

A Caregiver's Guide to Huntington's Disease

March 2011



A Caregiver's Guide to Huntington's Disease

Introduction to Caregiving.....3

What is Huntington's Disease (HD)?.....3

The Genetics of HD.....3
Onset and Treatment of HD.....3

Symptoms of HD.....4

After the Diagnosis.....4

Team-based Care.....4

The Multidisciplinary Team.....4

Movement Disorder Symptoms.....6

Chorea.....6
Poorly Coordinated or Poorly Controlled Movements.....6
Abnormal Walking and Problems with Balance.....6

Living with the Movement Disorder.....7

Safety Modifications in the Home.....7
Seating.....7
Sleeping.....8
Communication.....8

Nutrition and HD.....9

Making Eating Easier.....9
Distractions.....10
Nutrition in Late-Stage HD.....10

Cognitive Problems.....10

Attention and Concentration.....10
Memory and New Learning.....11
Fixed Ideas and Obsessions.....11
Lack of Awareness of Symptoms.....11
Caregiving and Cognitive Issues.....12

Behavioral Problems.....12

Depression.....12
Apathy.....12

Anxiety.....13
Impulse Control.....13
Irritability and Outbursts.....13
Managing Outbursts.....13
Emotional Outbursts and Caregiving.....14
Avoiding a Crisis.....14

Navigating the Transitions.....15

Working Successfully with Medical Professionals.....15

Keeping Track — The Care Notebook.....16

Caring for the Caregiver — Respite and Support.....17

Respite Care.....17
Choosing a Source of Respite Care.....17

Joining a Support Group.....18

Sources for Support Groups.....18

Preparing for Late-Stage HD.....18

Out-of-Home Care.....18
Living Wills and Advance Directive.....19
Medical Power of Attorney.....19
Tube Feeding and Hydration.....19
Hospice Care.....19

Hope for the Future.....19

Resources.....20

Acknowledgements

WE MOVE and the Huntington's Disease Society of America recognize the following people for their contributions to the vision and creation of this booklet:

Medical Reviewer Samuel A. Frank, MD
Assistant Professor of Neurology, Boston University; Attending Physician and Director of the Dystonia and Huntington's Disease Clinic, Boston Veterans Administration Hospital; Consultant Neurologist, Tewksbury State Hospital, Director Huntington's Disease Clinic

Medical Writers Karen Tarapata | Catherine F. Murray, NASW, CSE

Content Reviewer Debra Lovecky, Director of Education, Programs and Services, Huntington's Disease Society of America

Editors Joy B. Leffler, MLA, NASW, Director of Education & Informatics, WE MOVE
Judith Blazer, MS, Executive Director, WE MOVE

WE MOVE and the Huntington's Disease Society of America wish to acknowledge Lundbeck, Inc. for providing the funding to create and distribute this publication to the HD community.

Introduction to Caregiving and HD

If someone you care about has been diagnosed with Huntington's disease (HD), this guide has been written for you.

In these pages, we will cover some of the major symptoms of HD and how the symptoms will change over time. We will also offer suggestions and strategies for coping with the daily challenges presented by HD. We have drawn from the experiences of countless other HD families and the medical professionals who serve them. As a caretaker, you have an essential role to play in your loved one's life and care. This guide was developed to help support you during the journey that is HD.

Just as importantly, this guide will help you to connect with the informed, welcoming community of HD professionals and families that you can turn to for support.

What is HD?

Huntington's disease (HD) is a hereditary brain disorder. Symptoms usually appear in the prime of life, between ages 30 and 50, but there have been individuals affected by the disease as young as two or as old as 80. HD causes widespread physical and mental disabilities that become more and more severe over a long period of time, up to 20 years or more.

The Genetics of HD

People sometimes refer to the "HD gene." Huntington's disease is not a gene, but a change in one of the genes that every human being is born with. Only people who are born with this genetic abnormality (also called a gene expansion) can develop Huntington's disease. Unlike viruses (like HIV or the "flu"), no one can "catch" HD from another person.

If an individual has been diagnosed with HD, they inherited the expanded gene from a parent. Each child of a parent with HD has a 50% chance of inheriting the genetic change that causes the disease. This means that within a family with HD, each child has the same

50/50 chance of being free of the disease. If children are free of the disease, they can never pass it on. HD does not skip generations. The disease affects males and females equally. HD can be passed down from either the mother or father.

There is a genetic test which can show whether a person has the expanded gene that will cause HD. Your loved one may have had this test to confirm his or her diagnosis of HD.

If the test has not yet been performed, or if a test is being considered for another family member, it is highly recommended that a genetic counselor and neurologist familiar with HD be consulted before moving ahead. The decision to test is highly personal and complicated. Because there is no cure for HD at this time, many people at-risk for the disease choose not to be tested.

Onset and Treatment of HD

What is called the "onset" of HD occurs when the expanded gene begins to trigger changes to the brain and the person shows outward signs of the disease. Exactly how, why and when this happens is the subject of intensive, ongoing research.

At this time, there is no cure or treatment that will halt the progression or delay onset of HD, but there are medications and therapies which may relieve some symptoms in certain individuals. These will be discussed throughout the guide.



Symptoms of HD

The most obvious symptom of HD is chorea — involuntary tics, twitches and jerky movements that flow between body parts. But the movement disorder is only one aspect of the disease. The progressive loss of nerve cells in the brain affects not only movement, but also the capacity to control emotions and behavior, as well as the ability to think and reason.

Disabilities in one area will have an impact on other areas. HD is a disease that affects the entire person.

General Symptoms of HD Include:

- ▶ Unsteady gait and involuntary movements.
- ▶ Clumsiness and difficulty with voluntary movements.
- ▶ Slurred speech and difficulty in swallowing.
- ▶ Personality changes, mood swings and depression.
- ▶ Impulsiveness and impaired judgment.
- ▶ Poor attention and repetitive thinking.

After the Diagnosis

A diagnosis of HD may cause a variety of emotions—anger, outrage, disbelief, sadness—and even, perhaps, relief. You may have already been noticing changes in your loved one's behavior, such as impulsiveness, lack of concentration, or unusual irritation. With a diagnosis of HD, you have confirmed that what you have witnessed is real.

