A Carer's Guide for Huntington Disease

Huntington Society of Canada
HUNTINGTON
Société Huntington du Canada

A Carer's Guide
for Huntington Disease

Editors, Canadian Edition:

Angèle Bénard RSW, Director of Family Services and Community Development, HSC

Jana Papke Family Services and Research Lead, HSC

Designer:

Tiffany Spencer Communications Coordinator, HSC

Disclaimer

Statements and opinions expressed in this book are not necessarily those of the Huntington Society of Canada (HSC). HSC does not promote, endorse, or recommend any treatment or therapy mentioned herein. The lay reader should consult a physician or other appropriate health care professional regarding any advice, treatment or therapy set forth in this book.

Note of Appreciation to Jim Pollard

HSC would like to extend its sincere appreciation to Jim Pollard for his permission to update the information in this resource.

Note of Appreciation to Dr. Alan Fung and Dr. Tiago Mestre

HSC would like to extend its sincere appreciation to Dr. Alan Fung and Dr. Tiago Mestre for their review of this resource.

Note of Appreciation for Canadian Reviewers

HSC would like to extend its sincere appreciation to the Family Services team members for their assistance in updating the content of this resource.

The Huntington Society of Canada would like to extend its appreciation to the

Jack and Barbara Hay Foundation

for their support in helping to produce this resource.

Huntington Society of Canada
HUNTINGTON
Société Huntington du Canada

Printed in Canada

Table of Contents

Preface:

A Carer's Guide for Huntington Disease

A Carer's Guide for Huntington Disease is adapted and abridged from the original *A Caregiver's Handbook for Advanced-Stage Huntington Disease* originally authored and edited by Jim Pollard, MA, with contributions by a number of professional experts, published in 1999. The book was also reprinted by the Huntington Society of Canada (HSC) in 2003/2011 with permission. A special "thank you" to Jim Pollard, MA, for his permission to revise and update this resource for our current use.

For those of you who are familiar with the original publication, you will note that we have updated the title of the document and refer to "carers" rather than caregivers. Carers Canada defines a "carer" as "a person who takes on an unpaid caring role for someone who needs help because of a physical or cognitive condition, an injury or a chronic life-limiting illness" (www.carerscanada.ca/carer-facts). While the focus of this book is on carers in an unpaid role (and the word "carers" will be used throughout this book), the term is still viewed as synonymous with caregivers or care partners for the purposes of our examples used in the book. While some material may be more suited to carers (unpaid), we believe that the content and strategies offered can also be applicable to paid persons who provide assistance and care for others.

The guide provides a brief summary of some of the topics that carers need to consider when working with a person who has Huntington disease (HD) and also refers to additional resources that can be obtained to gain more in-depth information as needed. The HSC has many fact sheets which are typically 2-3 pages and are updated regularly and available both online and in print. The fact sheets cover a variety of topics which provide practical tips and suggestions. Personal stories are also used throughout the book to bring some of the topics "to life" with relatable examples.

While some specific technical information is provided for certain topics, the Huntington Society of Canada strongly recommends that you consult with a healthcare professional - Neurologist, Speech Language Pathologist (SLP), Occupational Therapist (OT), Physiotherapist (PT), Registered Social Worker (RSW), Registered Dietician (RD), Registered Nurse (RN) and/or Psychiatrist (to name some of the professionals you may consult) as each person's needs will be different and unique.

Many thanks to the team of professionals who took time to edit and provide information updates and technical content for this current guide including Dr. Alan Fung and Dr. Tiago Mestre. Also, thanks to the Family Services team of the Huntington Society of Canada for their editing and revision of the document.

An Overview of HD

Huntington disease (HD) is a neurodegenerative disease. Cells in the central part of the brain known as the basal ganglia die. