, then gradually changed over to bolus feedings. A "bolus feeding" is a set amount of formula that runs down the feeding tube at specific times during the day. Bolus feedings are essentially the equivalent of a meal consisting of formula. These usually, but not always, correspond to breakfast, lunch and dinner times. Some regimens include one of more bolus "snacks" as well, for a total of four to six feedings per day. Bolus feedings are usually more convenient for caregivers, since feedings are administered only at specific times and larger amounts are given at each sitting.

CASE STUDY

Jeff, a 52 year old man, with early onset HD developed severe dysphagia and weight loss, requiring a PEG placement. Jeff was not keen on having a "stomach tube," but he was spending all his waking hours trying to consume enough food to prevent further weight loss. His family caregivers also constantly offered him food and supplements and encouraged him to eat, but his weight continued to decline. Jeff also stated that he found their constant reminders to eat "really annoying." After PEG placement, he was started on 4,000 calories of formula as five bolus feedings per day. He tolerated the feeding well, and continued some PO foods for enjoyment. His weight and strength increased with this regimen. Upon his return to the clinic he commented that he no longer had to spend his days trying to eat. When asked what he was doing with his time now that it was not being spent trying to eat, he replied "watching baseball…. those Yankees need to work on their relief pitching!"

Some additional tips for caregivers about tube feedings

- Position the person so that he/she is sitting up or at least so the upper-body is above the level of the stomach.
- Practice good sanitation. Wash your hands before handling the feeding equipment. Wash feeding bags with water. Do not use soap, as it will stick to the inside of the bag and get into the formula. This can cause diarrhea.
- Feedings should be given at room temperature to minimize the risk of cramping and/or diarrhea. Open cans of formula should be kept in the refrigerator, and discarded if they are not used in 24 hours. Refrigerated cans should be taken out 15-20 minutes before a feeding and allowed to warm to room temperature.
- Always flush the feeding tube with water after a feeding. This will help to prevent the person from getting dehydrated. It will also prevent the tube from getting clogged.
- If medications are to be given through the tube, use liquid forms if available. If not, always be sure that the pills are finely crushed. Flush the tube with water to wash them down.
- If the tube seems to be clogged and a small flush of water isn't effective, a flush of about 100mls of cola may do the trick. You can also try dissolving about ¼ teaspoon of meat tenderizer in a teaspoon of water and placing it into the tube. Wait five minutes before flushing again. If none of these strategies work, contact your healthcare provider(s) for assistance.



Summary Guidelines for Dental Care

The European HD Network recently wrote a comprehensive guideline for dental care in HD, which we have found to be very helpful for persons with HD, families, and dental professionals. See the link below to access this document.

http://mun-h-center.se/upload/MunhDoc/Guideline%20for%20oral%20healthcare%20of%20adults%20with%20Huntington's%20disease.pdf

Thnigs to Remember

- Diet can't cure HD but it can contribute to a better quality of life.
- Persons with HD tend to have lower body weight and have a need for higher calories to maintain their weight.
- Diets should be both nutrient and calorie dense for a person with HD.
- Persons with HD can maintain a good appetite but have impulsive behaviors that can increase the risk of choking.
- A registered dietician can adjust the diet for persons with swallowing problems. This includes the use of thickened liquids.
- Because a person with HD may expend a lot of energy and become fatigued when eating, a feeding tube may be an option when the person has substantial weight loss.

Food / Cooking References:

Many books and resources are available on-line. Additional recipes which may be appropriate can be found in:

The Non-Chew Cookbook By J. Randi Wilson

The I-Can't-Chew Cookbook: Delicious Soft Diet Recipes for People with Chewing, Swallowing, and Dry Mouth Disorders By J. Randi Wilson



The Dysphagia Challenge By Pam Womack, RD

Easy-to-Swallow, Easy-to-Chew Cookbook: Over 150 Tasty and Nutritious Recipes for People Who Have Difficulty Swallowing Donna L. Weinofer, MS, RD, Joanne Robbins, PhD, CCC-SLP, and Paula A Sullivan, MS, CCC-SLP.

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2010 Dietary Guidelines (pg 35)

http://www.cnpp.usda.gov/Publications/DietaryGuidelines/2010/PolicyDoc/PolicyDoc.pdf

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Chapter 8

The Role of Recreation Therapy and HD



Huntington's Disease Society of America

The Role of Recreation Therapy and HD

A typical person with HD in a care facility is likely much younger than the average nursing home resident. Care should be taken to find activities like music, games and movies that are age appropriate for this younger group. It is important to keep in mind the person's level of function, including the ability to use their hands for fine motor tasks, be ambulatory and vocalize or make their needs known in other ways. Getting input from other staff members about the person's possible behavioral issues or severe cognitive decline will assist as activities are planned. This chapter will share some activities that most persons with HD can participate in even with limited speech or need for adaptive equipment.

