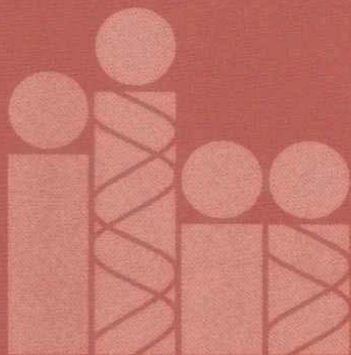


Huntington's Disease Society of America

Understanding Behavior in Huntington's Disease

Written and Edited by
Jane S. Paulsen, Ph.D.

Second Edition





Understanding Behavior in Huntington's Disease

*A practical guide for individuals,
families, and professionals
coping with HD*

Second Edition

by **Jane S. Paulsen, Ph.D.**

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The lay reader should consult a physician or other appropriate health care professional concerning any advice, product, treatment or therapy set forth in this book.

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Most of what I know about behavior in Huntington's Disease (HD) was taught to me by the HD individuals and families with whom I have worked. They have struggled with me to find solutions to their problems. It is with gratitude to this "HD family" that I pass this information along to others.

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This book is dedicated to John B. "Jack" Penney, a master and student of HD, whose compassion for sufferers of HD was exemplary, as was his quest to understand the basal ganglia.

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Preface

This book was written for the caregivers and people affected by Huntington's Disease (HD). Caregivers may include family members (partners, children, parents, and siblings), friends, and direct care staff (nurses, nursing assistants, physical and occupational therapists, speech therapists, and social workers). The materials will also be useful to other professionals, such as neurologists, general practitioners, psychologists and psychiatrists, who provide advice to persons with HD and their caregivers. This book was intended to help better understand the various causes of behaviors in HD and to provide strategies to cope with difficult behaviors.

People used to argue about whether behavior was "caused" by genes, diet, weather, politics, child-rearing practices, or a variety of other potential influences. Today we find it more useful to think of multiple causes of behavior, including past experiences, current health, and expectations for the future. Over the past several years, scientists have discovered many ways in which the brain contributes to behavior. Many of the behavior patterns observed in persons with HD are similar because the same part of the brain is affected in everyone with HD. Throughout this book I have addressed some of the major behavior patterns that we see in persons with HD.

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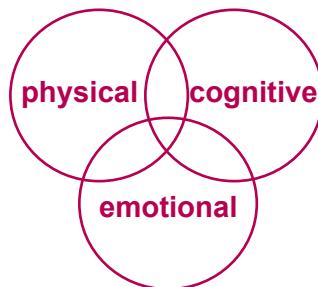
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■ *Difficult Behaviors in Huntington's Disease*

HUNTINGTON'S DISEASE: AN OVERVIEW

Huntington's Disease (HD) is an inherited brain disorder that causes progressive deterioration of the physical, cognitive and emotional self. It leads to severe incapacitation and eventual death 10–40 years after the onset of the disease. Although it usually affects adults between the ages of 30 and 45, symptoms can appear in young children and older adults. Common symptoms are uncontrollable movements, abnormal balance when walking, slurred speech, difficulty swallowing, thinking difficulties and personality changes. Each child of an affected parent has a 50% chance of inheriting the HD gene, which is located on chromosome four. There is no cure and no effective treatment, but scientists are exploring possible treatments and caregivers are developing new approaches to care.



DIFFICULT BEHAVIORS IN HUNTINGTON'S DISEASE

There are a number of behavior problems that can accompany HD. These behaviors may include apathy, agitation, rigid thinking, denial, depression, disorganization, paranoia and forgetfulness. Specific behaviors vary significantly from person to person and can change throughout the progression of the disease.

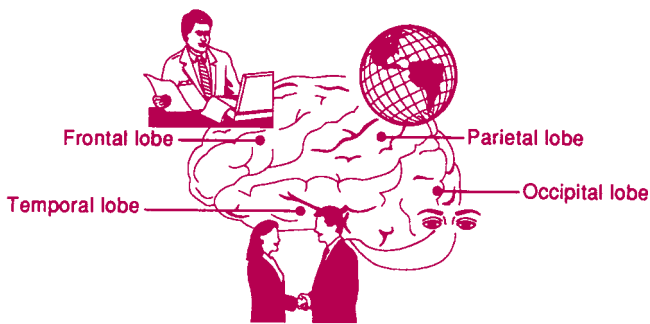
There are many reasons why a difficult behavior may be occurring. Typically, the behavior is related to changes taking place in the brain. In addition, the environment, including people, events and/or health issues, can contribute to the behavior being expressed.

Often, it is helpful for caregivers to try to understand why the person with HD is acting in a particular way. If caregivers can determine what might be causing or triggering the unwanted behavior, it becomes easier to cope with the behavior and, sometimes, to prevent the difficult behavior from recurring. Listed below are some possible reasons for behavioral difficulties in HD. A specific behavior may be the result of one or all of these reasons.

HOW DOES THE BRAIN CONTRIBUTE TO BEHAVIOR CHANGES IN HD?

UNDERSTANDING ANATOMY AND RELATED BEHAVIORS

Different parts of the brain are responsible for different aspects of our behavior. Below is a picture of a brain showing some of these different general responsibilities.



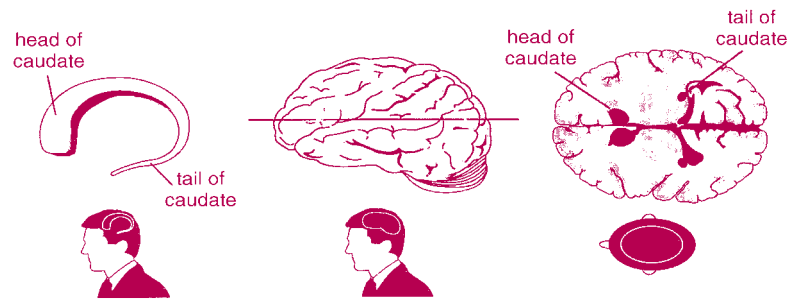
For instance, the occipital lobes contain the cells which help us to see. These cells tell us the shape, color and movement of things we see. The neighboring parietal lobes tell us the spatial location of things that we see, hear or touch, just like a map tells us the location of countries, cities and roads. The temporal lobes are very important for our memory, containing cells that help us

remember a face, a name or an event that has taken place. The frontal lobes make up the largest area of the brain and contain the highest levels of our behaviors. The frontal lobe area of the brain is often considered the “boss,” or chief executive officer (CEO), of the rest of the brain. The following list includes executive functions of the frontal lobes.

- Organizing
- Establishing the priority of various events
- Controlling impulses
- Monitoring self-awareness
- Beginning and ending activities
- Creative thinking
- Problem-solving

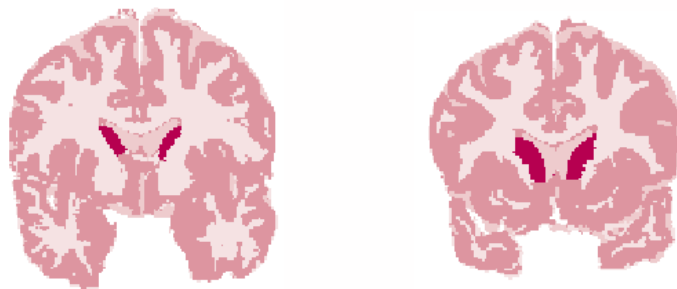
Although the effects of the HD gene are not fully understood, the primary site of known pathology is the caudate nucleus in the brain, where the greatest number of brain cells die because of HD. We cannot see the caudate from our picture of the brain above because it is located deep within the brain. Let’s take a closer look

at the caudate and its importance in the brain by studying the pictures shown below. The figure on the left shows the shape of the caudate, which is located under the brain's surface and runs through the entire brain. The middle figure shows how the brain would be sliced to reveal the caudate. The figure on the right shows the caudate.

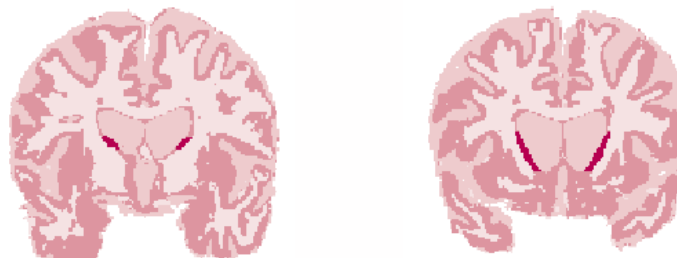


When doctors take a picture of the brain to “see” the effects of HD, they typically look at pictures like the ones below. The first row of diagrams shows a magnetic resonance image (MRI) scan of the brain of a person with no disease and the second row contains MRI scans of the brain of a person who has HD. The brain tissue colored dark black in the scans represent the caudate. You will notice that the caudate is significantly reduced by the disease: the reduced black sections in the brain pictures on the bottom indicate a great deal of atrophy or brain cell death. We are just beginning to understand why so many behaviors are affected when the caudate is damaged.

Black represents the caudate



MRI scan of an individual with no brain disease



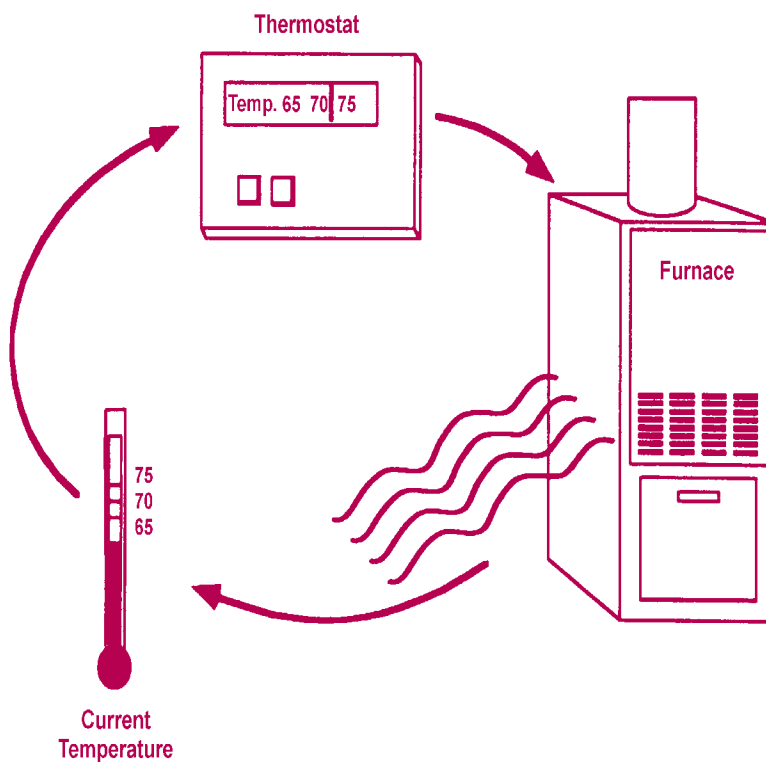
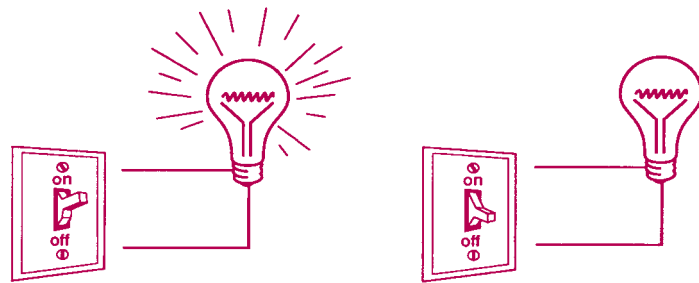
MRI scan of an individual in the early stages of HD

UNDERSTANDING BRAIN COMMUNICATION AND CIRCUITRY

The caudate is a very important structure because it has multiple connections with, or circuits to, other parts of the brain. A circuit is a pathway by which information travels in the brain. These circuits run through the caudate carrying information from one part of the brain to another. Brain circuits operate much like other circuits that we are familiar with in our daily lives. For example, a light bulb can fill a room with light with the flick of a switch that allows the electrical current to travel through the circuit. The light bulb cannot give off light if the circuit is disconnected or blocked in any way.

The brain works in a similar manner such that behavior occurs without difficulty when the circuits are operating properly. As the caudate deteriorates, however, some circuits may “flicker” on and off and other circuits will cease working, with the result

that the messages just don’t get through from one part of the brain to the other. Consider the circuits operating in your home. The circuits that operate your lights are “one-way” circuits, light switch to light bulb. There are also circuits that allow two-way communication, or feedback, to occur. A good example of a two-way circuit is the heating and cooling system in your home. A thermostat is used to set the desired temperature in your home. The furnace turns on and off to heat the home. The room temperature is recorded on a thermometer, which is read by the thermostat. When the desired room temperature and the current room

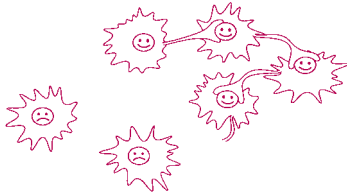


temperature are consistent with one another, no heating is required. When the room temperature is lower than the desired temperature, however, the heating system turns on. This circuit, as pictured on the left, requires that the heating system (the furnace) receive information from the thermostat to complete the circuit and produce the desired behavior (heating).

A brain circuit is made of neurons that communicate with one another when they fire or “talk.” Neurons, which are microscopic, look like this:



They “talk” via chemical substances called neurotransmitters to get information transferred in the brain like this:

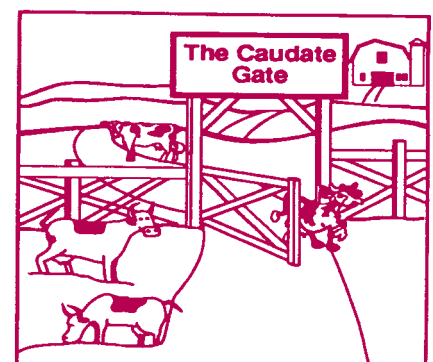
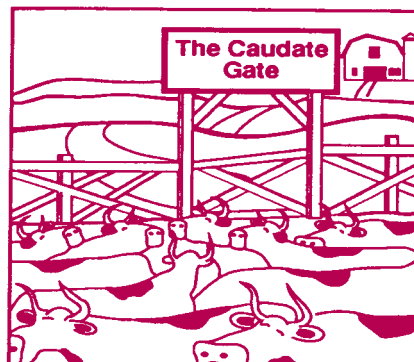
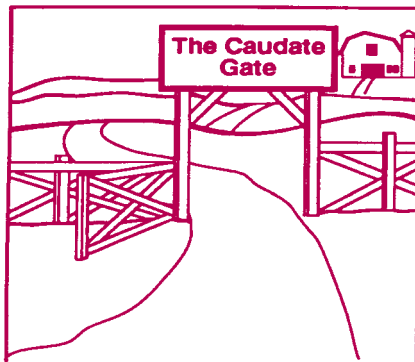


UNDERSTANDING THE ROLE OF THE CAUDATE IN THE BRAIN: THE CAUDATE GATE

Given the numerous connections of the caudate with other areas of the brain, it has a great deal of impact on behaviors. The caudate is one of many “gates” in the brain. Gates are used to regulate, organize, and filter information. The caudate has been found to be a very important brain gate because it has the most connections with the frontal lobes and it influences mood and cognition as well as motor skill. In general, the caudate regulates all information that passes through it, including movements, thoughts and feelings. When information that travels along the circuit arrives at the caudate, the caudate’s job is to “regulate” or “gate” the information before sending it along to the next station.

For many of our day-to-day behaviors, regulation is critical in determining whether the behavior will become a problem. For instance, a little anger (often called annoyance) is acceptable, whereas a temper outburst is a problem. A little concern (often called worry) is normal, whereas non-stop repetition of a concern (often called obsession) can become a problem.

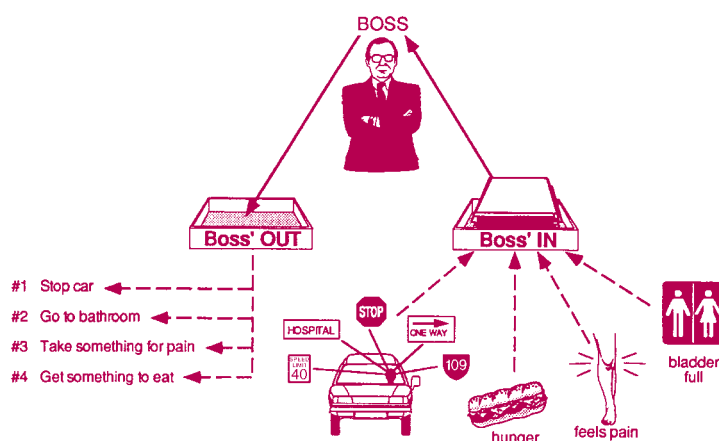
Being from Iowa, I think of the caudate gate much like I think of the gates on our farm. The gate was our method of controlling the cows. For example, when it was time to milk at the end of the day, the gate was opened wide, allowing all of the cattle to cross the road and come into the barn. When it was time to wean the calves from their mothers, the gate was opened and closed quickly, allowing one calf at a time to come through the gate into the neighboring paddock. At Fair time, we only wanted the prize heifer out, so we opened the gate only once to lead the winning heifer to the Fair. The caudate is necessary to regulate information in the



brain much like a gate is used to control, or regulate, our livestock. If the gate breaks down, the cows cannot be controlled. They may get out and get into the corn, they may not come into the barn at milking time, or we would be unable to wean calves from their mothers. When the caudate breaks down, the person with HD cannot control the feelings, thoughts or movements that are experienced or expressed.

The Caudate Gate and the Boss

An additional function of the caudate gate is to assist with the organization of information that travels to and from the “boss of the brain,” or the frontal lobes. One of the primary functions of the frontal lobes is to organize or prioritize the transfer of information from one area of the brain to another. For instance, there may be several information bits travelling through the brain on their way to the boss to make a specific request. The pain center may be communicating a tension headache and requesting Tylenol. The hunger center may be communicating hunger and requesting a stop at McDonald’s. The bladder may be full and signalling for a pit stop. The visual association cortex may be scanning some road signs and decoding language into meaningful spatial locations. The olfactory cortex may be detecting some mild body odor and requesting that the window be opened for some fresh air. Obviously, all of these requests cannot be met at once. The information needs to be organized and prioritized. The caudate is thought to assist with the transfer of information to and from the frontal lobes. A problem arises when certain circuits aren’t functioning because neurons or brain cells have died, interrupting the flow of messages through the brain.



This organization of information may be similar to the sorting of letters as they come into and out of the boss’s office. It is possible that the connections through the caudate require similar organization as does the mail. For instance, the caudate needs to determine which request to send up to the boss for action immediately, and which requests can wait until later. Using the example above, the caudate would need to prioritize the full bladder and send the request up to the boss to find a restroom. Next the caudate would need to decide whether the pain request or the hunger request were more important, as well as continue to organize the ongoing interpretation of street signs so that driving can occur safely. With the progression of HD and increasing brain cell death, it becomes increasingly difficult for the brain to carry out its functions to regulate and organize. As circuits are damaged, the brain can’t transfer information as effectively as before.

HOW CAN THE ENVIRONMENT CONTRIBUTE TO BEHAVIOR DIFFICULTIES IN HD?

ROUTINE

Since the caudate is unable to regulate properly (in other words, the gate needs oil), people with HD need a regulated environment that provides daily structure and routine upon which they can depend. Because the caudate cannot get the information from the frontal lobes to decide what is the most important thing to do next or how to order the day's activities, a person with HD relies on external sources to structure the day and make decisions.

DISTRACTIONS

When the caudate cannot regulate what is travelling along the brain circuitry, it becomes very easy for the circuits to become overloaded. For instance, if the television and radio are on, the roommate is talking on the telephone and a friend comes in and asks whether the person would like to go outside, the person may respond with frustration or anger. In this case, the circuits are overloaded and the caudate is unable to decide what is important and what is not. Unfortunately, the friend may not realize that this environment is too busy—too stimulating—



and may decide that the person is a grump. A particularly important time to minimize distractions to diminish “circuit overload” is during mealtime. Swallowing difficulties due to impaired muscle control and functioning create the need for concentration when eating meals. Distractions while eating can increase the chances of choking and inhaling food or liquid into the lungs.

Consider the family who complained that they couldn't get along any more. She reported that he never listened and couldn't be relied upon to “do simple household chores like going to the store.” He stated that she lost her temper when he forgot one item in a list he considered to be too long. After some discussion it became evident that the caudate gate was not working properly. As he tried to listen to the store list, several things (such as background noises) competed with his wife's voice for his limited attention. He described it as “a wide-open gate—with everything rushing in at once.” When distractions were minimized and requests were written in short lists, this family got along much better.

HOW CAN INDIVIDUAL FACTORS CONTRIBUTE TO BEHAVIOR DIFFICULTIES IN HD?

UNIQUE PERSONALITY

Although sometimes it might seem that it would be easier if all people with HD were exactly the same, having HD fortunately does not take away individuality.

We Are All Unique: Joan's Story

Alice, a staff member in a nursing home, was rearranging the night table of Joan, a woman with Huntington's disease. Joan had been busy in the adjoining bathroom while Alice tidied up her room and threw away some wilted flowers on the night table. Suddenly, without warning, Joan leaped out of the bathroom and wrestled Alice to the floor. Joan attacked her with such force that Alice was sent to the hospital. The staff at the nursing home agreed that this was a product of the irritability and aggression of Huntington's disease.

Unfortunately, this staff's interpretation of the aggressive outburst is not uncommon. The disease is not the person, however. All individuals have a history and a life course. They have personal experiences and objectives and it is important to pay attention to how the disease has interfered with the individual's life. It turned out that the bouquet of wilted flowers was the only gift Joan had received from her son that year. Their removal by Alice was the source of her anger. Her response was certainly exaggerated, but the nursing staff viewed her behavior quite differently knowing the full story. "When we see things in a new way, we can intervene in a different way," says Dr. Allen Rubin, who shared this story.

When an individual is diagnosed with HD, it is important to remember that there is still a person, an individual, behind the HD. Oftentimes, the diagnosis of HD is all the caregiver pays attention to, and the personality of the individual is neglected.

STAGE OF ILLNESS

The type, frequency, and severity of behavioral problems that occur often vary with stage of illness. One survey suggests that persons with early HD are more concerned about depression, anxiety, and apathy, whereas persons with later-stage HD report agitation, irritability, and disinhibition as the most prominent behavioral concerns. The ways of responding to the various stages of HD, however, can vary greatly from person to person. It is usually helpful to consider what stage of illness the individual is in to better understand the behaviors that occur.

Based upon the rating of Total Functional Capacity (TFC; Shoulson, 1981), patients are considered to be in one of five stages of the HD illness. In general, descriptions of the stages are as follows:

Stage I: (0–8 years from illness onset) Maintains only marginal engagement in occupation having part-time voluntary or salaried employment potential, *and* maintains typical pre-disease levels of independence in all other basic functions, such as financial management, domestic responsibilities, and activities of daily living (eating, dressing, bathing, etc.); *or* performs satisfactorily in typical salaried employment (perhaps at a lower level) and requires slight assistance in only one basic function: finances, domestic chores, or activities of daily living.

Stage II: (3–13 years from illness onset) Typically unable to work but requires only slight assistance in all basic functions: finances, domestic, daily activities; *or* unable to work and requires major assistance in one basic function with only slight assistance needed in one other; at least one basic function is handled independently.

Stage III: (5–16 years from illness onset) Totally unable to engage in employment and requires major assistance in most basic functions: financial affairs, domestic responsibilities, and activities of daily living.

Stage IV: (9–21 years from illness onset) Requires major assistance in financial affairs, domestic responsibilities, and most activities of daily living. For instance, comprehension of the nature and purpose of procedures may be intact, but major assistance is required to act on them. Care may be provided at home but needs may be better provided for at an extended care facility.

Stage V: (11–26 years from illness onset) Requires major assistance in financial affairs, domestic responsibilities, and all activities of daily living. Full-time skilled nursing care is required.

These stages are very broad guidelines and individuals can vary a great deal in the course of the illness. For instance, one patient has had chorea for 15 years yet he is still employed (in a reduced capacity) at his regular job.

Angry at Albert ... And HD

“At first I thought it was just his imagination. He complained that he wasn’t able to work as effectively as he used to. When he “quit” his job of 20 years I was concerned, but he assured me that his new job was a “good opportunity.” Only after losing this job, too, did I realize that my high-level executive husband was no longer able to function in his regular capacity. He tried several jobs over the next few years –just to keep busy and to keep money coming in. Finally, he became too frustrated to perform the way he wanted. I decided that he could take care of the kids and the house if he was going to stay home. He stayed home but was constantly pacing the house and became agitated at the drop of a hat. There was too little money and the home front was falling apart. I was angry with him for losing jobs and not helping me out at home. He just didn’t look that sick, so I couldn’t figure out why he was being so impossible. I wanted Albert to make money, be a parent, and be a husband for me. I was angry that all of this was gone. We went through five miserable years before I could begin to accept that HD was part of our family. I yelled a lot and I cried a lot. Even now, just when I start to figure things out, everything changes and I have to start over. It’s a long process of accepting and coping, over and over again.”

Loss

Thinking about HD in terms of stages emphasizes the losses that occur as the disease progresses. Over time, individuals with HD lose their capacity to work and to function independently. Each loss is a death of sorts, and the natural reaction to loss is to grieve. Coping with loss is one of the greatest challenges for HD persons and families. The author Elisabeth Kubler-Ross has suggested that individuals progress through stages of loss: denial, anger, bargaining, depression, acceptance. Experience tells us that everyone expresses these stages of loss in varying degrees and at varying times. Illnesses that keep changing, like HD, can bring grieving and re-grieving.

Losing the things you depend on is scary—whether the things are your partner’s abilities or your own. Coping with loss is a big job for individuals with HD as well as for their family and friends. In the case described on the previous page, we hear a wife describe her own methods of responding to her husband’s loss of work capacity. She experiences denial (considers it his imagination—he looks just fine), bargaining (if he stays home, he takes care of the kids and house), anger, and depression. When she finally has acceptance, she finds that the situation has changed and the new situation requires more grieving, anger, depression, denial, and coping. When HD strikes, we are faced with multiple losses: the loss of a healthy loved one, the loss of income, the loss of control, the loss of our own independence and the loss of plans for the future.

SAFETY AND SELF-ESTEEM

There are two primary issues that need to be addressed in managing the decline associated with advancing HD. First, what activities can the HD person continue to pursue in a safe manner? Second, how can we maintain safety and protect an individual’s sense of self-esteem? There are several complex tasks that could be dangerous as an individual loses control over his or her emotions, motor skills, and judgement. Unfortunately, many of these activities may mean a great deal to a person. Perhaps the activity was important in their role in the family (e.g., Mom’s great cooking), or perhaps the activity helped define their self-esteem (such as Dad’s driving to community events). The challenge is to assure safety at all times while maintaining respect for the individual’s personality.

Before an activity is no longer safe for an individual to perform, the activity should be modified (instead of totally forbidden) in preparation for its total restriction. For instance, driving is often an activity that needs to be restricted when an individual is impaired by consequences of a disease. If driving was very important to an individual (as it often is), a decision may first involve limiting driving to surface streets within one mile of the home, excluding freeway driving, downtown driving and high-speed driving. Alternatively, driving may be restricted to one specific route (e.g., to the doughnut shop for morning coffee). Eventually, however, even modified driving becomes unsafe. When an activity is no longer safe in any form, plan its restriction in conjunction with an exaggeration of other activities that help define the individual’s esteem. For instance, one family formally restricted driving immediately prior to a family vacation but suggested their father highlight the course on the map. In this manner, it was clearly stated that he would not be driving but that his input was a valuable contribution to the family.

In general, maintenance of an individual's personality is critical to coping with loss due to HD. Often people worry that if they get HD, they will lose their individuality and be just like their affected parent. Whether or not this is a rational fear, efforts can be made to assure people with HD that their unique aspects as a person are appreciated, even though they have HD and may no longer be able to participate directly in favorite pastimes. What activities does the individual most enjoy? Is the person a sports fan? An opera or theatre buff? A rap enthusiast? A heavy metal fanatic? Country-western star wanna-be? Tape players can help provide much enjoyment by maintaining favorites. Books on tape can be enjoyed when eye tracking makes reading difficult. Videos can provide humor, ball games and old classics. What colors does the individual most enjoy? Wild, busy fabrics in oranges and reds, or solid, calm blues and purples? Colors, special decorations, and pictures from the kids and grandkids should all be used to maintain the identity of an individual who happens to have HD.

OTHER HEALTH FACTORS

ACUTE AND CHRONIC ILLNESS

Although HD can become the primary focus of attention, people with HD can have other medical problems. This is important to remember and consider when trying to understand behaviors. Pneumonia, fever, urinary infection, hemorrhoids, a cold or flu, gynecological problems, chronic illness such as arthritis and diabetes, and the natural aging process, including menopause, can lead to changes in behavior. It is not always easy to identify health problems in people with HD due to speech problems, word-finding problems and lack of self-awareness. Health conditions should be watched, and any sudden changes in behavior should be reported to a doctor.

FOOD REACTIONS

It is wise to be aware of the tendency of friends and relatives to suggest the need for herbs, special diets and so on. These suggestions should not be forbidden but generally they should be discussed first with the primary physician. Some herbs and foods (such as grapefruit juice) have major interactions with some medications.

DEHYDRATION

Persons with HD, in the mid to late stages particularly, may not recognize the sensation of thirst or be able to initiate getting a drink. As a result, they may become dehydrated. The symptoms of dehydration may include dizziness, confusion, refusal to drink, dry skin, fever, flushed appearance and a rapid pulse.

FATIGUE

Persons with HD often need to spend greater amounts of energy to perform regular activities of day-to-day life, to control emotions and to adjust to the continual changes in their capabilities. Many individuals require additional sleep to avoid fatigue that can contribute to increased behavioral difficulties. Plan daily routines to

include rest times, make sure that outings have some quiet time to relax and remember that when you're exhausted, nothing is fun.

PHYSICAL DISCOMFORT

As the disease progresses, physical discomfort may increase due to musculoskeletal shifts from walking and/or sitting in awkward positions. There is some evidence that HD disrupts the normal temperature regulation mechanisms of the brain. Adjustment of the thermostat may be helpful to limit physical discomfort for the individual with HD. Discomfort may also be caused if the person's immediate needs are not being met. For example, the person may be hungry, thirsty, too warm or need to use the bathroom.

UNDETECTED VISUAL OR HEARING IMPAIRMENTS

Unfortunately, it has been found that individuals with illnesses such as HD often have undetected vision and hearing impairments, the correction of which could greatly improve quality of life. Vision and hearing problems are not related to HD but are common problems overlooked in persons with any illness. Behavior problems often increase if the individual becomes frustrated. Basic vision and hearing problems should be checked and corrected so that these difficulties do not further limit the capacity of the individual with HD.

THE EFFECTS OF MEDICATIONS

Medications may be prescribed for certain symptoms of HD such as chorea, depression, aggression, and temper outbursts. Unfortunately, side effects can accompany medications; over-medicating is a risk as well. Drowsiness, decreased motivation, nausea, dizziness and depression can be side effects and should be reported to a doctor. Persons with HD often become unable to tolerate as much alcohol (or other sedative medications) as they could in the past. This is especially important for persons who are still driving.

■ Approaches to Solving Behavior Problems in HD

GUIDELINES TO PROBLEM SOLVING

Caring for and treating people with HD can be both challenging and rewarding. At times, the lack of an apparent remedy can be frustrating. We all must be creative in our search for solutions. Careful attention to the changing symptoms, as well as good communication between family members, professionals and the individual with HD, can contribute to the successful management of the disease. Listed below are some general guidelines to consider when a problem occurs.

STEP 1: *Identify the main problem.* Is the problem observable? Is it measurable? Can others see it? Avoid constructing a long list of problems—you will only be overwhelmed and unable to address any one behavior adequately.

STEP 2: *Gather information about the problem* and break the difficulty down into discrete components. Ask yourself:

- When does the problem occur?
- Where does the problem occur?
- What precedes the behavior?
- Who was involved?
- What follows the behavior?
- What emotion (fear, anger, frustration) was expressed?

Open your mind and develop new ways of seeing: with your eyes, your mind and your heart. Remember, the problem is occurring because of changes in the brain and the impact of HD. The patient with HD isn't deliberately trying to cause problems. Try to work out the problem with the person with HD. Together, you might be able to help solve it.

STEP 3: Review possible causes of behaviors including brain changes, environmental causes, individual contributions and health-related considerations. Evaluate any communication or triggers that might have caused the behavior.

STEP 4: Set a realistic goal—one that you have a chance of achieving. Develop a list of possible responses to the behavior and prioritize them. Try environmental changes and try to change your own behavior.

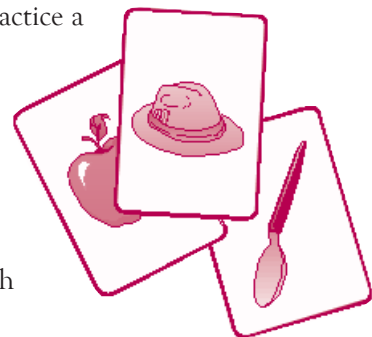
STEP 5: Be flexible and ready to try several strategies. Allow yourself several attempts and adequate time to use a trial and error process. Sometimes the best strategy may be time.

STEP 6: Reassure all individuals involved after a stressful event that you care and are trying to understand how to make things better. Don't carry the burden alone. Ask for help, advice, assistance, and support. HD is a multidisciplinary, multigenerational disease. Use every friend, family member, and professional to assist in making your life and the life of the patient with HD as pleasant as possible.

APPROACHES TO GOOD COMMUNICATION

SKILLS OF COMMUNICATING

- Be calm, gentle, matter-of-fact and relaxed.
- Use humor or gentle teasing. Convincing someone to get out of bed or go to the bathroom is usually easier if you can make a game or joke of it. Use touch to show that you care, even when your words don't show it or when they are not understood. Some people may shy away from contact, but most find a gentle touch reassuring.
- Start the communication socially. Trust is more easily established if you practice a balance of getting the task accomplished and chatting. Try spending a few minutes talking about the ball game, the school activity, family members, a movie, the family pet or even the weather.
- Use good eye contact and try to be at eye level. Keep your energy focused upon the individual.
- Keep the rate, pitch and volume of your speech steady and low. Lower pitch and conversational tone are easier to hear.
- If you are both getting frustrated, it may be a good idea to drop it and try again later. Sometimes a hug and a change of subject can make you both feel better. Other times you may need to leave the room and calm down.
- Never give up. Even when communication is difficult, set up a signalling system that will allow the person with HD to communicate nonverbally. Make flashcards of specific objects to which they can point.



LISTENING SKILLS

- Listen actively. Don't try to do two things at once—make listening a priority. If you do not understand something, apologize and ask the person to repeat it. Repeat back or rephrase what you hear so the person can know what parts you understand and what parts need to be repeated. Try to focus on one word or phrase that makes sense. Repeat this back to help clarify what is being said.
- Respond to the emotional tone of the communication. You may not understand what is being said, but you may recognize that it is being said angrily or sadly. Saying, “You sound very angry,” at least acknowledges the feelings, even if you cannot decipher the words.
- Remember that poor articulation is one of the primary problems caused by HD. The person is not doing this on purpose and is probably even more frustrated than you. Your calmness and patience will help create a caring atmosphere that will encourage the person to keep trying.
- Praise and encourage all efforts. A simple “thank you” or “you did a nice job” is good.

CREATE A MESSAGE CENTER

To reduce confusion and “he said, she said” disagreements, put routines, appointments and “to do” lists in writing. Establish a fairly large message/communication center in a visible, uncluttered place in the home or residence. Create the routine in a calendar format. Keep the information simple, easy to read and to the point, since eye movements may make reading difficult, the attention span may be short and a lot of written information may be visually overwhelming and confusing.

ENVIRONMENTAL CONCERNS

SIMPLIFY THE ENVIRONMENT

Keep the environment as simple as possible. Remove as many distractions as is necessary to calm the person. Try to limit the number of activities to one at a time. Work on keeping the number of individuals in the room to a minimum. In addition, avoid busy and noisy settings when possible. Remove furniture that is no longer useful. Lighting, visual contrasts between floors and walls, and the use of color can affect a person's behavior and level of functioning. For example, inadequate levels of light may affect a person's ability to concentrate when trying to eat. Patterned tile floors can look like steps, causing the person to trip or become uncertain. Avoid visual and auditory clutter!

ESTABLISH ROUTINES

Establish a regular weekly routine with scheduled activities (e.g., go to the movies on Monday, walk in the park on Wednesday, go shopping on Saturday) and routine assignments for household chores (e.g., change bed sheets on Tuesday, do the laundry on Thursday, water the plants on Saturday). The routine should have priority and should only be altered when unavoidable. A daily routine is equally important (e.g., awake at 7:00, breakfast at 8:00, morning show at 9:00, walk at 10:00).

PREPARE FOR CHANGES IN ROUTINE

Although routine is preferable for many people with HD, change is unavoidable at times. For example, some family members with HD need to move from an upstairs bedroom down to the ground floor to lessen the use of stairs and diminish falls. Expect this change to create some stress for them and try to make the transition as gentle as possible. Preparation is important: tell them well ahead of time and tell them more than once. This will help them make the mental adjustment before they must actually physically change rooms. Focus on the familiar and try to make the changes one at a time. For example, do not move the person and purchase new bedroom furniture at the same time. When moving to a nursing home make sure to bring photos, personal items and other objects that are familiar to the person. Of course, when at all possible, the person with HD should be involved in the entire process of selecting where he or she will live.

Moving to a Care Facility: Helen's Story

One family was confronted with resistance from their family member, Helen, when it came time to discuss the topic of placement in a skilled nursing facility. Helen's family was not confrontational with her. They simply shifted the discussions over a several-month period from "if you need to be placed" to "when you need to be placed" to "it's *getting to be the time* you will need to move into a facility" to "Where *you are going* to live is an important decision and I do not want to make it without you. I have found four places I want you to see." Placement was discussed at various times from the beginning of the disease, so the topic was not a surprise. Although there was some argument when the time came, Helen was given time to process the change as well as the power to choose the facility.

CARE FOR CAREGIVERS

The primary responsibilities of caregivers are twofold:

- 1) To advocate for the needs of and to provide care for the person affected with HD.
- 2) To advocate for the needs of and to provide care for oneself.

These two aspects of caregiving are equally important and essential.

Caregivers are individuals who assist with the care of a person or persons with HD. Caregivers can be family members, friends, professional staff or volunteers. Caring for someone with HD is physically and emotionally challenging. Paying attention to one's own needs and limitations is important for the health and happiness of both the caregiver and the HD-affected individual. When caregivers become ill, overloaded or depressed, they are unable to safely and effectively provide for the needs of the HD-affected individual. **For many of us, it is difficult to consider caring for oneself as a way of caring for someone else, but this is probably the most important aspect of caregiving.**

A RECIPE FOR CAREGIVER SURVIVAL

- 1) Use the team approach: enlist the help of others. The more players on the team, the better. Put an announcement in the church bulletin or the local paper. The local girl or boy scouts can adopt you to help run errands, rake leaves, shovel snow or whatever other help you may need. Take the initiative to assign some tasks to family members and friends—you'll soon realize which individuals are good team players and which ones are not. Allowing others to do easier tasks can leave you more energy for the more difficult tasks.
- 2) Plan time in your schedule for activities that you enjoy. Maintain this schedule and make it a priority. Some caregivers join a photo club, others have purchased season tickets at the local symphony or theater, whereas others schedule in time to take long walks, go shopping or meet with friends. Make the breaks in your schedule part of the daily routine.
- 3) Arrange backup caregiving that is regular. Have Aunt Shelley come the first Saturday of every month from 8–10 p.m.; have a volunteer from church take the HD-affected individual for a walk every Monday after work from 5–5:30 p.m.; join the local Alzheimer disease “morning out” program and take the HD-affected individual every Wednesday and Friday morning; have your son stay with Dad while you attend the local support group meeting once each month; and so on.
- 4) Many activities will change from the way that you've done them in the past but just because something is different doesn't mean that it's not as enjoyable. Find new ways to enjoy old things. Be prepared for holidays to be different for both you and the person with HD because some family traditions will need to be varied to accommodate the changing needs of both of you.
- 5) Change your expectations for success. Success changes with ability. If you feel a day (or a week, or a visit, etc.) has to go just as it did before onset of HD to be a “good” one, you will spend a lot of time being upset and the person with HD will be the more frustrated for not being able to live up to your expectations. If, however, you redefine success as the situation changes, you'll both be happier. A successful meal is not one completed in 30 minutes, but one completed without major incident. A successful day is not one without frustration, but one with some resolution, communication, or whatever works in your individual circumstance.
- 6) Use humor as much and as often as possible. Rent funny movies, read the comics each day, share the best ones with each other, cut them out and put them on the refrigerator. Don't be afraid to laugh at yourself and even at HD. It's probably the best known treatment for HD at this time.
- 7) Enjoy several methods of relaxation—yoga, meditation, bubble baths or just sleeping in.

If a loved one with HD resides in a care facility, recognize that it is common to “put off” visiting. Sometimes the atmosphere is difficult for visitors to observe. It may be reminiscent of where we all believe we will spend our final days and we may not want to think about it. Some family members share that “I hate to

visit because all I hear are gripes and problems that I can't solve." Avoiding visitation, however, is not helpful. Feelings of guilt or dread typically strengthen, rather than weaken, with time. This potential for emotional build-up and drain surrounding the visit can heighten the possibility of stressful visits. Structure the visits and limit the time of the visits. It may be helpful to consider the visit as "refilling the gas tank" for the HD person. Many families have found it helpful to have two people visit at one time, rather than one, as they can "carry" the conversation together. Bring photos, news clippings and videos of people that are important to the HD-affected person. Inform them about the news on the home front. Avoid topics that are typically argumentative. Plan ahead of time how long the visit will last. Frequent short visits, if feasible, seem to work better than long visits. Mark on the calendar when the next visit will occur and stick to it!

■ Common Behavior Concerns in HD

COMMUNICATION

Although there are many potential causes for behavior difficulties, the number one reason for any problem is poor communication. It is important to recognize that communication problems are not unique to HD. Leading marriage therapists say that poor communication is the number one reason for divorce; some sociologists believe that poor communication is the foundation for international tension and, possibly, war. Over the years, several strategies have been developed to assist people with communication. Radio, television, newspapers, magazines, telephones, cell phones, pagers, computers, the Internet, e-mail, fax machines, video tape and audio tape have all been created or used to assist in communication. So, even without HD, communication is challenging.

DEFINITION

Communication, or the transfer of information from one person to another, requires a complex integration of thought, muscle control, and breathing. HD can impair all three of these functions. There are two main aspects to communication: getting the information *in* (understanding) and getting the information *out* (expressing).

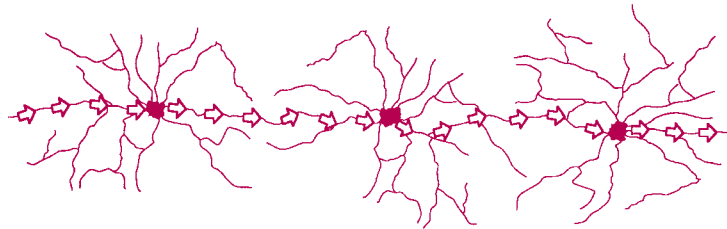
POSSIBLE CAUSES OF MISCOMMUNICATION

Communication with a person affected by HD can be a difficult task because the main aspects of communication can be impaired by the disease. The most prominent language difficulties in people with HD are:

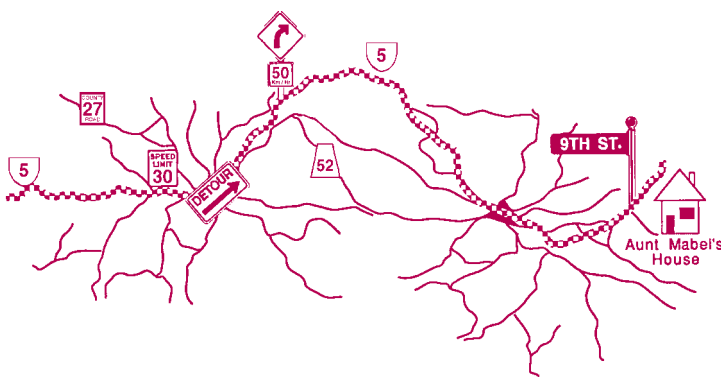
- Speaking clearly (articulation)
- Starting conversation (initiation)
- Organizing what is to be said
- Understanding what is being said

Articulation impairments have become one of the hallmark symptoms of HD. Many persons affected by HD have been accused of “drinking too much” because their speech is slurred. One of the “circuits” that relays information through the caudate is the motor circuit—the pathway for motor information that tells the body how to move specific muscles at precisely the right time. When the caudate becomes affected by HD, it is unable to regulate how motor information flows from one part of the brain to another. One consequence is that some motor movements occur randomly (this is known as chorea). Another consequence is that the brain is unable to control motor movements throughout the body. Talking requires very complex motor movements of the mouth and tongue, breathing control and so on. Without the caudate to regulate all of the different aspects of talking, speech becomes disorganized, poorly timed and not adequately supported by breath. Unfortunately, slurred speech is one of the main reasons that unaffected individuals stop communicating with HD-affected persons. Unaffected individuals are uncomfortable that they cannot understand what is being said to them and, oftentimes, they stop trying.

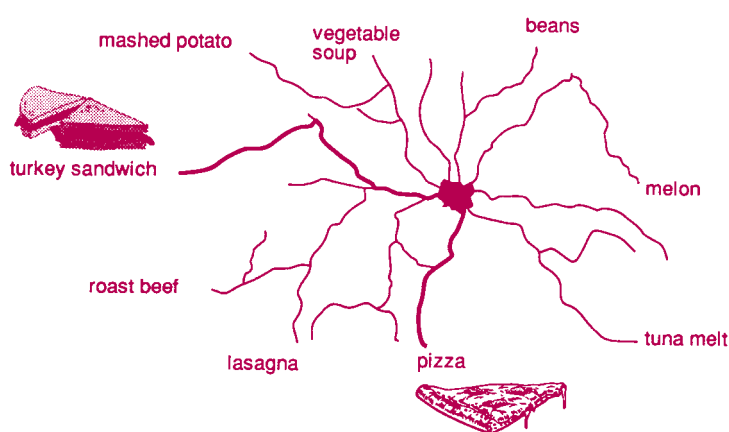
Initiating, or starting, speech is also severely impaired by the damage to the caudate in the brain. The degeneration that occurs in the caudate is due to the death of the medium spiny neurons (a specific type of brain cell). Here is a picture of three of this kind of neuron. Notice the number of long dendrites that flow off of the center, or body, of each neuron. These long dendrites are used to send information through one neuron and on to the next.



As a neuron degenerates, or dies, the information cannot be passed along the circuit as easily. Imagine the dendrites of the neuron to be roads in a large freeway system. The nucleus of the neuron would be a large city and the roads (dendrites) lead to neighboring towns. Let’s ask this neural circuit a question, like one you would ask a person with HD. “How do you get to Aunt Mabel’s house?” Typically, we take Highway 5 north to Lakewood Exit and then take County Road 52 to Vinetown, where Aunt Mabel lives on 9th Street. We begin the journey in the HD “freeway system” only to find that Highway 5 is closed for road construction, and County Road 52 has been washed out by a recent mud slide and 9th Street is being resurfaced and cannot be accessed until Thursday. Try to find a detour that can get you to Aunt Mabel’s house. It is difficult *and* it takes longer to get there.



As HD causes the neurons in the caudate to degenerate, the ability of the brain's circuits to communicate becomes more and more impaired. It may take longer and longer for the person with HD to answer your question. He/she will experience increasing difficulty simply finding the right words. It may take the person with HD much longer to respond. Consequently, several common scenarios occur: (a) the person with HD becomes frustrated and/or fatigued and gives up; (b) the listener becomes frustrated and/or fatigued and gives up; (c) the listener assumes that the person with HD cannot answer the question and continues with yet another question; (d) the listener assumes that the person with HD cannot answer the question and makes the decision unilaterally.



Here is another example of a typical question asked to a person affected by HD: “What would you like for dinner?” It is easy to assume that people with HD no longer have preferences for food and no longer care about their diets. However, it is more likely that the degeneration of the neurons causes such long delays to get their preferences out that one of the communication failures cited above occurs (i.e., patient or listener gives up or listener assumes an answer). HD does not

typically erase an individual's personal opinions. The method of asking and answering questions may need to be altered to have successful communication. For instance, it might be more effective to give the person with HD a choice of available menus for dinner. “Would you prefer pizza or turkey for dinner?” Giving such cues can assist the neuronal communication in the brain. As the brain hears the word “pizza,” specific pizza neurons activate and are more easily retrieved.

Organizing your words and ideas logically is what helps others understand what you mean to communicate. As the caudate deteriorates, access to the part of the brain that helps to sequence, organize, prioritize, and delegate tasks (the frontal lobes, or “boss”) is compromised (see p. 2). Without the boss to tell the brain what information should go first and what information should wait until later, the output, or speech, of the HD-affected person becomes garbled.

Understanding of speech can become impaired if the organization of input is affected by HD. Sometimes, individuals with HD have difficulty understanding or acting upon information that is given to them. Typically, the HD-affected person understands each unique word without difficulty, but complex sentences and paragraphs become mixed up without the “boss” to organize them. Similar to the organization of the information that is going out of the brain, the information that is coming into the brain is difficult to organize when the caudate cannot regulate information for the frontal lobes. Although the person with HD can hear information without difficulty, the components of the information become jumbled and mixed up without the caudate and frontal lobes to keep them separated.

Tim Doesn't Help with Chores: Lazy or Unable?

Beth asked her brother, Tim, to help clean the house. Five minutes later she returned to find him watching TV, not a dust rag in sight. She became very frustrated and angrily asked him why he would not help with the housework.

Understanding, or comprehension, can be impaired by HD. In Tim's case, he has difficulty doing multi-step tasks (e.g., dusting the furniture in the house). What seems to be a simple request may be too complex for the individual with HD to start and complete without assistance. The instructions may be too complicated or not sufficiently detailed. We often underestimate how complicated a task is. A task we can perform daily without thinking can be quite complex without a boss or brain organizer. For instance, getting dressed has a number of steps (e.g., consider the weather and the day's activities, select clothes, assess their colors and patterns for matching, assess cleanliness and then organize the order to put them on—pants before shoes, etc.). In this case, Tim is not opposed to helping with housework and has the physical capabilities to dust but he is unable to organize the steps needed to do the task (i.e., go to the pantry, choose a dust rag, open lower right-hand cabinet and choose dust spray, go back to family room and dust shelves, TV, chair rails, picture frames, knickknacks and window sills).

A slowed response time may erroneously suggest that comprehension is impaired. For example, Dr. Swenson asked Mr. Wilson to name the current month. He did not respond. After three-and-a-half minutes, however, when Dr. Swenson was no longer in the room, he did answer correctly: Dr. Swerdlow asked about his mood, to which Mr. Wilson responded "October." Dr. Swerdlow might consider Mr. Wilson to be demented or confused if he failed to appreciate the circumstances. As the caudate deteriorates, the organization of information going in and coming out of the brain takes much longer. Other consequences of the impaired caudate may be a lack of response or an outburst, possibly due to overload, i.e., too much information or information presented too quickly.

COPING STRATEGIES FOR COMMUNICATION

Although we are unable to stop the progression, we can manage the speech difficulties associated with HD, thus enabling the individual to maintain communication as long as possible. It is important to remember that the disease process can be as long as twenty years or more. Maintaining communication from the beginning is essential. Some professionals have described the late-stage HD patient as "locked in," meaning that the individual is unable to communicate the most basic requests or desires despite relatively normal understanding of his or her surroundings. When people cannot control the muscles or find the words to express their own thoughts, it can be painful, frustrating and embarrassing for everyone.

SUGGESTIONS FOR COMMUNICATION

- Allow the person enough time to answer questions or express him/herself. Remember, it is not necessary to speak slowly yourself, since the person with HD generally understands well. He or she just needs additional time to respond.
- Offer cues and prompts to get him or her started. Fill in words that may be on the tip of the tongue, but be sensitive about “putting words in someone’s mouth.”
- Give choices. Rather than asking, “What do you want for dinner?” give specific choices like, “Do you want lasagne or spaghetti?” or, “Would you like Mexican or Japanese food?”
- Break the task or instructions down into small steps. Remind yourself that most of our daily tasks are very complex activities.
- If the person is confused, modify what you are saying by making it simpler and shorter.
- If asking a question, phrase it in a “yes or no” or a “this or that” format.
- Ask the person to repeat what you did not understand; don’t fake it.
- Ask them to give you the first letter of a word you don’t understand or to spell the word.
- Modify the steps of a request as the person becomes more impaired. Use simple words and short sentences.
- Demonstrate what you are saying or use visual cues.
- Alphabet boards, yes/no cards or other technical communication devices should be made available to every person with HD to allow for at least a simple method of communication.
- Once a person with HD loses the ability to communicate verbally, *do not stop talking to them*. This can only intensify the feelings of isolation that accompany losing speech.
- Request speech therapy.

MEMORY

DEFINITION

Memory is the ability to learn and remember information. The primary memory problems in individuals affected by HD are the following:

- 1) An impaired ability to learn new information.
- 2) An impaired ability to recall remembered information.

POSSIBLE CAUSES

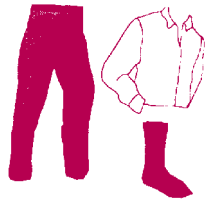
1) An impaired ability to learn new information. This is most likely due to the disruption of the circuits connecting the frontal lobes and the caudate in the brain. Without efficient usage of the frontal lobes, persons with HD experience impairments in the ability to organize and sequence the information to be learned. When information is not organized in an efficient manner, retention and recall of the information is very difficult.

For example, try to learn this list of words: *pants, shirt, socks, peach, cherry, apple, hammer, wrench, pliers, drum, flute, and trumpet.*

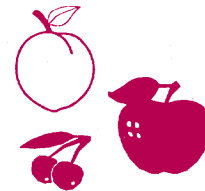
Now try to learn this list of words: *computer, toothpaste, horse, butter, truck, field, arm, ball, coffee, pencil, straw, and uncle.*

computer
toothpaste
horse
butter
truck
field
arm
ball
coffee
pencil
straw
uncle

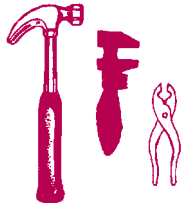
vs.



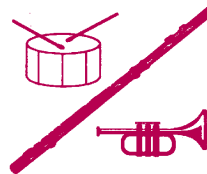
pants
shirt
socks



peach
cherry
apple



hammer
wrench
pliers



drum
flute
trumpet

It is much easier to learn the list of words with shared categories (clothing, fruits, tools, musical instruments) because you can organize the similar items together as a “chunk” and remember the four categories rather than the entire 12 words.

Learning may also become more and more difficult because the impaired caudate makes it difficult to use “divided attention.” **Divided attention** is one type of attention that we use most of the time to attend to more than one thing at a time. For instance, we often drive a car and listen to the radio, we talk on the phone and watch TV, or we prepare the family meal and talk. When the caudate becomes unable to “filter” or regulate information as it travels in the brain, it becomes impossible to use divided attention because all of the information tries to come through the circuit at the same time, causing overload.

2) An impaired ability to recall the remembered information. Oftentimes it appears that persons with HD have difficulty remembering information, but the reason for this deficit is that HD disrupts the search mechanism to find the desired word. For instance, when you ask Katie to tell you what she had for breakfast, she may not respond. It would be a misconception to think that Katie did not remember what she had for breakfast. When you ask Katie whether she had pancakes or cereal for breakfast, she is able to identify the correct choice. As described in the previous section on communication, HD can disrupt the ability to freely locate the right word for something. When people are offered a choice from which they can recognize (rather than recall) the memory, they usually perform normally.

COMPARISONS BETWEEN HD AND ALZHEIMER DISEASE

The table below compares some of the problems of persons affected by HD with those of people affected by Alzheimer disease. It is very important to discriminate the ability to learn from the ability to remember. Typically, persons with “true” memory problems have difficulty with the latter—remembering. For instance, persons with Alzheimer disease have severe problems remembering information that was once learned. The comparison is not made to note the similarities between HD and Alzheimer disease, both commonly referred to as “dementia.” Rather, the table points out the numerous differences between these two disorders. From everything we know about Huntington’s disease, it does not involve a primary memory deficit, and, consequently, it is not useful to refer to HD as a dementia.

ABILITY	HUNTINGTON’S DISEASE	ALZHEIMER DISEASE
<i>Speed of processing</i>	Slow, but relatively accurate	Slow, often inaccurate
<i>Speech output</i>	Slurred and slow but accurate	Normal in clarity and rate; often the incorrect word
<i>Learning new information</i>	Disorganized and slow but can learn	Rapid forgetting, defective storage of information
<i>Free recall of memory</i>	Impaired: cannot find the right word; can recognize with choices, benefits from cues	Impaired: memory store is defective; cannot recognize, cues don’t help
<i>Motor memory</i>	Impaired: cannot learn or recall motor memories	Intact: can learn and retain motor memories

EXECUTIVE FUNCTIONS

DEFINITION

Executive functions are critical to our ability to care for ourselves as well as manage our job, home and family. These functions fall into four categories—organization, self-regulation, attention and problem solving—but include a long list of skills. These skills, which we often take for granted, include the following: planning, thought organization, sequencing, prioritizing, follow-through, problem solving, decision making, controlling feelings, judgement, creativity (i.e., generating new ideas, coming up with options for life problems), attention/concentration and abstract thinking.

POSSIBLE CAUSES

The frontal lobes are often referred to as the boss of the brain and are responsible for executive functions. When the frontal lobes or their connections with the caudate are damaged, even simple tasks that we would normally take for granted can become difficult and frustrating.

Before we proceed with some of the behaviors that can change due to frontal lobe disconnections, it might help to review our understanding of brain functions and circuits. Remember, a circuit is a pathway by which information travels in the brain. Brain circuits operate much like other circuits that we are familiar with in our daily lives (see pages 2-8).

APATHY AND DIMINISHED ABILITY TO INITIATE ACTIVITIES

A Problem with Initiation: Elsie's Story

Elsie did not have a problem with apathy. She still enjoyed and cared about many things in her life. She was very disappointed, however, that she never did anything anymore. She seemed to just sit around the house all day, sometimes never getting up from her easy chair except to use the toilet. After complaining about her inactivity to several others, including her family, it was agreed that the family would try to be Elsie's "boss" and begin activities for her. This strategy worked very well. Elsie was a good "follower" and enjoyed all of the activities that her family members encouraged her to attend or participate in. After about six weeks, however, it became evident that the family was experiencing a significant amount of distress having to guide and begin every one of Elsie's activities. First of all, the family was small. Secondly, all of the other family members had full-time commitments to work and/or school. They grew tired of providing constant initiation for Elsie and began to neglect her. One day Elsie's daughter came home from work with an abandoned puppy which had been left at school. At the family's protest, Elsie adopted the puppy. The next time Elsie and her family were seen in support group, the whole family announced that the puppy had become Elsie's "boss." He initiated playtime, dinnertime, time to go outside to the bathroom and time to go for a walk. Elsie was no longer inactive!

DEFINITION

Apathy is similar to inattention, indifference and lethargy. Apathy is present when individuals with HD seem to have diminished concern for things about which they used to demonstrate a great deal of care and concern. A lack of initiation often accompanies apathy but can also occur by itself in the absence of apathy. The ability to begin, or initiate, behaviors, conversation or activity is a very complex function which is frequently compromised in persons with various types of brain dysfunction, including mild head injury, Parkinson disease, multiple sclerosis, stroke and, of course, HD.

POSSIBLE CAUSES

The circuits in the middle and bottom sections of the frontal lobes are very well-connected to the limbic system, or the "emotional lobe," of the brain. The caudate degeneration can result in disconnections in these circuits, causing the frontal lobe boss to be disconnected from the "feelings" of the brain.

A common, and normal, response to having HD is sadness, but this sadness can sometimes be a warning sign of a more serious problem called depression. Often, when individuals become depressed, they also have reduced concern and energy for other aspects of life.

COPING STRATEGIES FOR APATHY

- Do not interpret a lack of initiation or activity as “laziness.” Educate family members and friends about the inability to “begin” or “initiate” behaviors and ask others to encourage participation in activities. Gently guide behaviors but respect “no.”
- Use calendars and routines as suggested earlier (see pages 7, 19, 20).

ORGANIZATION

DEFINITION

Difficulties in planning, thought organization, sequencing, and prioritizing can affect attempts to follow a recipe, to complete a list of household errands, to apply for social security benefits or to put on a dinner party. These same organizational impairments affect problem solving and logical thinking (deductive reasoning). When the brain cannot sequence bits of information, many aspects of intellectual, social and personality functions are impaired.

EXAMPLES

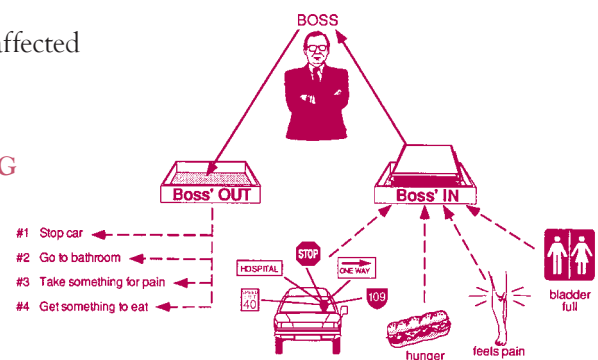
Errands that used to be completed in two hours now take all day and even then may not be completely done. Problems might include the following: the errands were not written down and some were accidentally skipped; they were not organized in a logical and efficient way, e.g., from closest to farthest from home; necessary papers were overlooked and left behind, so a trip back home was needed.

COPING STRATEGIES FOR DISORGANIZATION: SUGGESTIONS FOR IMPAIRED PLANNING

- Make lists that help to organize the individual tasks in the order needed to do an activity.
- Prompt each step of an activity.
- Rely on routines that can be much easier for the HD-affected person to initiate or continue without guidance.

SUGGESTIONS FOR IMPAIRED DECISION-MAKING

- Offer some choices but not too many at once.
- Avoid power struggles.
- Avoid open-ended questions.
- Use short, simple sentences.



IMPULSE CONTROL

DEFINITION

Dysfunction of the caudate nucleus and frontal lobe of the brain may cause difficulty regulating or controlling emotions and impulses. This is called impulsivity or disinhibition. Impaired impulse control may be the reason that some persons affected with HD easily lose their temper, begin to drink too much or have inappropriate sexual relations. Also, disinhibition can sometimes contribute to illegal behaviors such as stealing. Disinhibition usually presents itself as trouble controlling a sudden desire to do or say something that comes to mind, even when the behavior is hurtful, repetitious or socially or sexually inappropriate.

POSSIBLE CAUSES

The living or home environment can contribute to disinhibited responses if it is chaotic or without a reliable routine. Environments without routine (i.e., meals are at different times every day, activities are not planned but spontaneous) may provoke greater confusion or anxiety which, in turn, may lead to a greater number of outbursts and behavior problems. Mild feelings of confusion, annoyance, frustration, irritability, or anxiety may be expressed as strong feelings such as anger, rage or fear.

Damage to the caudate nucleus or circuits connecting the caudate and the frontal lobes can also contribute to impulsivity. One of the primary functions of the caudate is to regulate, or control, the information from the rest of the brain. As the caudate is affected by HD, the regulation mechanisms of the brain break down. Without the caudate, the brain cannot regulate how much movement, how much feeling or how much thinking is required in a certain situation.

EXAMPLES

“He was arrested for stealing a pair of blue jeans while a salesperson was right behind him.”

“She had a tantrum because the family chose Taco Bell over McDonald’s.”

“He asks women he’s never met to have sex.”

“She spent half her rent money on a new television.”

“He yelled at our neighbor friends, ‘Turn down that music or I’ll call the cops!’”

COPING STRATEGIES FOR IMPULSIVITY

- Remember, although the things being said are hurtful or embarrassing, generally the person is not doing this intentionally.
- The person may be remorseful afterward. Be sensitive to any efforts to apologize.
- It is possible that a behavior is a response to something real that needs your attention. Don’t be too quick to discount it as an outburst.
- Since individuals with HD cannot control their responses, a routine and predictable daily schedule can reduce confusion, fear and, as a result, outbursts.

- Medications may be helpful for outbursts and sexually inappropriate behavior. Talk to your neurologist or psychiatrist.
- Let the person know that yelling is not the best way to get your attention and offer alternative methods for getting your attention.
- *Stay calm and in control.* This will help you remain able to think and not react emotionally and impulsively yourself. It will also reduce the chances of reinforcing maladaptive attention-seeking behavior. In addition, staying calm may help the person calm down or at least not enrage him or her more. Develop a thick skin. This is the HD talking, not your loved one. Do not badger the person after the fact. It won't help. Remember, this lack of control most likely is not by choice.
- Label the feeling for the HD person: "I realize that you are feeling angry/frustrated."

FRUSTRATION, IRRITABILITY, ANGER AND TEMPER OUTBURSTS

Frustration, irritability, anger and temper outbursts are behaviors that most of us experience in our lives regardless of whether we have HD. These behaviors seem to increase when a person becomes affected with HD because of the deterioration of the caudate. Aggressive behaviors can be particularly disturbing to the family because they can be a source of fear and tension in the household or in the care facility. Irritability can take different forms. Irritable responses can become exaggerated in intensity and duration or punctuated by episodes of explosiveness.

EXAMPLES

"When I have trouble understanding what he is saying he quickly becomes frustrated and sometimes violent. I guess he feels his actions speak louder than words."

"She insists upon balancing her own check book even though she can no longer perform this task successfully. When I try to help out she becomes angry with me."

"When I tried to introduce a new activity he became irritable and refused to make an attempt to participate."

"I asked him to get dressed for the recreation center. He said 'No,' so I said, 'Come on, you need to get dressed,' and he slapped me."

POSSIBLE CAUSES

- Feelings of frustration and anger, and their subsequent reactions, are usually triggered by real and legitimate events in life but the brain cannot control the intensity of the response.
- Frustration and irritability can arise from circumstances that emphasize a loss of ability to function physically, cognitively or emotionally. Underlying causes or triggers of this kind of behavior include hunger, pain, inability to communicate, frustration with failing capabilities, boredom, difficult interpersonal relationships and, in particular, minor changes in routine. Usually, anger is an emotion that "covers" for

another emotion. Typically when an individual experiences anger the underlying feeling is disappointment, grief, hurt, frustration or anxiety.

- Frustration, irritability, anger and temper outbursts are expressed in response to a true feeling, but with HD they become exaggerated due to a loss of control from the caudate.

COPING STRATEGIES FOR FRUSTRATION

- An awareness of the person's capabilities is very important so that he/she is encouraged to be as independent as possible and is allowed to take risks without constantly exposing him/herself to failure. Consider a "happy medium" of responsibilities. A non-institutionalized adult should be responsible for something. Choose tasks that are appropriate (e.g., water a few plants, sweep a small area) but not overwhelming (weed the garden, clean the whole house). Close attention should be paid to the signals, verbal or nonverbal, that the patient is upset or wanting something so that he/she does not have to get to the stage of making a fuss before receiving attention.
- Knowledge of the person and sensitivity to his/her needs means that some situations can be anticipated and potential frustration defused. It may be possible to identify situations that trigger frustration and either avoid them or provide diversional activities.
- Often an overachiever needs help to give up peripheral responsibilities. For example, one caregiver encouraged his wife, "Let's ask Julie to take over the telephone chain for the women's auxiliary. Of course, you will still try to go to as many meetings as possible."