Now that there is a diagnosis, you can begin to educate yourself about the disease and your role as caregiver. Reading this guide is just a beginning. We recommended that you access the large and active HD community of caregivers and medical professionals for ongoing support and informed guidance.

There is much to understand about living in a family affected by HD—not only how to manage your loved one's symptoms, but also how to create a community of support for yourself.

One of the first steps after a diagnosis of HD will be setting up the multidisciplinary team who will provide the varied aspects of medical care.

Team-Based Care

The Multidisciplinary Team

HD is a complex brain disorder that affects all aspects of life. Providing the best care for a person with HD takes the expertise of many professionals in different areas of medicine and healthcare.

Your loved one may already be seeing a neurologist, psychiatrist or physician who specializes in brain disorders as part of the diagnostic process. Over the course of the disease, this doctor (also called the primary managing physician) may need to call upon other medical specialists and allied health professionals to address the treatable symptoms of HD. As the caregiver, you may be the one to ask the doctor to make referrals for your loved one.

Here are some of the health professionals who may be part of the team:

Neurologist

- ▶ May be the primary managing physician and team leader.
- ▶ Provides management of neurological symptoms such as chorea and other movement disorders.
- ▶ Provides assessment and counseling about changes to reasoning (cognition).

Psychiatrist

- ▶ May be the primary managing physician and team leader.
- ▶ Addresses conditions that may be caused by or associated with HD such as: depression, anxiety, behavioral difficulties, obsessions, compulsion or psychosis.

Mental Health Therapist/Psychologist

- ▶ Provides supportive counseling at initial diagnosis or during difficult transitions.
- ▶ Provides assessment and treatment for frequent or severe irritability/anger.

Social Worker

- ▶ Assists with applications for disability and other benefits.
- ▶ May serve as the primary therapist and/or supportive counselor.
- ▶ Provides guidance, financial planning and advance medical directives.
- ▶ Helps in accessing community services.
- ▶ Aids in finding placement outside the home.

Occupational Therapist

- ▶ Suggests strategies to increase functional independence in daily life.
- ▶ Assesses home safety and makes recommendations (furniture placement, handrails, etc.).
- ▶ Recommends adaptive equipment for seating, feeding, hygiene, etc.

Physical Therapist

- ▶ Evaluates difficulties in walking and the need for home mobility equipment (scooters and wheelchairs).
- ▶ Develops an exercise program to improve balance, reduce pain, increase flexibility, build strength, improve function and increase range of motion.

Speech and Language Pathologist

- ▶ Helps address speech difficulties through exercise.
- ▶ Suggests strategies to reduce choking and swallowing problems.
- ▶ Can help find and train to use communication devices when needed.

Dietician or Nutritionist

- ▶ Looks at diet to address weight loss or gain.
- ▶ Adapts diet to address swallowing difficulties.

Nurse

- ▶ Provides case management, in-home care and telephone support.

In addition, because HD progresses over so many years, your loved one will need to continue to have general medical checkups, eye care and dental care.



The Need for Team-Based Care

Even doctors who treat brain disorders may not be familiar with HD and the many interrelated symptoms that it causes. They may need to be convinced of the need for referrals to other health professionals. Here are some points to make when discussing the need for team-based care:

- ▶ HD is a complex disorder with symptoms that affect more than just movement. It is a condition that causes severe disabilities in reasoning (cognition) and behavior as well.
- ▶ There is currently no cure for HD, but there are many symptoms of the disease that can be treated.
- ▶ HD is a disease that progresses over many years to decades and the goal of HD management is to maintain optimal functioning and comfort.
- ▶ Physical therapy, occupational therapy, speech therapy, swallowing therapy and mental health therapy are among the treatments that can help address some of the symptoms of HD.
- ▶ The collected expertise of a team of allied health professionals will provide the highest level of care to the HD patient.

Movement Disorder Symptoms

Huntington's disease is a condition that affects the entire person. The disease has an impact on movement, the capacity to control emotions and behavior and the ability to learn, think and reason. Although each of these areas is associated with its own unique disorders, symptoms in one area can worsen symptoms in another area. For example, a person's emotional outbursts might be triggered by difficulty with chewing and swallowing. Understanding the connections between the various disorders can help you see the bigger picture when looking for solutions to problems that arise.

The most obvious symptoms of HD are related to movement. HD often causes muscles to move by themselves (involuntary movements) and also affects control over purposeful physical actions (voluntary movement).

As HD causes changes in the brain, you may notice your loved one making involuntary movements, such as facial twitching or flinging of an arm. These movements can be seen more when a person is nervous or excited. You may also see stiffness or involuntary posturing (dystonia), such as arching of the back or extending a finger for several seconds at a time.

Reduced control over voluntary movements can cause people with HD to become clumsy or uncoordinated. They may begin to have difficulty walking, playing sports, eating or writing. Over time, this loss of motor control will make it difficult to move the fingers for fine motor tasks like using a cell phone, typing on a computer keyboard, buttoning a shirt, using a remote control or even going to the toilet without assistance. It will also affect the person's ability to speak clearly, chew or swallow.

Some of the Abnormal Movements Caused by HD Are:

- ▶ Involuntary jerky movements (chorea)
- ▶ Restlessness
- ▶ Stiffness
- ▶ Poorly coordinated or poorly controlled movements
- ▶ Walking and balance problems
- ▶ Slurred or indistinct speech

Chorea

Chorea is a term for involuntary jerking or twitching movements, which can be small or large and tend to flow between body regions. You may notice it in finger flicking, shoulder shrugging, facial grimacing or even flailing of the arms or legs. We now know that chorea also affects speaking, chewing, swallowing and even the small eye movements needed for reading. If chorea occurs near someone, the person with HD can accidentally bump into them. This type of physical contact may not be purposeful and the person with HD may not be aware of the contact with the other person.

There are several types of medications that may be used to keep chorea under control. Because these medicines have potential for side effects, it is important to work closely with a physician (preferably a neurologist) familiar with HD when considering chorea treatments. The goal of reducing chorea needs to be discussed with the patient. Some common reasons to consider treatment of chorea include physical injury, difficulty with dressing or other activities of daily living, falling or social embarrassment.

Poorly Coordinated or Poorly Controlled Movements

Huntington's disease affects coordination and control of small muscles. People can become clumsy and slow moving. They may have difficulty controlling the force of their movements. They may drop things or have difficulty using tools because of trouble keeping muscles contracted (called motor imperistence). Writing and typing on a keyboard may become a challenge. Problems with coordination can also affect speech and the ability to properly chew and swallow. Treatment may include speech therapy, special eating utensils and devices to ease communication.