Exercise Programs

It is important to ensure safety for those with an unsteady gait for any exercise activity. Generally, group exercise programs are done with everyone seated, including those who can walk. This provides safety along with maintaining eye contact of all participants. An instructor can also work with an ambulating person with HD doing one-on-one exercise. If there are concerns about whether a particular activity is safe or not, or what activities are most appropriate, referral to the physical therapist for an individualized assessment can be helpful.

Guided imagery can be used asking persons to make concentrated slow, guided movements of one limb at a time. It is important to also concentrate on guiding their breathing while performing these movements. For those persons who are unable to easily move their limbs, they can be asked to close their eyes and, through guided imagery, concentrate and be aware of each limb and think of themselves moving that limb.



Exercise programs like Yoga can be used as a tool to offer support and behavioral coping strategies to find and sustain healthy mind/body balance. One possible outcome could be a calmer and steadier baseline in response to situations more reflective and less reactive. Breath awareness, centering, meditation (usually guided), and repetition of sounds or mantra can be used with possible added hidden benefits of assisting a person with speech.

The yoga instructor works with several persons with HD to do adaptive yoga.

Current Events

Many persons with HD continue to appreciate news updates. Newspapers can be delivered to those who can read on their own or be read highlights by a Recreation Therapist, family member or caregiver. Many persons value hearing about medical research, stem cell research, articles which focus on HD or other related neurological diseases as this provides them with hope that there are possible future treatments.

Sensory Massages

The use of massage and the power of touch remain important throughout one's lifetime. It can offer persons with HD the sense of compassion and being cared for. Communication during massages or touch builds a bond of trust between the person providing the stimulation and the receiver. Some persons with HD may experience extensive choreic movements causing tired, sore muscles that keep the person awake for hours. After a massage, a person with HD may experience relief and rest comfortably. Some persons with HD exhibit dystonia and contraction of their body and benefit from a slow, deep massage to aid in lengthening, relaxing and loosening their muscles in their extremities, shoulders, neck and jaw.

Water Play



The Recreational Therapist assists a person with HD during a group water activity.

The facility where I work has tried different methods of water play. Water balloons proved difficult for persons with chorea as their hand movements caused the balloons to break on themselves. Many also had trouble throwing wet sponges or utilizing a water gun. The game that seemed the most successful was hot potato using a large beach ball and a stereo. Initiate a circle, placing persons who may have trouble passing the ball between individuals with better hand coordination to improve the passing flow. When the music starts the ball is passed around the circle and when the music stops the individual with the ball gets blasted with water from those who are able to use a water gun (not in the face). After everyone is wet, the game continues and the rules change. The one left holding the ball when the music stops points or looks to the person they want to get wet. Nursing staff should be informed about these water activities as the group will likely be drenched.

Card Games-Black Jack

Black Jack is a game that can be played using only the dealer's hands. This simplified way of dealing can be played as a group activity or one-on-one to try to maintain the person's attention span. Some persons with HD require verbal cueing to maintain their focus on the task. It may be helpful to call their name when it is their turn and then wait before you begin to speak to get their full attention. Start by

dealing the first person two cards laying face up in front of them, so they can see the cards, and tell them the total of the cards in front of them. Next, tell the person what cards they will need to stay under 21. Ask if they want to 'hit' (for another card) or 'stay' (to hold). The dealer is likely to encounter a delayed response from the person which is common in HD. Those persons with limited speech can touch the table or chair for yes or use eye contact to confirm 'hit' or 'stay.' Once the first player completes their turn, others around the table can get a turn. After everyone has played the round, the dealer can flip his/her cards trying to get close to 21. If the dealer goes over, every player who did not gets one chip. If the dealer does not go over 21, every player who gets closer to 21 than the dealer gets a chip and those who did not get close to 21 get nothing. Cards are collected and a new round starts. This can be played for as long as you and the person wishes to play and the winner is whoever has the most chips. Other varieties of 21 (like Five Card Charlie and Six Card Bertha) can increase the number of chips available. Other card games can be played with only the dealer manipulating the cards including Five Card Stud and Seven Card Stud Poker.