COPING STRATEGIES FOR IRRITABILITY

- Restructure the person's interactions, expectations, and responsibilities. Such restructuring may need to take place frequently as more activities become difficult.
- Family members and caregivers should learn to respond diplomatically, acknowledging the patient's irritability as a symptom. Confrontations and ultimatums should be avoided unless the issue is crucial. The environment should be as calm and structured as possible.
- In addition to support by clinicians, caretaker/family support groups can be invaluable, both in providing emotional comfort and in sharing strategies that members have found effective in their own households.

COPING STRATEGIES FOR TEMPER OUTBURSTS

- Redirect the HD person away from the source of anger.
- Try to identify circumstances that trigger temper outbursts and then avoid them.
- Assess your own expectations regarding the HD-affected individual. A family member may be unwilling or unable to accept the patient's new limitations. Therefore, there must be a restructuring of interactions, expectations and responsibilities.
- Confrontations and ultimatums should be avoided.

- The environment should be as calm and structured as possible. People with HD tend to become mentally inflexible and are typically comforted by stability. Establish daily routines and break them as infrequently as possible.
- There are medications that can control irritability. It is important to see a physician who has current knowledge of Huntington's Disease.
- HD in itself does not cause an individual to become dangerous but a loss of normal regulation of impulses can contribute to unsafe situations. Family members should be responsible for providing a safe environment so that no person is ever in danger. Remove potential weapons from the house and have emergency numbers near the telephone.

DENIAL AND UNAWARENESS

DEFINITION

Denial in an individual with HD is common. There are at least two reasons why denial can occur. Commonly, denial is considered a psychological inability to cope with distressing circumstances. We often see this type of denial in cases such as loss of a loved one (denial that they are gone), terminal disease (denial of cancer or HD diagnosis), serious illness, or injury. This type of denial, however, typically decreases over time as the individual begins to "face reality." In contrast, individuals with HD often suffer from a lack of insight or self-awareness. They are unable to recognize their own disabilities and are unable to evaluate their own behavior. This type of denial is sometimes called organic denial and is a condition that may last a lifetime. Given that we typically assume that denial is under the control of the individual, the term may not be useful for persons with HD suffering from this organic type of denial. Therefore, we recommend that "unawareness" be used to describe this behavior in HD.

Unawareness often plays a significant role in difficult and seemingly irrational behavior. On one hand, unawareness is beneficial because it keeps individuals motivated to try things and to avoid labelling themselves as "affected" or "impaired." Unawareness can also be a useful defense mechanism against depression. On the other hand, anger may develop from unawareness because individuals with HD cannot understand why they cannot go back to work or live independently. HD persons with unawareness sometimes feel that people are unjustifiably keeping them away from activities that they could do (e.g., driving, working, caring for children). You may hear an individual with HD enumerate a long list of people who are at fault for his or her failure to return to work, to drive, to travel or to live alone.

This type of unawareness can become dangerous if the individual with HD (and unawareness) attempts to do things independently that are not safe. Often, these individuals are considered to have poor judgement. Judgement is impaired, in this case, because of the unawareness of limitations that HD can create.

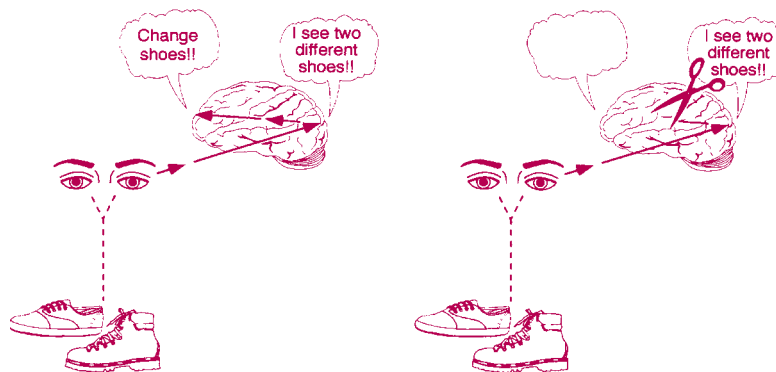
Unawareness is not only a problem for the individual with HD, but also for health professionals, friends and family members. There are some family members and/or health professionals who delay making the

diagnosis or keep the diagnosis from the affected individual because they are concerned that he/she “cannot handle it.” Some people interpret the unawareness as a sign that the individual “does not want to know” or will “get depressed” if he/she receives the diagnosis of HD. There is no evidence to suggest, however, that talking about HD to a person with unawareness will cause negative consequences.

POSSIBLE CAUSES

- Damage to circuits connecting the caudate nucleus and the frontal and parietal lobes.
- Normal psychological response to an overwhelming situation.
- The circuit that relays information from other centers in the brain to the frontal lobes (where appropriate actions are considered and initiated) is interrupted by caudate degeneration secondary to HD.

In the final case, although the person with HD may still be able to see and hear accurately, the information seen and heard is often not available to the boss or decisionmaker of the brain. Consider the example below. The eyes see that the person has two different shoes on and the visual information gets sent to the occipital cortex, which has cells to read shape, color, and movement. This information then travels toward the frontal lobes, where the boss will instruct the “motor controller” to change shoes. When an individual has HD, however, the cells in the caudate are breaking down and the information never gets relayed back to the frontal



lobes.

EXAMPLES

Miles continues to say that he will live with his wife and she will care for him, even though she has filed for divorce.

Bob bathes and shaves only when asked, due to his difficulty recognizing how he smells or when his hygiene is poor.

Claire will not go to the doctor. She says, “Nothing is wrong with me, I’m not sick.”

Steve will not say he has HD.

When asked, Beverly says she has no change in her speech or walking and no uncontrollable movements, although she has severe chorea and slurred speech.

Mary notes that her speech and coordination are not the same as they used to be but says it is because of the “fender bender” she was involved in two years ago.

Rosie never complains. She denies needing help at home, yet her legs, arms and hips are badly bruised. (Many HD patients are unrealistic about the potential for falls and injuries in the tub or shower. Don’t expect them to raise this issue.)

STRATEGIES FOR UNAWARENESS

- There is no single way to cope with this difficulty. At times it may require the caregiver to use creative thinking to get the person with HD to co-operate with a request.
- Accept “unawareness” of illness as a component of HD that is not a treatable obstacle. Often, the person with HD does not seem to change or “accept” the HD. Stop expecting the awareness to “kick in”—the “ah-hah!” may never occur.
- Avoid interpreting non-compliance with therapy or nursing care as intentional. It may be helpful to develop a contract that includes incentives for compliance. It is important that the rewards (e.g., foods, activities) be things that the person likes, not just things the caregiver chooses.
- It may be that the person can talk about his or her problems, but not acknowledge that he or she has HD. This being the case, address the problems and avoid the HD label.
- A formal written agreement that explains expectations will increase the chances for success. Make goals realistic and avoid requiring awareness and/or acceptance.
- Counselling may help someone with HD come to terms with his/her diagnosis but may have little impact on specific insight.

REPETITION AND PERSEVERATION

DEFINITION

A person with HD may perseverate or become “stuck” on one idea or activity. Individuals may become rigid in their behavior and become unable to change easily from one activity or idea to another or to alter their routines. While true obsessive-compulsive disorder (OCD) is uncommon in HD, the behaviors associated with OCD are often seen. OCD is an obsessive preoccupation with certain ideas, which can be seen in persons with HD who perseverate about obtaining cigarettes, getting coffee refills, having a meal or using the lavatory. Individuals may become irritable when these requests are ignored or denied.

POSSIBLE CAUSES

- Damage to the frontal lobes or their circuitry
- Legitimate needs of the individual are not being met and, consequently, the HD individual continues repeating the request in hopes of being understood.

EXAMPLES

“He’ll chain smoke if I let him.”

“She repeatedly tells me that she wants to get her own apartment and to start her own business.”

“She worries about how she’ll pay for a car although she no longer has her driver’s license.”

“She incessantly uses demonstratives such as ‘honey’, ‘baby’ and ‘dear’ even in situations where these are inappropriate.”

COPING STRATEGIES FOR REPETITION

- Attempting to alter these behaviors is a difficult challenge. While rational argument does not have much effect, providing the individual with a sense of security and an alternative way to express his or her feelings may be an effective way to reduce frustration for both the patient and caregiver.
- Distract the individual. Maintain a list of activities or have various foods that are of interest to the individual. These items can be used to gently shift patients off a perseverative topic. Humor may also effectively break a cognitive “set” (a topic that the person is stuck on).
- Explain that this topic has been discussed. If no acceptable conclusion was agreed upon, gently remind the individual where you are in the process of problem-solving.
- Some perseverations are unsolvable. Try setting limits on how long you will talk about this.
- Sometimes “real life” experience can stop a repetitive notion. For example, help the person who wants to drive but is clearly unable to, to go through the tests at the local motor vehicle licensing authority and let the authority make the decision. Oftentimes your physician can be the “bad guy” and limit activities that the individual refuses to limit at your request. In this manner, you can maintain a positive relationship with the HD-affected person.

DEPRESSION

Depression is a common problem for persons with HD. There are two reasons for this. First, a saddened mood is a natural and understandable emotional response to the diagnosis and symptoms of HD because it progressively alters an individual’s role in the family, work and social realms. For example, the individual’s role in their family may change from breadwinner and household manager to a dependent person requiring supervision. Secondly, the brain changes that occur in HD can directly alter the neurotransmitters or the chemicals that regulate moods. Consequently, even persons who are naturally optimistic and happy can experience severe depression secondary to the brain changes in HD.

DEFINITION

Many of the symptoms of HD resemble and may disguise the symptoms of depression. Some of these include memory loss, lack of concentration, apathy, weight loss, and sleep disturbance. It may be difficult to

tell whether a person's symptoms are depression, HD or a combination of both. It is important that the doctor be regularly informed of the symptoms and changes in behavior. The suicide rate for persons who are symptomatic with HD is seven times the national average. Therefore, do not hesitate to contact the family doctor if the HD affected person may be depressed.

SIGNS OF DEPRESSION

Seeing two or more of these symptoms almost daily over a two-week period is good reason to contact your family doctor:

- Sleeping most of the day or rarely sleeping
- Decreased ability to find pleasure or interest in usual activities
- Depressed mood most of the day, nearly every day
- Significant decrease or increase in appetite or weight
- Fatigue or loss of energy nearly every day
- Restlessness or slower movements
- Feelings of worthlessness
- Decreased ability to think, concentrate or make decisions
- Recurrent thoughts of death or committing suicide

RISK FACTORS FOR SUICIDE

- Depressed mood
- Increase in life stress or upcoming stressful event
- Withdrawal from activity and the desire to be alone
- Talk about "ending it all," "not dealing with it anymore" or death
- Talk about or active pursuit of a plan
- "Putting their life in order" by saying goodbye (although the signs are not usually this obvious)

SUICIDE PREVENTION

If you consider your loved one to be at risk:

- Talk about it!
- Ask your primary physician for a referral to a mental health specialist.
- Keep in touch with a doctor to periodically evaluate mood changes.
- Remove all potential weapons from the home, including guns, bullets, rope and medications.
- Put household cleaners and other solvents in locked cabinets.
- Post emergency phone numbers by the phone.
- Make a "contract" with the person to let you know if he or she begins to feel badly.
- Arrange for supervision.

Many chronically ill patients hide multiple medication bottles “in case I need them” all over the house, apartment or room. An occasional unobtrusive check while the person with HD is bathing or eating may be wise. It is not only suicide that is problematic but a decision such as “I’m not right today—I need to take a few of those pills.”

ANXIETY

DEFINITION

It is common for an individual with any chronic progressive illness to experience some excess anxiety, or worries, about the future. Oftentimes, however, symptoms of anxiety can become so severe that they are problematic. Anxiety can present in a variety of ways, including general nervousness, restlessness, repetitive thoughts about troublesome topics, fidgeting hands, shallow breathing, sweating, rapid heart rate, fear or panic. Typically, anxiety symptoms are worse when individuals are in new situations or perceive themselves to have insufficient skill to handle a situation. Often, anxiety is associated with depression. Many individuals with HD find that they worry more than they used to. Even about seemingly trivial matters, excess worry occurs. For instance, several patients have reported that the day prior to a doctor’s appointment is particularly anxiety-ridden.

When anxiety symptoms become severe, other anxiety disorders can be present, such as panic disorder or obsessive-compulsive disorder. **Panic disorder** is characterized by an acute onset of overwhelming anxiety and feelings of dread often accompanied by rapid heart rate, sweating, hyperventilation, light-headedness or numbness and tingling of fingers and toes. The symptoms typically last about 15 minutes, but residual anxiety often remains. **Obsessive-compulsive disorder** is characterized by senseless recurrent intrusive thoughts or impulses (obsessions) that are anxiety-provoking. **Compulsions** are repetitive behaviors that are performed over and over, sometimes in response to an obsession or as part of a stereotyped routine that must be followed. The most common obsessions tend to focus on cleanliness (such as washing hands) or safety (such as checking to make sure the stove is turned off). Although true panic and obsessive-compulsive disorders are rare in HD, they can occur. More common are components of these anxiety disorders, such as obsessive preoccupation with particular ideas.

STRATEGIES TO AVOID ANXIETY

- Establish routines and stick to them.
- Keep the environment simple.
- Keep demands simple (one step at a time).
- Refrain from discussing future stressful events until the day before.
- Plan on changes in routine and compensate for these stressful times.

STRATEGIES TO MANAGE ANXIETY

- Remove excessive stimulation from the environment. For instance, turn off radios, limit conversations to one in the room, remove ringing phones, turn off the TV and so on.
- Take a deep breath of air into the lungs, hold it, and let it out slowly. Repeat as needed. Close eyes and focus attention on breathing.
- Make positive self-statements, such as, “You can do this,” “Be calm” and “It’s OK.”
- Discontinue the present activity that may be contributing to the feelings of anxiety.
- Pursue relaxation training and stress management counseling.