Abnormal Walking and Problems with Balance

Problems with coordination and balance will affect the way a person with HD walks. They may look unsteady, wobbly or even "drunk" when they walk. They may have trouble judging distances and bump into furniture and doorways. If they trip, they will have a hard time recovering their balance. A physical therapist may be able to help with balance and walking using strengthening exercises, special balance activities, and strategies for safely sitting down and standing up. The impulsiveness of HD can interfere with safe walking or the proper use of a walking aid.

Living with the Movement Disorder

There are many strategies available to the caregiver that can help the person with HD manage their symptoms in daily life. The movement disorder is only one component of the disease; cognitive and behavioral aspects of HD impact the physical parts of the disease. With that in mind, some strategies to help the movement disorder include:

- ▶ Safety modifications in the home.
- ▶ Changes to seating.
- ▶ Sleeping arrangements.
- ▶ Mobility aids.
- ▶ Communication aids.
- ▶ Changes to nutrition.

Safety Modifications in the Home

There are many simple changes and modifications that can be made in the home to increase safety and comfort for the person with HD. You may also wish to have a nurse or occupational therapist visit your home in order to identify areas that can be improved.

In the Kitchen

- ▶ Switch to less breakable plates, bowls and glasses.
- ▶ Make it “standard procedure” to set the kitchen timer when using appliances, as a reminder to turn them off.
 - ▶ Lower the temperature on the hot water heater to prevent scalding.
 - ▶ Use covered “travel” mugs and straws for liquids.
- ▶ Provide oven mitts rather than potholders.



In the Bathroom

- ▶ Put a non-skid mat in the tub or shower.
- ▶ Change to ‘soap on a rope’ or a wall mounted soap dispenser.
- ▶ Get a shower bench or chair.
- ▶ Install safety bars.



In the Living Room and Bedroom

- ▶ Remove tripping hazards, such as scatter rugs or thick-pile carpeting.
- ▶ Move tables and lamps away from walkways.
- ▶ Use chairs with high backs and armrests.
- ▶ Clear rooms of any unnecessary furniture.
- ▶ Pad doorways and furniture, if needed.
- ▶ Remove items on tables and lower shelving.



Seating

The abnormal movements and postures of HD interfere with proper sitting. In addition, HD causes weakness in the muscles of the torso and sitting upright for a long period of time can become uncomfortable.

Adaptive Seating

- ▶ Choose chairs with high backs, good seat depth and padded armrests.
- ▶ Make sure the chair seat is not too high. Solid foot support is important for balance.
- ▶ A small wedge cushion or lumbar support can make the lower back more comfortable.
- ▶ A waist belt may or may not help, depending on the individual. Putting cushioning on either side may work better.
- ▶ Make sure the chair has an open bottom to allow feet to slide under and get a better center of gravity before standing.

Safe Sitting and Standing Techniques

“Touch-Turn-Sit” Technique

This process is frequently taught to people with HD, so that they can safely sit down. This simple strategy solves both the problem of balance and the inability to judge distance.

1. Touch the chair (for stability).
2. Turn slowly (maintain balance).
3. Touch the chair with the other hand.
4. Guide yourself and sit down.

“Hands-on-Knees” Standing Technique

This technique will help the person stand up without losing their balance.

1. Place the hands on the knees.
2. Bring the weight forward and bend slightly at the waist.
3. Place feet under the chair.
4. Push up to a standing position.

Sleeping

Comfortable, safe sleeping can be challenging for people with HD. Falling may occur because of difficulty sensing the edge of the mattress, lack of coordination, or problems with balance. In addition, the inability to control the force of their movements may cause a person with HD to “vault” out of bed when all they wanted to do was roll over or sit up.

Reducing Falls in the Bedroom

- ▶ Avoid high bed frames. Choose a low, padded bed frame or place the mattress on the floor.
- ▶ Move bedroom tables and lamps away from the bed.
- ▶ Side rails on the bed may help or may be a dangerous obstacle. If you install a rail, cover it with at least two inches of foam padding.
- ▶ Keep bedding lightweight and loose.

Communication

As HD progresses, a variety of symptoms may arise to make communication difficult. Speech will be harder to understand and there can also be problems with reasoning (cognition). A speech-language pathologist (SLP) can provide exercises and strategies that can help improve the ability to communicate.

Common Communication Problems in HD

- ▶ Muscular weakness or lack of coordination in the lips, tongue, throat and jaw.
- ▶ Slurred or indistinct speech.
- ▶ Talking too fast or too slow; too loud or soft.
- ▶ Difficulty finding words.
- ▶ Stuttering.
- ▶ Difficulty changing topics.
- ▶ Reduced concentration.
- ▶ Problems with short-term memory.

Improving Communication

Ask the person with HD to:

- ▶ Say one word at a time.
- ▶ Speak slowly and exaggerate the sounds.
- ▶ Take a deep breath before speaking.
- ▶ Repeat the word or sentence, if needed.
- ▶ Rephrase the thought, if needed.

When speaking to a person with HD:

- ▶ Ask one question at a time.
- ▶ Use yes/no questions if possible.
- ▶ If you do not understand, restate what you believe you heard, “Did you say...?”
- ▶ Allow extra time for the person to respond. Wait. A lack of response is not a “no” response.
- ▶ Reduce distractions by turning the volume down on the tv or radio.

Additional communication strategies will be discussed in the section about the cognitive disorder.

Nutrition and HD

As HD progresses beyond the early stage, nutrition can become more of a concern for the caregiver. People with HD can lose weight because of the involuntary movements and an increased metabolism of HD. In addition, the symptoms of HD can interfere with the person's ability to eat appropriate portions of food. Healthy, but calorie rich foods can help keep the weight on, and adapting textures can help reduce chewing and choking problems.

HD Symptoms That Can Interfere with Nutrition Include:

- ▶ Involuntary movements.
- ▶ Difficulty holding eating utensils.
- ▶ Problems with seating.
- ▶ Difficulty chewing and swallowing without choking.
- ▶ Distractibility.
- ▶ Unawareness of hunger.
- ▶ Poor dental hygiene.