Smoothie Socials

Smoothies can be made to order for all persons with HD whether they are on regular thin liquids or on pudding thick liquids. Persons can choose to add ingredients like blueberries, thought to be a source of antioxidants. Try a recipe with one cup of frozen blueberries and a banana placed in a blender with the addition of heavy cream covering the fruit. Heavy cream is used for its high calorie count. Blend well. Add flavored yogurt to the top of the blender. Continue blending until smooth. The smoothie will have a thick consistency that can be enjoyed by most. If needed, add less heavy cream and more yogurt to make pudding thick or more heavy cream and less yogurt for those on thin liquid diets. For those on thick liquid diets who are also lactose intolerant, chocolate flavored soy milk can be used with extra frozen blueberries and extra bananas to make the smoothie thick.



Horticulture

Horticulture Therapy can be used with persons with HD who can manipulate tools or use their hands easily. Some persons prefer to use their hands while others prefer oversize handles on scoops. Large, deep, high-sided bins with one low side offer access when working with soil and water indoors. Persons can place their arms easily inside the bins to work with the soil to plant. Once they are done potting a plant, the soil is taken out and the same bin can be used to hold the potted plant. Persons can use a watering can without worrying about spilling water on the table. Persons with extensive chorea may require hand-over-hand assistance. Persons who enjoy the water fight activity may also enjoy horticulture.



The Horticulture Therapist works with persons with HD doing adaptive horticulture

It is important to choose sturdy plants that can be handled without breaking the stem. Plants should be pleasing to all of the senses including scented, flowering, herbs, vegetables and tactile plants such as the velvety leaves on an African violet to the healing qualities of an Aloe Vera plant. The best soil option is a soil-less potting mix that is sterilized and perfect for growing new plants and seeds.

Supervision may be needed so that confused persons with HD do not mistake the scoop for a spoon and the smells of the plant as food (especially if the activity is being held in a dining area).

Horticulture outside is another option. Those in wheelchairs can work in raised gardens and use a hose to water plants.



The Horticulture Therapist works with a person with HD as they water the plants in the facility's day room.

Animal-assisted Therapy

Persons with HD often respond with great joy to therapy dogs, who respond to touch and do not mind an occasional choreiform movement or spill on the shirt or floor. Even a person who is mute can express enthusiasm with an affectionate dog. Others enjoy the warmth of a cat on the lap.



Music Therapy/Live Music

Some persons with HD may be able to play an instrument but for others their ability may have declined. Music Therapy provides persons with HD with an opportunity to continue playing instruments adapted to their abilities. Some may be able to hit keys on a keyboard, hit a drum with or without a drumstick, strum a guitar or other stringed instrument while others enjoy bells strapped to their wrists that make music with every movement.

Other persons may benefit from music at their bedside from a stereo, MP3 player or from live musicians, concerts or themed parties.

Arts & Crafts

Persons with HD may vary in their ability to do arts and craft projects. Use caution when using paintbrushes or other sharp objects, such as scissors, that could cause harm to a person with excess choreic movements. It is best to use paintbrushes with small round handles that fit in the palm as opposed to long stick brushes. One-on-one assistance may be needed but also the use of verbal cuing can help the person to maintain their focus for continued participation. Some persons may require hand-over-hand assistance while others, who may be unable to do the project physically, may have the ability to verbalize their desire of colors or design. Try to persist with some persons with HD who may need a lot of encouragement to speak their choices.

Computers

Many people know how to use a computer and many now use social networks like Facebook and e-mail. Once a person with HD can no longer maneuver a keyboard, live video conferencing such as Skype can become an important way they can continue to communicate with family and friends. Even if the person has lost the ability to speak, it is important to continue to be able to see one another live and share a moment or blow a kiss.

Millennials who have HD have grown up using the computer or other platforms for playing games. Computer gaming does not help a person to socialize, but depending on the game, may keep the mind and hands occupied in a useful way. For the person with a tendency toward compulsive behaviors, access to computer games may have to be monitored or restricted.

It is important to keep in mind that those who may kick or swing out be kept at arms/legs length away from others at a table or in a circle setting. Try to keep those who do not get along a fair distance from one another or out of their line of sight to avoid conflict. A person with HD who perseverates may require a lot of verbal redirection. It is often best to give the person an answer in response to what they are focused on before attempting to divert their attention back to the program. For example; if the person is focused on their computer not having adequate volume, respond with a time you are able to look at their computer, such as after the activity, and continue to reiterate this every time they ask. If their request can be fulfilled, then do so.



It is important to have background information on a person to better understand their temperament. Some persons may need more personal space to do well in their current environment. Maybe a person loves to talk about their family, sports or other hobbies. Having this knowledge can be useful as a distraction technique. Knowing when to change the subject and converse with them about what they enjoy can create a positive response. Some quality of life exists for all person with HD and an astute caregiver can assist a person with HD to continue to enjoy their passions at any level.