PSYCHOSIS: HALLUCINATIONS AND DELUSIONS

DEFINITION

Seeing, hearing or experiencing things that are not real is considered a hallucination. For instance, some individuals may see illusions or hear voices and feel bugs crawling on them when nothing is really there. Thoughts about unreal situations and relationships are considered delusions. For example, thoughts that someone is out to get you, watching you or reading your mind are usually paranoid symptoms of a delusion. Fortunately, psychosis is rare in HD but it can occur.

STRATEGIES TO MANAGE PSYCHOSIS

- Consult a psychiatrist. Psychosis is typically managed well with medications.
- Provide frequent reality checks and orientation cues.

ALTERED SEXUALITY

Although changes in sexual behavior are often uncomfortable to discuss with family, friends and professionals, they are very common in persons with HD. Changes in the brain can be associated with changes in sexual interest and functions. Some persons with HD report that they have increased sexual drive whereas others report diminished sexual interest. Increased promiscuity can be secondary to disinhibition, poor judgement or impulsivity. Decreased sex drive can be secondary to depression, apathy or an inability to initiate activity. Although the reasons for sexual behavior changes are not fully understood, changes in sexual functioning often need to be addressed.

POSSIBLE CAUSES

- The brain is no longer able to regulate, or gate, the amount of sexual drive a person has, resulting in too much or too little.

- The delicate balance of hormones in the brain is disrupted by the presence and progression of Huntington's Disease, resulting in variations in behaviors typically regulated by hormone levels.

EXAMPLES

Bob is "single" for the first time in 20 years but does not have the social skills to appropriately initiate relations.

Jim is taking a tricyclic antidepressant for a moderate depression and still desires intercourse with his wife but is unable to sustain an erection.

Julie is embarrassed about the way her body looks with constant movements; her husband is saddened that she has pulled away from their sexual intimacy.

Terry has increased his sexual relations dramatically; his family is concerned about sexually transmitted diseases.

ADDRESSING THE CHANGES IN SEXUAL FUNCTIONING

Each individual has the right to achieve his/her highest reasonable potential on the continuum of human sexual development. It is a misconception that inheriting a degenerative disease will cause an end to one's sexuality. There are several ways to better adjust to the changes that Huntington's can bring. Most important is the need to maintain communication. Readers may want to review the bullets that highlight improving communication and do so with your sexual relationships in mind (see page 14). Access to supportive services, educators and counselors can also be valuable, and community health centers should be able to provide a list of local resources. Sexuality is a lifelong process of learning about oneself and growing as a social and sexual being. All people have a right and a need to be fully and accurately informed about the unique pleasure, joy, and pain this aspect of identity can bring. Remember to allow yourself to respond to change and adjust as needed. Respect the space and development of those around you, and keep in mind that rarely is it just one party who is affected by change.

SLEEP DISTURBANCES

A normal sleep cycle is divided into two distinct periods with brief intervals of wakefulness. A typical night's sleep involves multiple cycles. One part of the cycle is non-rapid eye movement (NREM), entered when the body begins to slow down and eye movement stops. The brain activity, heart rate, blood pressure and metabolism are slowed to enter into a deep restful state. Another part of the cycle is rapid eye movement (REM). This stage is characterized by fast eye movement, dreams lasting up to thirty minutes and the brain being 20 percent more active than when we are awake. NREM and REM sleep alternate in cycles throughout the night with occasional brief intervals of wakefulness.

Over 50% of older persons report that they have disturbances in their sleep. Normal aging brings about changes in sleep patterns. Older people tend to wake up more often, have lighter sleep and require a longer

time in bed to get adequate rest. A shift in the body's daily rhythms occurs which makes adults prone to waking in the early morning and fatigued in the early evening.

Many persons with Huntington's Disease complain of disturbed sleep. Although the reason for this is not fully understood, there are some causes of disturbed sleep that can respond to medical treatment. Thus, all problems of this sort should be promptly examined by a doctor. Restlessness, wandering and disturbances in sleep are often just a part of the progressing disease and may not respond to medical treatments. All family members need to practice good sleep hygiene because poor sleep can lead to memory impairment, inability to concentrate, interpersonal problems and increased risk of serious illness and accidents.

STRATEGIES TO ENCOURAGE A GOOD NIGHT'S REST

- Maintain a regular schedule, rising and retiring at the same time every day.
- Make the bedroom conducive to sleep by having it cool, quiet and dark at night, but full of light in the morning.
- Avoid excessive napping in the early evening.
- Eat a proper diet that is devoid of heavy, hard-to-digest food as well as alcohol, salty foods, and stimulants (coffee, chocolate, tobacco) before bedtime.
- Exercise regularly, but not within two to three hours of bedtime because it can cause overstimulation.
- When using over-the-counter sleep medications, do it sparingly. Never use them for more than four days unless directed by a physician. One becomes tolerant to the effects of sleep medications over time. That is, if they are used frequently they do not work as well. Too much use of some of these medications can lead to addiction.
- Discuss your medication schedule with your physician. Certain antidepressants and asthma drugs may be too stimulating for before-bedtime use. Try to minimize the number of medications being used.
- Take a warm shower or bath before going to bed. This will ease the transition to sleep.
- To reduce night waking due to a full bladder, limit the amount you drink after dinner. Make sure to use the toilet before bed.
- Establish a safe environment in case of night-time wandering. There are a number of ways to make it hard to get out of the house or yard without using restraints (e.g., bells on doors, hard-to-open doorknobs, deadbolt locks, oddly placed latches)

AUTHOR'S NOTE

I am very aware that this book will not have all the answers. I know that we will continue to be frustrated and to marvel at the challenges that HD brings. I trust that the HD family will continue to meet often, share behavior concerns associated with HD, and offer insights and support. I am certain that the readers of this book will also continue to learn and share information about persons with HD. Our fight against HD must continue to support the practitioner and the scientist, the person and the neuron, the care and the cure for this disease and the people and caregivers living with HD on a daily basis.



Appendices

Appendix I

Voluntary Organizations and Other Sources of Help

National and International Lay Organizations

The national HD lay organizations offer a range of services and care programs to benefit people with HD and their families. In addition, they operate research, education and advocacy programs, and are a useful source of information and referrals for both families and health care professionals.

HUNTINGTON'S DISEASE SOCIETY OF AMERICA

505 Eighth Avenue, Ninth Floor
New York, NY 10018
Phone: 800-345-HDSA
Fax: 212-239-3430
Email: hdsainfo@hdsa.org
Web site: www.hdsa.org

HUNTINGTON SOCIETY OF CANADA

151 Frederick Street, Suite 400
Kitchener, Ontario N2H 2M2
CANADA
Phone: 519-749-7063
Toll-free in Canada: 800-998-7398
Fax: 519-749-8965
Email: info@hsc-ca.org
Web site: www.hsc-ca.org

For information on, or referral to, lay organizations in other countries, contact:

HUNTINGTON'S DISEASE SOCIETY OF AMERICA

Go to the HDSA national website at www.hdsa.org and click on "About", click "About HDSA" and then "Helpful HD Links" or contact"

INTERNATIONAL HUNTINGTON ASSOCIATION

www.huntington-assoc.com

Appendix II

Brain Tissue Banks/DNA Bank and HD Research Roster

Brain Tissue Banks

The greatest gift to research and future generations is the donation of the HD patient's brain. For information on brain tissue donation, write or call:

University of California at San Diego
Alzheimer's Disease Research Center
(also performs HD research)
Phone: 858-622-5800
Email: jlgoldstein@ucsd.edu

Massachusetts General Hospital
Department of Neurology
Charlestown (Boston), MA
Phone: 617-726-1254
Email: hersch@helix.mgh.harvard.edu

**Harvard Brain Tissue
Resource Center**
McLean Hospital, Belmont, MA
Phone: 1-800-Brainbank
Email: btrc@mclean.org
Website: www.brainbank.mclean.org

Buckeye Brain Bank
Ohio State University
Department of Neurology
Phone: 614-293-8531
Note: Due to storage limitations, the Buckeye
Brain Bank is restricted to OSU patients only.

**The New York Brain Bank at
Columbia University**
New York, NY
Phone: 212-305-5779
Beeper: 917-889-2045 (emergency donations)

Sun Health Research Institute
Sun City, AZ
Email: lucia.sue@sunhealth.org
Phone: 623-876 5328
Website: www.sunhealth.org/shri

University of Rochester
Department of Neurology,
Movement Disorders Center
Rochester, NY
Phone: 585-341-7500
(if clinic closed ask for movement disorders
physician on call)

**National Neurological Research
Specimen Bank**
VA Medical Center, Neurology Research,
Los Angeles, CA
Phone: 310-268 3536
Email: brainbnk@ucla.edu
Website:
<http://www.loni.ucla.edu/~nnrsb/nnrsb>

University of Washington
Laboratory of Neuropathology
Phone: 206-731-6315
Email: cfederha@u.washington.edu

**Loyola University Medical Center/
Hines VA Brain Bank**
Department of Pathology
Maywood, IL
Phone: 708-216 8270
E-mail: dmagnus@lumc.edu

DNA Bank and HD Research Roster

The roster is a vital link between scientists and HD families to facilitate research. All information is strictly confidential. The DNA Bank was established for the purpose of storing genetic material for possible future use. Cost to store a sample is \$70.00. For information contact:

Indiana University Medical Center

975 West Walnut Street

Indianapolis, IN 46202

HD Roster: 317-274-5744

Email: sfox@medgen.iupui.edu

DNA Bank: 317-274-5745

Email: sraig@medgen.iupui.edu

Appendix III

HDSA Chapters

HDSA chapters provide sources and referrals for local and community resources. Information may change over time. Please visit the HDSA national web site at www.hdsa.org for updated chapter information or call 800-345-HDSA to locate the HDSA chapter closest to you.

ALABAMA

Alabama Support Group

5750 Picketts Lane
Pinson, AL 35126
Phone: 205-325-3877

ARIZONA

Arizona Affiliate

P.O. Box 7666
Phoenix, AZ 85282
Phone: 888-267-3411
Fax: 480-394-0511
HDSA_AZ@hotmail.com
www.hdsa-az.com

ARKANSAS

Arkansas Affiliate

c/o Sandra Boll
HDSA Midwest
466 Fox Trail Drive
St Louis, MO, 63367
Phone: 800-558-3370
800-536-1728 (Pin no. 5935)
hdsa-arkansas@comcast.net

CALIFORNIA

Greater Los Angeles Chapter

9903 Santa Monica Blvd.
Suite 106
Beverly Hills, CA 90212
Phone: 888-4-HDSA LA
888-443-7252
800-686-9868

Northern California Chapter

3940 Industrial Blvd, Suite 100 D
West Sacramento, CA 95691
Phone: 916-372-1895
Fax: 916-371-2468
Helpline: 888-828-7344
www.hdsanortherncalifornia.org

Orange County Affiliate

c/o Dennis Mesnick
HDSA Regional Development Director
1017 F Street
San Diego, CA, 92101
Phone: 619-544-1792
hdsamesnick@cox.net

San Diego Chapter

P.O. Box 19524
San Diego, CA 92519-0524
Phone: 760-752-1844
800-473-4014
www.hdsasandiego.org

COLORADO

Rocky Mountain Chapter

6545 West 44th Ave., Unit 1
Wheat Ridge, CO 80033
Phone: 303-321-5503
877-740-HDSA
303-837-9937
www.hdsarockymountain.org

CONNECTICUT

Connecticut Affiliate
P.O. Box 719
Southington, CT 06489
Phone: 508-872-8102
Fax: 508-872-8103

FLORIDA

South Florida Chapter
12555 Biscayne Blvd.
North Miami, FL 33181
Phone: 305-274-7411
Fax: 305-665-3038

GEORGIA

Georgia Chapter
P.O. Box 15298
Atlanta, Georgia 30333
Phone: 770-729-9207
Fax: 678-461-3518
www.hdsaga.org

ILLINOIS

Illinois Chapter
PO Box 8383
Rolling Meadows, IL 60008
Phone: 630-443-9876

INDIANA

Indiana Chapter
P.O. Box 2101
Indianapolis, IN 46206
Phone: 317-271-0624
Fax: 317-722-7614
www.hdsaindiana.org

IOWA

Iowa Chapter
600 N. 21st Street
Clarinda, IA 51632
Phone: 866-248-IAHD (4243)
712-542-4976
Fax: 712-379-3317
hdsaiowachapter@hotmail.com

KANSAS

Kansas Affiliate
c/o Sandra Boll
HDSA Midwest Regional Development
Director
466 Fox Trail Drive
St Louis, MO, 63367
Phone: 314-313-3644

KENTUCKY

Kentucky Chapter
c/o Kosair Charities
982 Eastern Parkway
Louisville, KY 40217
Phone: 502-637-4372
800-784-3721
Fax: 502-637-4310
HDSAKyChapter@aol.com

MARYLAND

Maryland Chapter
The Rotunda
711 West 40th St
Baltimore, MD 21211
Phone: 630-443-9876
410-467-5388
Fax: 410-467-4143

MASSACHUSETTS**Massachusetts Chapter**

1253 Worchester Road, Suite 202
Framingham, MA 01701
Phone: 508-872-8102
888-872-8102
Fax: 508-872-8103
www.hdsa-ne.org

MICHIGAN**Michigan Chapter**

Sparrow Dimondale Center
4000 N. Michigan Road
Dimondale, MI 48821-9774
Phone: 517-646-0920
800-909-0073
Fax: 517-646-0885

MINNESOTA**Minnesota Chapter**

22 27th Avenue
Suite 212
Minneapolis, MN 55414
Phone: 612-371-0904
612-371-6268
www.hdsa-mn.org

MISSOURI**St. Louis Chapter**

8039 Watson Road
Suite 132
St. Louis, MO 63119-5325
Phone: 314-961-4372
866-707-HDSA
Fax: 314-961-5754
hdsa@stlouis.missouri.org