Making Eating Easier

Utensils

It may be hard for your loved one to handle food or change the tableware. Clumsiness and involuntary movements can make mealtime messy and frustrating. Changing the dishes and flatware may help.

Select less breakable dishes and glasses.

Soup plates or pasta bowls may be easier to use than flat plates.

Avoid tall or "stemmed" glasses that are top heavy.

"Travel" cups with a cover and attached straw can help prevent spills.

Try spoons and forks with large handles or utensils made for people with arthritis.

Replace placemats with a small non-skid mat to keep dishes from slipping.

Meal Size and Frequency

Smaller, more frequent meals can solve many problems. Offering six to eight small meals a day can solve several of the problems associated with nutrition and HD.

More meals equal more calories, which can help prevent weight loss.

Smaller meals may take less time and may cause less frustration; however, be careful as some people with HD eat too quickly.

Hunger can make anyone irritable and anxious. People with HD can be unaware of their own hunger. Frequent meals can help prevent emotional outbursts.

Ask for a referral to a dietician or nutritionist familiar with HD who can assist in developing a menu that will provide the nutrients and calories needed by your loved one to maintain weight.

Choking

If choking is a problem, try changing the size and texture of food. Soft, moist, tasteful, hot or cold foods are easiest to eat. Identify and eliminate foods that trigger coughing and choking. Typically, these are foods that are crispy, flaky or crumbly. Bland, lukewarm foods tend to be less appetizing.

Serve small pieces of meat and vegetables, and encourage the person to take small amounts in each bite.

Serve soft, moist foods like mashed potatoes, noodles, soup, casseroles, yogurt, pudding, gelatin, ice cream, milk shakes, smooth cooked cereals and macaroni and cheese.

Make foods more smooth and moist by adding gravy, cream sauce, salad dressing, broth, sour cream, mayonnaise or butter.

Offer a covered "travel mug" for hot liquids and a straw with cold liquids to prevent choking.

Avoid acidic beverages if they irritate the throat, cause heartburn or cause choking. These may include orange juice, grapefruit juice, lemon juice and tomato juice.



Avoid spicy and peppery foods if they cause choking.

If liquids and beverages cause choking, try adding one of the thickening products available at drugstores. Cornstarch can also be used as a thickener.

Ask for a referral to a speech-language pathologist to ensure that there are no physical reasons for choking and then ask for techniques that can ensure safe swallowing.

Distractions

People with HD can be easily distracted and concentration may be a problem. To help your loved one get enough calories each day, you may need to create a quiet environment for meals.

During meals:

- ▶ Turn the tv off.
- ▶ Keep conversation light — no big decisions.
- ▶ Maintain the focus on the food.
- ▶ Be encouraging about eating.

Nutrition in Late-Stage HD

Eating will become more difficult as HD progresses. In the late stages, chewing and swallowing are challenging and choking may be a constant threat. By this time, the person will no longer be feeding themselves and the caregiver may need to consider enteral feedings (tube feeding). Tube feeding may be needed as a supplement to regular diet, to provide fluids without the need to swallow, or as the sole means of calories, nutrition and hydration. The possibility of tube feeding is one that should be discussed in advance with the person with HD (if possible.) The person's wishes on the subject should be included in a legal document called an Advance Directive (see page 19 for more information.) You may want to ask a doctor or nurse to lead the initial discussion. Sometimes, once patients stop walking, they burn fewer calories and can increase weight. Ideal body weight (not too heavy, not too light) should be maintained throughout the disease, including in the late stages.

Cognitive Problems

The ability to understand information, organize thoughts and make reasoned decisions is called *cognition*. The cognitive problems caused by HD affect an individual's ability to organize and control their thoughts and actions. Changes to cognition and behavior can be among the earliest symptoms of HD. They may even appear before the abnormal movements.

The Cognitive Disorder

Symptoms include difficulties with:

- ▶ Multi-tasking, attention and concentration.
- ▶ Impulse control.
- ▶ Irritability/angry outbursts.
- ▶ Short term memory/new learning.
- ▶ Beginning/ending activities.
- ▶ Obsessing on a thought or idea.

Attention and Concentration

Huntington's disease interferes with the ability to concentrate and prioritize. People with HD will have difficulty performing a sequence of tasks, especially if they are distracted by noise or activity. If you ask individuals with HD to do several things at once, they will have a hard time organizing them in order of importance. They may skip important steps or forget tasks entirely.

This can be frustrating for both you and the person with HD. Try to remember that these are symptoms of the disease and that the best thing to do is to adopt coping strategies.

To improve attention and understanding:

- ▶ Reduce noise and other distractions when thinking is required.
- ▶ Avoid the need to multitask — do one thing at a time.
- ▶ Give directions in short sentences containing 1 to 2 pieces of information.
- ▶ Break tasks into small steps and write them out.
- ▶ Offer limited choices and avoid open-ended questions. For example, ask "Do you want beef or chicken for dinner?" instead of "What do you want to eat?"
- ▶ Adopt simple "to-do" lists and use a calendar or appointment book.

- ▶ Plan activities so that the same type of thing happens at the same time every day.
- ▶ Regular routines can help patients and caregivers by removing the reliance on memory.

Memory and New Learning

Huntington's disease affects short-term memory and the ability to learn and remember new information and skills. A person with HD may seem to learn something new, then spontaneously forget it. This can be more pronounced in children and young people with HD, who are still learning important skills. In addition, there can be a slowness of recall of information.

New Learning

Here are some strategies for introducing new skills to a person with HD:

- ▶ Break the skill into simple steps.
- ▶ Allow plenty of time for practice.
- ▶ Repeat the learning over several days.
- ▶ Use something, like a checklist, to show when each step is complete.
- ▶ With adaptive equipment, such as a walker, wheelchair or special seating, try to introduce it before it is absolutely needed, to allow time for learning and relearning.

Fixed Ideas and Obsessions

Another symptom the caregiver has to manage is the tendency for the person with HD to get caught up with one idea or activity. This may take the form of:

- ▶ Worrying endlessly about one thing.
- ▶ Thinking and talking about one subject.
- ▶ Performing the same action over and over.
- ▶ Believing something is true when it is not, despite reassurance.
- ▶ Because these are symptoms of the disease, the caretaker cannot simply tell the person with HD to "stop it." There are ways to modify the behavior and, if those do not work, there may be medications the doctor can prescribe to help manage the symptoms.