Chapter 9

Hospice Care in Huntington's Disease



Huntington's Disease Society of America

Hospice Care in Huntington's Disease

As a neurodegenerative disorder for which there is currently no cure, everyone with Huntington's Disease (HD) will eventually die within 10-25 years of diagnosis. Because of the relatively young onset of symptoms, most persons with HD are otherwise relatively healthy, and die of medical or traumatic complications of HD, such as aspiration pneumonia, other infections, malnutrition/cachexia related to dysphagia, or from complications of falls or other trauma.

The general principle underlying this guidebook is that good care from knowledgeable providers improves the lives of persons in the late stages of HD; similarly, hospice care can improve the lives of persons in the terminal stage of the disease. We discuss below how to recognize that a person is in the terminal stage of HD, what hospice care can provide for this person and the family, and some of the unique challenges that HD presents to the hospice provider.

Recognizing the Terminal Stages of HD

In the late stage of HD, a person requires assistance with all activities of daily life, due to the combination of dementia and loss of motor control. Usually, by the terminal stage, a person is unable to walk and/or unable to talk, and difficulty with swallowing may be causing aspiration pneumonia. Chorea, which may be very prominent in the early or middle stages of the disease, sometimes subsides towards the end, and is replaced by rigidity and hypokinesia (lack of spontaneous movement). In others, the chorea seems to worsen as the person moves into the latest stages of the disease. Although some persons in the late stages of HD seem to lose awareness of their surroundings, many seem to retain some interactiveness and personality even in the very late stages, when they are mute and bed-confined.

Because the progression of the disease is so gradual, over many years, it can be difficult to determine if a person in the late stage of HD has become hospice-appropriate. Hospice care is appropriate in the terminal stages of HD.



The typical characteristics would likely include some or all of the following: nonambulatory, with reduced or absent ability to speak intelligibly; recent history of aspiration pneumonias or other major hospitalization; multiple recent hospitalizations or visits to the emergency room, and weight loss despite aggressive attempts to feed.

These characteristics describe a person who has the neurological features of late stage HD, who is also showing medical instability and deterioration despite appropriate or aggressive medical care. As has also been reported in Alzheimer's disease, persons with HD can develop "terminal agitation," an unexplained escalation in agitation, screaming, or crying. While this may be difficult to recognize in a person who has substantial behavior issues at baseline, in some, the recognition of agitation as a sign of progression to the terminal stage permits an important acknowledgement that death is coming and a shift is needed in care priorities.

Long before the terminal stage of HD arrives, the person with HD and family should create a Living Will or Advance Directive, which addresses how they want care to be provided during the late stage of the disease. As dysphagia is such a common complication of HD, which in turn leads to weight loss and aspiration pneumonia, it is particularly important for the Advance Directive to address whether the person wants a feeding tube in the late stage of his or her disease. HD does not lead to situations in which cardiac resuscitation, dialysis, or intubation would be necessary, but recurrent infections of different kinds

can occur as the person becomes bedbound in the late stages. Persons with HD should also make clear who will make medical decisions on their behalf if they are no longer able either because of dementia, communication difficulties, or both.

A social worker, nurse, or other care provider who is new to a person in the late stage of HD, and who needs to discuss complex issues such as end-of-life care, may find it helpful to use a validated scale to measure cognitive function and level of depression, as they assess the person's competency to make decisions. The Mini-Mental Status Exam, Montreal Cognitive Assessment, and St. Louis University Mental Status (SLUMS) are all commonly used cognitive assessment tools, and the Beck Depression Inventory or The Terminally Ill Grief or Depression Scale can be used to assess depression.

Complicating the conversation with the HD-affected person is a tendency to unawareness or denial of symptoms, and to impulsiveness, which can lead to bad judgment or poor decisions. Children of the affected person, who are themselves at-risk for developing the disease, may have difficulty making decisions on behalf of their parent. At-risk children may make decisions for their loved one based on their own fears and engage in denial to escape their perception of their own future.

CASE STUDY

Beth was a regular visitor of her mother until her mother's symptoms worsened and Beth couldn't face her mother's decline and her own fears of inheriting the HD gene.

Early, ongoing, and open discussions are important to ensure that end-of-life care decisions are made in a timely and appropriate way. Communication between the hospice team and the treating physician can provide insight to family dynamics and provide continuity of care to enhance the hospice experience for the family and person with HD.