NEW JERSEY**New Jersey Chapter**

114B South Main Street
Box 67A
Cranbury, NJ 08512
Phone: 609-448-3500
Fax: 609-448-3521
www.hdsanj.com
HDSAnjoffice@aol.com

NEW YORK**Long Island Affiliate**

c/o Michelle Crepeau
HDSA Greater New York Regional
Development Director
505 Eighth Avenue, Ninth Floor
New York, NY 10018
Phone: 212-242-1968
Fax: 212-239-3430

Upstate New York Chapter

115 Hardwood Lane
Rochester, NY 14616
Phone: 585-341-7400
www.hdsauny.org

NORTH CAROLINA**North Carolina Chapter**

P.O. Box 240353
Charlotte N.C. 28224-0353
Phone: 704-525-1835

OHIO**Central Ohio Chapter**

490 City Park, suite C
Columbus, OH 43215
Phone: 866-877-HDSA (4372)
614-460-8800
Fax: 614-460-8801
www.hdsacentralohio.org

Northeast Ohio Chapter

7059 Old Mill Road
Chesterland, OH 44026
Phone: 440-423-HDSA (4732)
Fax: 440-423-0515

Ohio Valley Chapter

3537 Epley Lane
Cincinnati, OH 45247
Phone: 513-741-HSDA (4372)
Fax: 513-741-4645

OKLAHOMA**Oklahoma Chapter**

1313 Val Genes Road
Edmond, OK 73003
Phone: 405-236-4372
www.okhdsa.org
okhdsa@sbcglobal.net

PENNSYLVANIA**Delaware Valley Chapter**

525 Plymouth Road, Suite 314
Plymouth Meeting, PA 19462
Phone: 610-260-0420
Fax: 610-260-0423

Western Pennsylvania Chapter

P.O. Box 110223
Pittsburgh, PA 15232
Phone: 412-833-8180
888-779-HDSA (4372)
www.hdsawpa.org

SOUTH DAKOTA**Sioux Valley Chapter**

PO Box 1311
Sioux Falls, SD 57101
Phone: 605-334-9917

TEXAS**Texas Affiliate**

PO Box 270261
Flower Mound, TX 75027
Phone: 972-724-1367
800-910-6111
www.hdsatexas.org

WASHINGTON**Northwest Chapter**

PO Box 33345
Seattle, WA 98133
Phone: 206-464-9598
888-264-4372
www.geocities.com/nwhdsanwhdsa@yahoo.com

WASHINGTON D.C.**Washington Metro Chapter**

8303 Arlington Blvd., Ste 210
Fairfax, VA 22031
Phone: 703-204-4634
703-323-1403
Fax: 703-573-3047

WISCONSIN**Wisconsin Chapter**

2041 N 107th St
Wauwatosa, WI 53226
Phone: 414-257-9499
877-330-2699
www.hdsa-wi.org

Appendix IV

HDSA Centers of Excellence

HDSA Centers of Excellence for Family and Services are established at major medical institutions that HDSA has identified as having expertise in Huntington's Disease or movement disorders. The Centers provide a multidisciplinary approach to the treatment of HD and their services include professional social workers, genetic counseling & testing, speech, physical and occupational therapies, educational programs and family support groups. **New Centers are added on a regular basis. Call (800) 345-HDSA or visit www.hdsa.org for updates.**

ALABAMA

HDSA Center of Excellence at the HD Clinic at the Children's Hospital of Alabama

1600 7th Avenue South, CBH 314
Birmingham, AL 35233
Contact: Donna Pendley
Tel: 205-996-7850
Fax: 205-996-7867
Email: dpendley@peds.uab.edu

CALIFORNIA

HDSA Center of Excellence at University of California Davis Medical Center

Department of Neurology
4860 Y Street, Suite 3700
Sacramento, CA 95817
Contact: Terry Tempkin
Tel: 916-734-6278
Fax: 916-734-6525
Email: teresa.tempkin@ucdmc.ucdavis.edu

HDSA Center of Excellence at University of California - San Diego

200 West Arbor Drive, Outpatient Center,
Third Floor, Suite 1
San Diego, CA 92103
Contact: Jody Goldstein
Tel: 858-622-5854
Email: jlgoldstein@ucsd.edu

HDSA Center of Excellence at UCLA

Department of Neurology
The Regents of the University of California
300 UCLA Medical Plaza, Suite B200
Los Angeles, CA 90095
Tel: 310-794-1589
Fax: 310-794-7491
Genetic Counselor: Michelle Fox
Tel: 310-206-6581
Email: mfox@pediatrics.medsch.ucla.edu

COLORADO

HDSA Center of Excellence at the Colorado Neurological Institute

Movement Disorders Center
701 East Hampden Avenue, Suite 530
Engelwood, CO 80110
Contact: Sherrie Montellano
Tel: 303-788-4600
Fax: 303-788-8854
Email: montellano@megapathdsl.net

FLORIDA

University of South Florida Huntington's Disease Clinic

Health Sciences Center
12901 Bruce B. Downs Blvd (MDC 55)
Tampa, FL 33612
Contact: Marci McCall
Tel: 813-974-6022
Fax: 813-974-7200
Email: mamccall@hsc.usf.edu

GEORGIA

HDSA Center of Excellence at Emory School of Medicine
Wesley Woods Health Center
1841 Clifton Road
Atlanta, GA 30329
Contact: Joan Harrison
Tel: 404-728-6364
Fax: 404-728-6685
Email: jharri2@emory.edu

ILLINOIS

HDSA Center of Excellence at Rush University Medical Center
1653 W. Congress Parkway
Chicago, IL 60612
Contact: Jean Jaglin
Tel: 312-563-2900
Fax: 312-563-2684
Email: jean_a_jaglin@rush.edu.

INDIANA

Indiana University HDSA Center of Excellence
Indiana University School of Medicine
Department of Medical and Molecular Genetics
975 West Walnut Street
Indianapolis, IN 36202-5251
Contact: Carrie McGinnis
Tel: 317-274-3487
Fax: 317-278-1100
Email: cmcginni@iupui.edu

IOWA

HDSA Center of Excellence at University of Iowa Hospitals and Clinics
Psychiatry Research 1-145 MEB
Iowa City, IA 52242
Contact: Anne Leserman
Tel: 319-353-4307
Email: hdinfo@uiowa.edu

MARYLAND

HDSA Center of Excellence at Johns Hopkins
Johns Hopkins Hospital
Ross 618
720 Rutland Avenue
Baltimore, MD 21205
Contact: Debbie Pollard
Tel: 410-955-2398
Fax: 410-455-8233
Email: dpollard@jhmi.edu

MASSACHUSETTS

New England HDSA Center of Excellence
MGH East Bldg. 114, Suite 201
114 16th Street
Charlestown, MA 02129
Contact: Yoshio Kaneko
Tel: 617-724-2227
Fax: 617-724-1227
Email: ykaneko@partners.org

MINNESOTA

HDSA Center of Excellence at Hennepin County Medical Center
701 Park Avenue South
Minneapolis, MN 55415
Contact: Shelly Anderson
Tel: 612-873-2595
Fax: 612-904-4270

MISSOURI

HDSA Center of Excellence at Washington University School of Medicine
660 S. Euclid
Campus Box 8018
St. Louis, MO 63110
Contact: Melinda Kavanaugh
Tel: 314-362-3471
Fax: 314-747-3471
Email: kavanaughm@neuro.wustl.edu

NEW YORK**HDSA Center of Excellence at Columbia Health Sciences/ New York State Psychiatric Institute**

630 West 168th Street,
P & S Box 16
Contact: Debra Thorne
Tel: 212-305-9172
Fax: 212-305-2526
Email:
thorned@sergievsky.cpmc.columbia.edu

George C. Powell HDSA Center of Excellence at North Shore University Hospital

300 Community Drive
Manhasset, NY 11030
Contact: Mary Ellen Benisatto
Tel: 516-869-9527
Fax: 516-869-9535

HDSA Center of Excellence at University of Rochester

1351 Mount Hope Avenue, Suite 220
Rochester, NY 14620
Contact: Leslie Briner
Tel: 585-273-4147
Fax: 585-341-7510
Email: leslie.briner@ctcc.rochester.edu

OHIO**HDSA Center of Excellence at Ohio State University**

1581Dodd Drive
371 McCampbell Hall
Columbus, OH 43210
Contact: Nonna Stepanov
Tel: 614 688-8672
Fax: 614-688-4060
Email: stepanov-1@medctr.osu.edu

TEXAS**HDSA Center of Excellence at Baylor College of Medicine**

6550 Fannin, SM 18011
Department of Neurology
Houston, TX 77030
Contact: Christine Hunter, RN
Tel: 713-798-3951
Fax: 713-798-1488
Email: chunter@bcm.tmc.edu

VIRGINIA**HDSA Center of Excellence at University of Virginia**

500 Ray C Hunt Drive
Charlottesville, VA 22903
Contact: Pat Allinson
Tel: 434-924-2665
Fax: 434-924-1797
Email: psa9m@hscmail.mcc.virginia.edu

WASHINGTON**HDSA Center of Excellence at University of Washington**

1959 North East Pacific Street
CHDD Bldg., Room 411
Seattle, WA 98195
Contact: Debbie Olson
Tel: 206-616-2135
Fax: 206-616-2414
Email: olsondl@uwashington.edu

Appendix V

Rehabilitative/Adaptive Equipment and Product Information

The following list is provided for reference purposes only. HDSA does not endorse or recommend any product, service or company listed. This list was accurate as of publication. HDSA is not responsible for any changes subsequent to that date.

Bedding, Padding, Low Beds, Bed Enclosures

NOA Medical Industries

1601 Woodson
St. Louis, MO 33114
800-368-2337
Low Beds

Vail Products

235 First Street
Toledo, OH 43605
800-235-VAIL
Bed enclosure

Profex Bumper Pads

PO Box 16043
165 N. Meramec, Suite 120
St. Louis, MO 63105
800-325-0196
Foam products; adaptive equipment

Chairs

Broda Seating

385 Phillip Street
Waterloo, Ontario N2L 5R8
519-746-8080
800-668-0637 (Canada and US)
Specialized HD Chairs

Gunnel, Inc.

8440 State Street
Millington, MI 48746
517-871-4529
800-551-0055
Gunnel Custom Recliner, customized wheelchairs

Hill Rom

1069 State Roads 46 East
Batesville, IN 47006
800-445-3730
Customized wheelchairs

May Corporation

Industrial Park South
PO Box 140
612-944-6450
800-525-3590
Posture Guard (wheelchair with body guard); customized wheelchairs

PDG, MedBloc

700 Ensminger Road, Suite 112
Tonawanda, NY 14150
888-433-6818
Bently Chair

Schwartz Medical

1032 Stuyvesant Avenue
Union, NJ 07083
908-687-1122
800-4SCRIPT
Customized wheelchairs

Walking Devices

Sunrise Medical

7477 East Dry Creek Parkway
Longmont, CO 80503
818-504-2820
800-255-5022
Grandtour walking device, rolling walker,

Guardian Products

745 Design Court, Suite 603
Chula Vista, CA 91911
800-423-8034
Walkers; adaptive equipment

Sammons Preston

PO Box 5071
Bolingbrook, IL 60440
800-323-5547
*Strider walker, standard walker,
adaptive equipment*

Rehabilitation Aids and Safety Products

Access to Recreation

2509 East Thousand Oaks Blvd., Suite 430
Thousand Oaks, CA 91362
800-634-4351
*Adaptive equipment for recreation and
activities of daily living*

North Coast Medical

18305 Sutter Boulevard
Morgan Hill, CA 95037
800-821-9319
Adaptive equipment

Alimed

297 High Street
Dedham, MA 02026
800-225-2610
Adaptive equipment

J.T. Posey Co.

5635 Peck Road
Arcadia, CA 91006
800-44-POSEY
Positioning devices; Adaptive equipment

Skil-Care

29 Wells Ave.
Yonkers, NY 10701
800-431-2972
Positioning devices; safety products

Smith & Nephew Rolyan
 N104 W13400 Donges Bay Road
 Germantown, WI 53022
 800-558-8633
Padding materials; Adaptive equipment

Food Preparations

Food Thickeners:

Diafoods Thick-It Food Thickener
 Precision-Milani Foods, Inc.
 2150 North 15th Avenue
 Melrose Park, IL 60160
 800-333-0003

Consist-Rite
 Donmar Fodds
 150 Industrial Parkway North
 Aurora, Ontario L4G 4L3
 905-726-3463

Thick 'N Easy Instant Food Thickener
 Hormel Health Labs
 1 Hormel Place
 Austin, MN 55912
 800-866-7757

Food Molds:

Culinary Puree, Inc.
 6001 Felstead Road
 Evansville, IN 47712
 800-981-7744

Cookbooks:

The Thick N' Easy Recipe Book
 Hormel Health Labs
 1 Hormel Place
 Austin, MN 55912
 800-866-7757

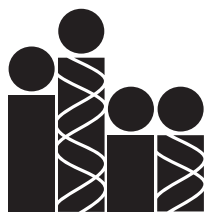
Blending Magic
 Bernard Jensen Products
 PO Box 8
 124 East Cliff Street
 Solana Beach, CA 92075

Non-Chew Cookbook
 J. Randi Wilson, 1985
 Local bookstore or online bookseller

Information and Supplies for Pureed Foods

Diamond Crystal Specialty Foods
 10 Burlington Avenue
 Wilmington, MA 01887
 800-255-0592
 (Institutions only)

MedDiet Labs
 3600 Holly Lane, Suite 80
 Plymouth, MN 55447
 800-633-3438



Huntington's Disease Society of America
505 Eighth Avenue, Ninth Floor
New York, New York 10018
212-242-1968 • 1-800-345-HDSA • www.hdsa.org

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