Working with Obsessions

To reduce the impact of fixed ideas and obsessions, try these strategies:

- ▶ Try not to "dismiss" the concern. Attempt to reassure them.
- ▶ Ask the doctor to discuss the concern, if needed. Sometimes a non-family member can get better results.
- ▶ Establish routines — e.g., locking the door and leaving the house the same way each time. This will allow you to reassure the person that the task has been completed.
- ▶ Set fixed time limits — i.e., scheduling a certain time and number of minutes to talk about the topic each day.
- ▶ Ask for a referral to an expert in obsessive compulsive disorders (OCD) or delusions.

Lack of Awareness of Symptoms

People with HD sometimes do not notice their own symptoms. They may notice the fact that they are dropping things or falling, but they may not notice the chorea or other symptom that is causing the incidents. They may experience arguments with people at work, but not see that their emotional outbursts are creating the problem. The science around this is not certain, but the current thinking is that this happens because changes in the brain prevent them from consciously recognizing their movements or behavior as abnormal. There can also be psychological denial, but not recognizing symptoms is more common.

Working Around Unawareness

When dealing with unawareness, it may be wise to:

- ▶ Focus on what happens, like falling, and talk about prevention, for example, "This rug seems to be a tripping hazard. Why don't we remove it?"
- ▶ Remember that the person with HD may not experience the changes to their abilities the way that you do. It may not be possible (or necessary) for you to convince them of their disabilities.
- ▶ Ask a medical professional for help, if possible. Sometimes information is taken more seriously when it comes from someone outside the family.

The unawareness of symptoms can be very frustrating to the caregiver, especially if you find that you are reporting symptoms to the individual's doctor and the individual is saying that "there is nothing wrong." You may have to request a neurological or cognitive test for your loved one, in order to show the doctor that you are not overreacting and that the symptoms you are reporting are real. Keep in mind that in addition to unawareness, there can also be denial of symptoms as a psychological reaction.

Caregiving and the Cognitive Issues

The cognitive issues in HD can be difficult for the caregiver to deal with. It is easier for most people to work around a physical disability, like the movement problems, than it is to accept the disorganization, forgetfulness and apathy that are also symptoms of HD. In addition to strategies such as using simple sentences, adopting checklists and calendars, etc., it may help if you:

- ▶ Remember that HD causes nerve damage in the brain.
- ▶ Understand that the person with HD has not become "lazy" and that they cannot simply "try harder."
- ▶ Accept that changes in the ability to think and organize will be frustrating to both you and your loved one.
- ▶ Stay calm when your loved one gets irritable.
- ▶ Allow extra time for just about everything.
- ▶ Be kind about your loved one's fixed thoughts and worries.
- ▶ Keep trying different strategies to help keep your loved one (and your life) on track.

Behavioral Problems

Changes in behavior and changes to cognition are often among the earliest signs of HD. These changes may be more disruptive to daily life than the movement problems. Your loved one begins to do things that seem entirely "out of character" even before the movement issues are noticeable.

Changes to behavior caused by HD may include:

- ▶ Depression
- ▶ Apathy
- ▶ Anxiety
- ▶ Impulsive behavior and poor judgment
- ▶ Emotional and temper outbursts
- ▶ Ruminative thoughts, compulsive behavior
- ▶ Irritability or agitation
- ▶ Recklessness
- ▶ Aggression

Try to keep in mind that people with HD are not choosing to lose control of their behavior — it is a symptom of the disease that must be managed. A physician may be able to recommend medications for some of the behavioral problems associated with HD.

Depression

Depression is very common in HD. You may hear people say, "Who wouldn't be depressed if they HD?" but medically, it appears that depression in HD can be caused by physical changes to the brain. Depressed people with HD typically respond very well to treatment with antidepressant medication, psychotherapy or a combination of both. There are potential side effects with all antidepressants, so the choice of treatment should be carefully discussed with the physician or mental health provider.

Apathy

Some people with HD will become apathetic — unmotivated and uninterested in work and in daily life. They just don't seem to care. This can be frustrating to the caregiver, who may feel that their loved one is simply being "lazy," although this is not the case. Loss of motivation and the ability to start

activities are symptoms of the brain disorder caused by HD. Often, the person with HD will participate in an activity if someone else starts it and keeps the energy up. Scheduling activities to occur the same day each week or at the same time every day may help. Sometimes, it may be easier to allow the person with HD to simply relax and watch TV.

Is It Apathy or Depression?

Symptoms of apathy and depression are similar. If a person with HD stops doing the activities they used to do, ask them about their mood — are they feeling sad or guilty, even suicidal? Those are serious signs of depression and should be discussed with the doctor. Some medications can also cause apathy.

Anxiety

Changes to cognition caused by HD can make it hard to “keep things straight” in the mind of the person with the disease. This feeling of disorientation can cause extreme anxiety. People with HD may become agitated and anxious over simple tasks or events. You may be able to reduce anxiety in the life of a person with HD if you can:

- ▶ Establish predictable routines for daily life.
- ▶ Make life less complex. Sometimes handing off a job that has become too difficult will significantly decrease anxiety in the person with HD. Break up even simple tasks into smaller parts to make it easier to do.
- ▶ Pay attention to what makes your loved one anxious — some individuals worry less when they know in advance about changes to their routines; others do better when they are told right before the event happens.
- ▶ Talk to the doctor if anxiety becomes overwhelming or disruptive. It may be a sign of depression or other psychiatric condition. The doctor may recommend counseling or medication to relieve the problem.

Impulse Control

Reduced impulse control is a very frustrating aspect of HD for the caregiver. Symptoms of HD can lead to thoughtless decisions, recklessness, temper outbursts and inappropriate sexual behavior. Some of these behaviors can be dangerous, both to the person with HD and to others.

As a caregiver, you may need to:

- ▶ Set predictable routines for daily living, which can help you to manage your loved one’s activities.
- ▶ Take over management of finances and restrict access to money.
- ▶ Keep a watchful eye on who your loved one spends time with — they may be easily influenced by peers to make poor decisions.
- ▶ Get rid of weapons and dangerous objects, or at least, put them under lock and key.
- ▶ Watch for developing obsessions with things like smoking, caffeine or even Internet use and keep the doctor up to date on unusual behavior.