Goals of Hospice Care

In the terminal stage of HD, an affected person is receiving full assistance for all activities of daily living due to their inability to walk or talk, general medical decline, such as weight loss, recurrent hospitalizations or someone who has developed terminal agitation. Some receive this care at home, but many are living in a long-term care facility by the time hospice care becomes appropriate. Hospice may be provided at home, in the hospital, in the long-term care facility, or in a residential hospice. The specifics of cost to the person, insurance coverage, services provided, medical services and providers included or excluded within the hospice program, and duration of service, may vary depending on the state, the insurance plan, the hospice program, and the location where the care will be provided.



Just as for people with other terminal illnesses, the goals of hospice are to guide the affected person and the family through the dying process with comfort and dignity. The terminal stage of HD often includes a slow reduction in food and water intake, with an increasing tendency to cough or choke when attempting to eat or drink. The person becomes increasingly immobile, sometimes with prominent rigidity or dystonia (awkward posturing of a limb or muscle). The person is likely unable to communicate if there is pain, so the care provider will have to make a judgment as to whether there is an appearance of discomfort,

or agitated behavior that might suggest discomfort. Some persons may have irritable or inconsolable behavior, but more commonly there are longer periods of sleep and fewer episodes of interaction. There may be a recognizable terminal event, such as a respiratory infection or pulmonary embolus. Because HD does not affect other organ systems, such as the heart, lungs, kidneys, or liver, and persons with HD are often relatively young and otherwise healthy, the dying process is often a slow, gradual one.

The other goal of hospice is to help family members through their loved one's dying process. The support of hospice can help a person with HD to remain at home until death, or help a family prepare for the death of a loved one who has previously moved to long-term care. The genetic nature of HD, however, can create some complex family situations and dynamics of which the hospice team should be aware.



CASE STUDY

Susan is the oldest of four children in the family. Her father resides in a care facility and had an increase in choking episodes and is steadily losing weight. She is the only one of her siblings to test negative for the HD gene. She is overwhelmed with the responsibilities of the whole family that is slowly taking its toll on her health, marriage and work.

The genetic nature of HD, and its relatively young onset age, produces unique psychosocial stressors within an HD family. At-risk family members may internalize their loved ones decline; therefore complicating their grief. Family members not at-risk may have severe guilt that HD will not affect them but have watched other family members die as a result of HD.

Denial and Avoidance

Denial and avoidance are common themes in death and dying. While denial can be a very effective coping skill, it must eventually be replaced with understanding and acceptance. Denial can be seen in patients, families, and caregivers. It is important to note that "lack of awareness," or anosagnosia, is common in persons with HD as a direct neurological result of the disease process. A person with anosagnosia may be unable to understand that they have HD, no matter how much evidence is presented to them. Curiously, there can be degrees of unawareness, so that a person who categorically says, "I don't have HD," may still be willing to discuss medication for treatment of their chorea, or the importance of using food textures that reduce the tendency to choke.

Denial may also be a strategy that families use to "normalize" the situation. By saying, "your father is just clumsy, like his brother," such families may minimize the changes that have occurred due to the progression of the disease, and are surprised or unprepared as the person moves to the late or terminal stages of the disease.

CASE STUDY

Sandra remembered the lack of information about her grandmother with HD who died alone and found comfort and relief from the hospice staff as she learned that her mother's death would be comfortable and peaceful.

Biological children caring for an affected parent are themselves at a 50% risk of inheriting the gene that causes HD. They may not want to see what their future might hold, or may choose not to see the person in an advanced state of HD because of their own denial. Children may be adjusting to their own feelings of fear, denial, anger, and depression and require more counseling, especially if the diagnosis has not been shared with them until late in the disease.

Some families use denial and avoidance in an attempt to protect their children. Parents report that their greatest fear is that their children are at risk for the disease. Parents may limit their discussions about HD with their children due to their own emotions and fears. Avoidance can also prevent a person with HD or family from reaching out to community resources for either hands-on or emotional support. This can make it difficult for a family to request hospice services.

Family Dynamics in the Terminal Stage of HD

It is normal during the course of a terminal illness for families to struggle with structure as they mourn and cope. Even a cohesive family may become disorganized or chaotic as it struggles with the dying of an elder, a sibling, or a child. As symptoms tend to occur at the height of a person's career, it causes financial strains on the family as the affected person becomes unable to work.

CASE STUDY

Richard was the primary wage earner and insurance carrier for his family prior to his diagnosis of HD. When he was no longer able to work the family found themselves in financial strain. Even with his disability payments, more care was needed and his spouse decided to stop working to provide his needed extra care.



This in turn can lead to changes in the roles of family members, as a spouse or teenaged child may become a caregiver for the HD person. Most HD families suffer from considerable financial and emotional strain as a result of HD. Hospice can assist the family with their grief due to the changes of a terminal disease.

About 50% of people at-risk for HD have some signs of psychiatric distress, such as anxiety and depression. Children are particularly vulnerable. Teenaged children have a growing awareness of their risk of developing the disease from which the affected parent is dying. If the parent was abusive, violent, aggressive, or neglectful in the earlier stages of the disease, the child may have fears of being like that as they grow older, or may be ambivalent about the parent's coming death. Hospice staff can be very helpful by providing education, grief counseling and support to the family, and age-appropriate information to the children.

It is important for a child's well-being and understanding to be involved in the family structure with open communication, and children should be told the truth. However, the child's developmental stage needs to be taken into account when addressing questions and presenting information. It is important to assess how the children are managing their own grief, and to provide emotional support or refer them for more intensive counseling, support, or treatment if necessary.

CASE STUDY

Rachel remembered stories about her grandmother who died of HD complications in her 60's. She had fond memories of her grandmother before the symptoms started. Watching her mother become more symptomatic has become very difficult for her. She is reminded of her grandmother and realizes her mother's fate. She was offered some counseling to understand the complicated grief from her grandmother's death that is making it difficult to accept and deal with her mother's increasing decline.

Guilt is a common theme in hospice, and perhaps more so in the HD family. The affected individual, aware of his own diagnosis, may feel that they are "responsible" for passing the gene on to future generations. The person may feel guilty for being unable to support the family or care for the children, for needing care, for dying at a young age. Spouses or other family caregivers may feel guilty for being unable to manage their loved one at home, or for asking for help. Education and counseling can be useful for the emotional well-being of both the person and the caregiver.

Grief and Mourning

As a neurodegenerative disease, HD leads to progressive losses over the years of its progression. Affected individuals and their families experience a loss of physical and cognitive abilities, loss of family roles, loss of income, loss of personality, loss of intimacy, and finally at the end, have to face loss of life. The multigenerational nature of the disease also means that there may be several affected individuals within an extended family who are at different stages of the disease simultaneously (for instance, an affected parent and child, or two affected siblings), complicating the situation further.

The term "disenfranchised grief," is grief that is not publicly recognized. The losses that accompany HD are gradual and there is no established social structure for the mourning of losses that continue for long periods of time. Hospice staff, by exploring these issues, can help persons with HD and families to work through the grief process. Hospice also allows for continued bereavement work with the family for up to 13 months after the death of the person.



Acceptance, planning, and emotional support were noted in one study as being the most helpful and commonly used coping strategies for HD families. The hospice team can facilitate the movement from a setting of denial and avoidance, to a family that feels supported, has made appropriate short-term and long-term plans for end-of-life care and for life after the death of the affected individual, and has an acceptance of the reality of the situation.

CASE STUDY

Hospice staff members worked well with the Thompson family to address each person's individual needs as Mr. Thompson's impending death became evident. The family was told about changes in Mr. Thompson's condition that suggested the time of death would likely be soon. Mrs. Thompson worked with the chaplain to discuss spiritual issues and the teenage children worked with the social worker to talk about how to talk about their father's death with their friends. Plans were made to have continued contact with the family after Mr. Thompson's death.

What Hospice Provides

Once the person has engaged in hospice care, the hospice team can support the person and family in many ways. We focused above on the special psychosocial stressors and situations that can exist in

HD families. Hospice providers can be of great help to these strained families by providing education, support, and continuity during this difficult stage of life. Helping a family to participate in a comfortable and peaceful death can go a long way to reversing generations of fear and suffering, and allow future generations to face their own mortality with much more confidence.



Music therapy, animal-assisted therapy, massage, aromatherapy, and other support activities can be as helpful for persons with HD as they are for other hospice patients. Involuntary movements and rigidity/dystonia can lead to discomfort, and therapies that relax muscles can be soothing to the person.

Comfort measures might also include the placement of an internal bladder catheter to relieve anxiety about incontinence, medications for constipation, comfortable clothing and padding of the walls, floor, or chairs to reduce the possibility of bruising due to involuntary movements. Joint cushions or pads and frequent turning can help to avoid pressure ulcers.