None of this will be easy. Previously independent adults may not be willing to give up control over their activities, particularly if they are unaware of their symptoms. You may need to negotiate changes over time. Ask a mental health professional to help you develop strategies for managing impulsive or reckless behavior, if needed.

Irritability and Outbursts

Irritability and emotional outbursts are common symptoms of HD that can be very upsetting to the caregiver. Your loved one may seem to “fly off the handle” over any little thing or when they don’t get their way. The disease will lesson control over emotions, similar to how it reduces impulse control.

Managing Outbursts

- ▶ Try to remain calm. Reacting emotionally may make things worse.
- ▶ Ask your loved one to stop yelling and offer other ways of getting your attention.
- ▶ Work to understand that it is the disease that is triggering the outbursts and angry words. Try not to take it personally.
- ▶ See if you can identify the situation (or person) that seems to trigger outbursts and then use that information to reduce the incidents.
- ▶ Use established routines to help you manage the irritability and outbursts of HD. They give you a reason to “stand your ground” and set reasonable limits on behavior.
- ▶ Outbursts can get out of control. If you fear violence, do not hesitate to leave or call for help. Safety first.

- ▶ Sometimes it is possible to identify circumstances that trigger outbursts in order to avoid them.
- ▶ Medication may also help. Irritability or even more severe symptoms such as delusions, violent outbursts and agitation can often be successfully treated with pharmaceuticals. Which medications are chosen will depend on the symptoms, as well as the potential side effects.

Other Causes of Behavioral Problems

Symptoms of HD are not the only reason that people with HD may become irritable or agitated. There are other possible causes, many of which can be treated. These include:

- ▶ Depression, not caused by the disease
- ▶ Frustration
- ▶ Fear
- ▶ Untreated medical conditions
- ▶ Physical discomfort
- ▶ Visual or hearing impairments
- ▶ Reactions to medication
- ▶ Dehydration
- ▶ Hunger
- ▶ Fatigue
- ▶ Financial concerns

Emotional Outbursts and Caregiving

The emotional outbursts caused by HD are among the most disruptive symptoms of the disease. Your loved ones may not consciously want to hurt your feelings or annoy you. They may not even realize how hurtful their words are. Often, they will forget about their explosive behavior as soon as it is over. This is because the loss of emotional control is a symptom of the disease.

Managing your loved one's emotional outbursts will be an ongoing job that requires emotional toughness and flexibility on your part. Consider finding a support group, therapist or counselor for yourself, so that you have an outlet for your own frustrations and feelings.

Avoiding a Crisis

While it is difficult to imagine, the loss of emotional control caused by HD may put you and your family in danger. Simple disagreements have been known to escalate out of control. It is important that you prepare for this possibility, however remote. We suggest that you create a file that contains items you might need in a crisis situation.

This crisis management file may include:

- ▶ Phone numbers for the doctor or psychiatrist.
- ▶ A one-page psychiatric and medical history summary.
- ▶ A copy of insurance card or policy information.
- ▶ A recent picture and description.
- ▶ A current list of medications (and past medications, if known).
- ▶ A copy of the criteria for emergency evaluations and civil commitments in your jurisdiction (a doctor, psychiatrist or social worker can help you get this information).
- ▶ If possible, get your loved one to sign a medical release form that allows you to see his or her medical information.

If a Crisis Occurs...

If behavior by the person with HD becomes seriously aggressive or violent:

- ▶ Do not hesitate to call for help or leave the house.
- ▶ Decide on a "safe room" (with a locking door) in the house where you can retreat until help arrives.
- ▶ If you call 911, be sure to explain that the person with HD is not in control of their actions.

Navigating the Transitions

As HD progresses, many things will change, big and small. Over time, your loved one will need to change what they do and how they do it.

These changes will include:

- ▶ Decision-making (finances, etc.)
- ▶ Working
- ▶ Driving
- ▶ Household chores
- ▶ Social life
- ▶ Personal grooming
- ▶ Eating

The change from independence to needing care and supervision may occur over a long period of time. HD can progress over two decades or more. There will be periods when things appear to be settled and times when things will seem to suddenly change.

It is important to know that you are not alone in facing the challenges of HD. There are HD support groups across the nation and online, where you can discuss the challenges you face with other HD caregivers — people who truly understand what you are experiencing and who will support your efforts with suggestions from their own experiences. A list of organizations offering support to HD caregivers is listed in the Resources section of this guide.

Working Successfully with Medical Professionals

As the caregiver, you are the eyes and ears for the medical team who may see your loved one infrequently and only for a short time each visit.

You are the one to see the symptoms of the disease on a daily basis and you will probably be the first one to notice changes in cognition and behavior. If doctors are not familiar with these aspects of HD, they may not be looking for these changes. In order to get the best possible care for your loved one, you may be the one who alerts the doctors to new symptoms.

Conveying the cognitive and behavioral issues to the medical team may not be easy. Doctors may not see the changes to reasoning and behavior that you do and you may be told you are “overreacting” to events. You may feel embarrassed to discuss changes in behavior with your loved one present. You may face many challenges.

There are things you can do that will help you to inform the medical team about the symptoms you see in your loved one. These include:

- ▶ **Become informed about HD.** The better you understand the symptoms, the better you can explain them to the doctors.
- ▶ **Keep track of symptoms.** When something occurs that looks like a symptom of HD, note the date and what happened in a private diary or notebook. Having a record of incidents will help you keep the doctors informed. Include symptoms such as:
 - Impulsive behavior or recklessness.
 - Forgetfulness or confusion.
 - Emotional outbursts or aggression.
 - Obsession or fixed ideas.
 - Apathy or depression.
- ▶ **Ask for a private consultation with the doctor.** If you feel you cannot speak freely in front of your loved one, ask the doctor for some time alone. Bring a summary of what you have observed and give it to the doctor. Talk to your doctors to see if email can be used to communicate about issues without involving your loved one.

- ▶ **Keep a record of any medications** your loved one has been prescribed, along with any side effects you have noticed.
- ▶ **Write down your questions and concerns** as they come up, so that you don't forget anything important you want to talk about with the doctor.
- ▶ **Ask the doctor to include the changes you have noted in their office notes and visit record.** These may be helpful if you later request a neurological, cognitive or psychiatric evaluation.
- ▶ **At the end of the consultation, repeat what you think the doctor said and write down any action plans**—including changes to medications, recommendations for evaluations, exercise plans, etc. This will make it easy to go over results at the next appointment.