The hospice or palliative care physician may be at a handicap trying to address the needs of an HD person who has recently moved to hospice care, because he is new to the person. The family can help the physician to know what their concerns are, what medications the person has used successfully in the past, and their care priorities. Medications are most often used in HD to treat chorea (neuroleptics, tetrabenazine, benzodiazepines), and behavioral and mood disturbances (antidepressants, anti-anxiety agents, mood stabilizers, antipsychotics). There is no specific medication that should be used, or avoided, in the terminal stage of HD. It is important to not abruptly discontinue medications in a person with HD, because of the potential for rebound symptoms, although it is often possible to taper down, stop, or switch from one medication to another, if the changes are made gradually and thoughtfully. There is no specific literature on medication use in hospice for persons with HD; our experience, however, is that agitation can sometimes be reduced with opiates (e.g. hydromorphone, fentanyl, morphine) or barbiturates (e.g. phenobarbital). A discussion between the hospice physician and the neurologist or psychiatrist who has treated the person in the past might reassure the family that there is continuity of care, and also provide the new physician with useful insights about the person and his past response to medications, and the family.



It is also important to offer grief counseling to staff members who may have cared for the person for many years. They too feel the loss, like family members, because of their long connection to the person.

Brain Donation

Research continues around the world to find better treatments for HD. A person with HD may wish to make a unique contribution to HD research in the form of a brain donation for research at the time of death. It is important to discuss brain donation in advance, so that the process moves smoothly at the time of death, and there are no unexpected expenses. An excellent resource for HD brain donation is the Harvard Brain Tissue Resource Center, which accepts brain donations from around the country, and provides samples of their brains to carefully selected researchers around the world. In some locations,

there may be a local or regional group or researcher accepting brain material. It is vitally important to contact the Brain Bank or the local researcher as soon as possible, long before the time of death, to learn more about the process, to sign consent forms and complete any other necessary paperwork, to understand what costs (if any) will be incurred with brain donation, and to ensure that everyone in contact with the person understands the correct procedures to use at the time of death. For more information, contact the Brain Bank at (800) Brain Bank (272-4622) or www.brainbank.mclean.org. Brain tissue needs to be obtained within 12-24 hours of the person's death to be of the most use.

Remember, there is never "nothing I can do" for a person with HD!

Things to Remember

- Hospice care is appropriate in the terminal stages of HD.
- Typical features of the terminal stages of HD include a person who is unable to walk or talk, and who has shown signs of medical decline, such as weight loss, recurrent hospitalizations, or a serious hospitalization; or who has developed terminal agitation.
- The genetic nature of HD, and its relatively young onset age, produce unique psychosocial stressors within the HD family.
- Hospice care allows the person with HD, and the family, to experience death as an expected, comfortable, and peaceful outcome of the disease.
- Communication between the hospice team and the prior treating physician creates continuity of care, and allows the new team to know as much as possible about the person and family they will be caring for.



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Appendix

Huntington's Disease Society of America

EXAMINATION GUIDELINES FOR
UNIFIED HUNTINGTON'S DISEASE RATING SCALE '99 (UHDRS '99)

MOTOR ASSESSMENT

#1 OCULAR PURSUIT – Ocular pursuit should be assessed over a range of approximately 20° with a target passing slowly at < 10° per second, which corresponds to about 2 seconds for moving an object from one shoulder to the other.

#2-3 SACCADE INITIATION AND VELOCITY – Saccade initiation should be tested over a 20° range, as for ocular pursuits. Saccade movement should be elicited by a sound (snapping fingers) or movement (wiggle fingers), but not by a verbal command to look to the right or left. Saccade velocity should be tested at a larger range of approximately 30° so as to be able to detect incomplete range.

#4-5 DYSARTHRIA AND TONGUE PROTRUSION - Self-explanatory

#6 FINGER TAPS – Subject taps thumb with index finger in rapid succession with widest amplitude possible, each hand separately.

#7 PRONATE/SUPINATE HANDS – This task requires the subject to alternately hit the palmar and dorsal surface of one hand against the palm of the opposite hand. Use the palm of the opposite hand as a target instead of some other surface such as the subject's leg or the table surface. The subject should do this task as quickly as possible over a five-second interval. The task is graded according to the degree of slowing and irregularity.