Keeping Track — the Care Notebook

A Care Notebook is a way to organize all of your loved one's medical information. Using a Care Notebook will make it easier to find and share important information with people who are part of the healthcare team.

A Care Notebook may be a three-ring binder, with dividers and plastic sleeves or it may be a file box or accordion file. The form doesn't matter. What is important is that you keep all the important information in one place.

Bring the Care Notebook to all medical appointments. As discussed above, try to have your questions written down in advance and leave room to jot down the doctor's response.

Make sure that a family member or someone else can locate the Notebook in an emergency and knows to bring it to the clinic or hospital.



Contents of a Care Notebook

Contact Information

- ▶ You and family members
- ▶ Voluntary caregivers
- ▶ Paid caregivers or agency
- ▶ Emergency personnel
- ▶ Hospitals
- ▶ Healthcare providers
- ▶ Therapists
- ▶ Pharmacy
- ▶ Insurance company or a copy of the insurance card
- ▶ Special transportation
- ▶ Equipment providers

Pages for Incidents

- ▶ Medication effects
- ▶ Behavior episodes
- ▶ Memory problems
- ▶ Sleep problems
- ▶ Activities/exercise
- ▶ Falls or balance problems

An Appointment Log

- ▶ Who the appointment is with
- ▶ The reason for the appointment
- ▶ The results of the appointment
- ▶ Any follow-up action that you need to take
- ▶ Questions for upcoming appointments

Medication Log

- ▶ Name of the medication
- ▶ Date the medication was first prescribed
- ▶ Who prescribed the medication
- ▶ Dose/timing/special instructions
- ▶ Date the medication was stopped and why
- ▶ Treatment results
- ▶ Test results
- ▶ Hospitalizations

Calendar

Copy of the Advance Directive, if one exists

Copy of the Durable Power of Attorney for Health Care, if one exists

Caring for the Caregiver — Respite and Support

Caring for a person with HD can be very stressful and tiring. The disease takes a very uneven course and you never know when symptoms will change. Some of the symptoms are difficult to manage.

Changing responsibilities also put pressure on the caregiver. You may be caring for someone who used to provide support to you. You may have to make decisions that previously would have been shared. Caregiving in HD can seem overwhelming.

Caregiver Burnout

Are you overwhelmed by caregiving? Signs of exhaustion and “burnout” may include:

- ▶ Being constantly tired, sad or depressed.
- ▶ Feeling helpless and alone.
- ▶ Eating more or less than usual.
- ▶ Feeling distracted or “spacey.”
- ▶ Skipping favorite activities or turning down invitations.
- ▶ Drinking alcohol to relax or deal with pressure.
- ▶ Thinking about death.
- ▶ Losing interest in caregiving.

To give your loved one the best possible care, it is important that you take care of yourself. This means finding enough outside help and support to allow you time to do things that give you pleasure and relaxation. Denying yourself the right to rest and recharge will not improve the care of your loved one, it will only add to “burnout” and make you a less effective, compassionate and loving caregiver.

Respite Care

Temporary, short-term care (also called respite care) may come from:

- ▶ Family and friends who can stay with the person with HD for a few hours or take them on outings, so that you get “time off.”
- ▶ Professional respite organizations or elder care referral services.
- ▶ Community or senior centers offering elder “daycare.”

To find respite care in your area, talk to your doctor, social worker or caregivers in your HD support group. Alzheimer’s organizations are also a good source of information on respite care facilities that can serve individuals with brain disorders, such as HD. Many insurance companies have policies and may cover respite care for you.

Choosing a Source of Respite Care

When looking into professional or community sources of respite care, you may want to ask about:

- ▶ Hours and fees.
- ▶ Types of programs for people with dementia and brain disorders.
- ▶ Staff training in dementia.
- ▶ Planning for emergency situations.
- ▶ Meals, specifically meals for people with choking problems.
- ▶ Transportation options.

Use the Power of “Yes”

When people offer to help you — say YES!

“Yes, you can pick up something at the grocery store for me.”

“Yes, you can come and sit with Joan while I go for a walk.”

“Yes, I would love to have you drop off a meal.”

Accept the offer of help as a generous gift from someone who cares about you. No one expects you to do it all.

Joining a Support Group

HD is not a common disease and you will meet many people who have no idea what the disease is about and what you do as a caretaker. It can be a great relief to have the opportunity to share your feelings and experiences with people who have faced the same challenges. A caregiver or HD support group can be your lifeline.

There are support groups that meet in person and those that communicate online. You may wish to join both types of groups. In-person groups typically meet once a month, while you can have an online chat with another caregiver almost any time of the day or night. What is important is that you **DO** talk to other caregivers and not try to bear this all alone. Plus, what you have learned as a caregiver may be of tremendous help to someone else.

Sources for Support Groups

The Huntington's Disease Society of America (HDSA) sponsors over 140 HD support groups nationwide. You can locate the nearest support group on the HDSA website (www.hdsa.org) in the section, "Living with Huntington's Disease."

WE MOVE sponsors an online discussion forum, (www.wemove.org/ubb/ultimatebb.php) designed for the free exchange of information and ideas among various movement disorder communities, including patients, families, and caregivers.

There are many other online HD support groups. You may have to try several before you find the right community for your needs. Begin your investigation by typing "HD Support Group" into a search engine, such as Google, Ask or Yahoo.

When you find a group to join, take a few visits to just read and learn what the group is about, before you jump in. Taking a moment to listen and learn will assure that you will be welcomed as a valuable new member of the group.

Finally — use your common sense. Remember that you cannot believe everything you read on the Internet. Individuals in a support group may propose treatments that are not backed by science. Use the support group to help you with your caregiving, not to replace the doctor's advice.

Preparing for Late-Stage HD

No one wants to think about end of life issues, but making decisions and completing paperwork will make it possible to provide your loved one with loving compassionate care until the end of life.

Some important end of life issues that will need to be discussed include:

- ▶ The need for skilled nursing/nursing home care.
- ▶ Living Wills and Advance Directives.
- ▶ Medical Power of Attorney/Health Care Proxy.
- ▶ Tube feeding and hydration.
- ▶ Hospice care.

Remember to take care of these issues for yourself as well. Medical issues can happen to any of us at any time. You should consider back-up plans to make sure the care and finances of your loved one are addressed if you are unable.

Out-of-Home Care

With the help of family and friends, a person with HD may be able to live at home for many years. However, as symptoms become more severe, home care may become too difficult for the caregiver. Sometimes it is the physical disabilities that become too much for the caregiver to manage. Other times, increasing behavioral problems make it impossible for the individual to remain at home.

Thinking about nursing home placement is often difficult for caregivers, who may feel that they "should" be able to take care of a loved one at home. However, if the goal of care is to maintain safety and comfort for the person with HD, professional care may be the most reasonable solution to a very difficult situation.

Finding a skilled nursing facility that can care for a person with HD may not be easy. People in the late stage of HD need help with eating, walking and communicating and may also require psychiatric care, occupational therapy, physical therapy and speech therapy. We recommend that you begin to look into nursing homes long before placement is actually needed. You may be able to get good information and suggestions on selecting a skilled nursing facility from caregivers in a support group or in an online HD chat room.

Paying for skilled nursing care is always a concern for caregivers. Medicare may be available to pay for part of the cost, if the individual has been declared disabled for more than two years or is over the age of 65. Even under the best of circumstances, Medicare pays for only a portion of skilled nursing care. Other funding sources for long-term care may be Medicaid, supplemental health insurance (often called “Medigap” insurance) or private long-term care insurance.

The first step may be to talk to someone at your local Medicare office or visit the Medicare website, www.medicare.gov, and then talk to your insurance agent.

Living Wills and Advance Directive

Your loved one should be given every chance to express their wishes about end of life care. A living will or Advance Directive for Health Care is a legal document that explains the level of care a person wants at the end of life. Do they want every effort made to extend life, even if it includes tube feeding or heart/lung support? Do they wish to withhold antibiotics or nutrition at a certain point? It will be very helpful to have your loved one’s wishes in writing, to support your decisions in difficult times. You may want to have a doctor or nurse start the conversation.

Each state has a slightly different Advance Directive form. Your doctor’s office should be able to assist you in getting the correct form for your state.

Medical Power of Attorney

Another important legal document is the Medical Power of Attorney or Health Care Proxy. This document allows the person with HD to decide who will make important medical decisions for them when they no longer can. This document must be signed by the person with HD and signed by witnesses. You may need to talk to a lawyer about completing a medical power of attorney form.

Tube Feeding and Hydration

As discussed before, tube feeding and hydration may need to be considered in the late stages of HD. People often have strong feelings one way or the other about tube feeding. If possible, find out what the person with HD feels about the issue and have their wishes written into an Advance Directive that is signed and witnessed.

Hospice Care

Hospice care emphasizes palliative rather than curative care and can be part of care anytime during a life-threatening illness. Although we think about end-of-life care, because the definition of hospice care was recently revised, hospice care can be initiated whenever needed. When the time is near, every family wants their loved one’s final days to be as comfortable and easy as possible. Hospice care works to relieve physical, emotional and spiritual distress in both the patient and the family. It can be provided at home or in a skilled nursing facility. Medicare and most private insurance cover hospice care.

Hospice is important because many doctors are uncomfortable dealing with end of life and may even be unhelpful. Hospice workers are trained to help individuals and their families during this difficult time. Hospice care is not about giving up hope; it is about providing the most compassionate, loving care to your loved one.

The American Medical Association (AMA) recommends the National Hospice and Palliative Care Organization (NHPCA), (www.nhpco.org) for information on hospice care, as well as state-specific Advance Care forms.

Hope for the Future

Every day, researchers around the world work tirelessly to find a cure for HD. Clinical trials and observational studies are essential to learning about the disease and to testing possible new treatments for effectiveness. If anyone in your family circle — with HD, presymptomatic, or at-risk — would like to learn more about participating in an HD research study, there is information available at ClinicalTrials.gov (www.clinicaltrials.gov), a service of the National Institutes of Health. Simply click on “Search for Clinical Trials” and enter “Huntington’s Disease” or visit the HD SA web site at www.hdsa.org

Resources

Family Caregiver Alliance (FCA)

www.caregiver.org

National Family Caregivers Association (NFCA)

www.thefamilycaregiver.org

National Alliance for Caregiving

www.caregiving.org

Huntington's Disease Society of America

505 Eighth Avenue, Suite 902

New York, NY 10018

Tel: 212-242-1968 or 800-345-HDSA (4372)

www.hdsa.org

Huntington Society of Canada

151 Frederick St, Suite 400

Kitchener, ON N2H 2M2

Tel: 519-749-7063 or 800-998-7398

www.huntingtonsociety.ca

National Institutes of Health

9000 Rockville Pike

Bethesda, Maryland 20892

www.health.nih.gov/topic/HuntingtonsDisease

The National Institutes of Health offer comprehensive information on Huntington's Disease and related disorders. The website includes links to other government agencies, including:

- National Institute of Neurological Disorders and Stroke (NINDS). See Enfermedad de Huntington below for Spanish.
- National Human Genome Research Institute (NHGRI)
- National Library of Medicine (NLM)

Medline Plus

www.nlm.nih.gov/medlineplus/huntingtonsdisease.html

Enfermedad de Huntington

www.espanol.ninds.nih.gov/trastornos/Enfermedad_de_Huntington.htm

Este panfleto fue escrito y publicado por el Instituto Nacional de Trastornos Neurológicos y Accidentes Cerebrovasculares (NINDS), el patrocinador principal en los Estados Unidos de la investigación sobre trastornos del cerebro y el sistema nervioso, inclusive la enfermedad de Huntington.

WE MOVE

5731 Mosholu Avenue

Bronx, NY 10471

www.wemove.org

www.mdvu.org (for medical professionals)

WE MOVE has made every effort to present information about Huntington's disease that is up-to-date and accurate. The material provided has undergone medical review. However, medical science is constantly changing. Therefore, WE MOVE does not warrant the information in this text is complete, nor is WE MOVE responsible for omissions or errors in the text or for the results of the use of this information. The information provided does not replace consultation with your personal physician or other medical professional. All medical procedures, medications, indications, and contraindications should be discussed with your personal physician.



www.hdsa.org



www.wemove.org