#8 LURIA – FIST-HAND-PALM SEQUENCING – Say "Can you do this?" Examiner puts hand into fist on flat surface (or in lap) and sequences as follows: fist, side, flat (DO NOT REPEAT THIS OUT LOUD). Watch to make sure that subject can mimic each step. Continue to practice Luria 3-step for 1-2 minutes. When subject is able to join you then say "Very good, now keep going, I am going to stop." Rest hand and start timing subject's sequences. A sequence is considered correct only if it is unaided by examiner model and in the correct order. Count completed sequences and score. If subject was unable to complete any sequences over a 10-second period, then continue as follows. Say "Now let's try it again. Put your hands like this. FIST, SIDE, FLAT." Watch to make sure the subject can mimic each step. Using the verbal labels, begin the sequences again and ask the subject to "Do as I do, Fist, Side, Flat" (repeat this as you continue). Continue to perform Luria 3-step. When subject is able to join you say "Very good, now keep going, I am going to stop." Rest hand and start timing subject's sequences. A sequence is considered correct if it is unaided by examiner model and in the correct order. Count completed sequences and score as above.

#9 RIGIDITY-ARMS – Rigidity is judged on passive movement of the arms with the subject relaxed in the sitting position.

#10 BODY BRADYKINESIA – Observe the subject during spontaneous motion such as walking, sitting down, arising from a chair, and executing the tasks required during the examination. This rating reflects the examiner's overall impression of bradykinesia.

#11-12 MAXIMAL DYSTONIA (TENDENCY TOWARD A POSTURE, POSTURING ALONG AN AXIS) AND MAXIMAL CHOREA (MOVEMENT) – Observe the subject during the examination; i.e., no particular maneuvers are required to illicit these features. Maximal dystonia and chorea are typically observed during demanding motor tasks such as tandem gait. Both dystonia and chorea are rated by specific regions. "BOL" refers to buccal-oral-lingual. Facial dystonia includes blepharospasm, jaw opening and closing. When rating dystonia (question #11) BOL and facial dystonia should be included in your assessment of the truncal region.

#13 GAIT – Observe the subject walking approximately ten yards as briskly as they can, then turning and returning to the starting point.

#14 TANDEM GAIT – The subject is requested to walk ten steps in a straight line with the foot placed (accurately but not quickly) such that the heel touches the toe of the other foot. Deviations from a straight line are counted.

#15 RETROPULSION PULL TEST – The subject's response to a sudden posterior displacement produced by a pull on the shoulder while the subject is standing with eyes open and feet slightly apart is assessed. The shoulder pull test must be done with a quick firm tug after warning the subject. The test may be repeated if the subject did not have sufficient warning or did not understand the test. The subject should be relaxed with feet apart and should not be learning forward. If the examiner feels pressure against his/her hands when placed on the subject's shoulders, the examiner should instruct the subject to stand up straight and not lean forward. The examiner should instruct the subject to take a step backward to avoid falling.

Examiners must catch subjects who begin to fall. To prevent either individual from falling to the floor, examiners should brace themselves with one foot back and/or stand between subject and a wall.

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However, adequate room is needed to test retropulsion and recovery. Subjects should be told that taking one step backwards is acceptable.

#16 WEIGHT - Self-explanatory.

#17 DIAGNOSTIC CONFIDENCE LEVEL

0 = Normal (no abnormalities)
1 = non-specific motor abnormalities (less than 50% confidence)
2 = motor abnormalities that *may* be signs of HD (50 - 89% confidence)
3 = motor abnormalities that are *likely* signs of HD (90 - 98% confidence)
4 = motor abnormalities that are *unequivocal* signs of HD (> 99% confidence)

The diagnosis of HD is based on the unequivocal presence of an otherwise unexplained extrapyramidal movement disorder (e.g., chorea, dystonia, bradykinesia, rigidity) in a subject at risk for HD.

The grade assigned by the investigator represents a level of confidence for the diagnosis of HD in a particular subject. Grade 1 represents a < 50% confidence level for a particular subject who may have non-specific motor abnormalities. Such abnormalities could include mild clumsiness or slowness that might be normal findings, or non-specific changes such as distal weakness. Grade 2 implies a 50 - 89% confidence level and should be assigned to a subject with suggestive but not definitive clinical findings. Such findings could include mild slowness and clumsiness with minimal non-specific oculomotor abnormalities. Grade 3 should be assigned to a subject that has motor abnormalities that are likely signs of HD (90 - 98% confidence). Such abnormalities could include intermittent movements that could represent chorea in the setting of mild motor slowing. Grade 4 should be assigned only to a subject with an unequivocal extrapyramidal movement disorder in the presence of a confirming family history or known positive gene test, when the examiner is ≥ 99% confident (only errs 1 in a 100 such instances) that the subject has HD. Such findings would include the presence of definite chorea or dystonia, usually with accompanying motor slowing.

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ISBN 978-0-9903220-1-6

Huntington's Disease Society of America

505 8th Avenue, Suite 902 NY, NY 10018
1-800-345-HDSA • www.hdsa.org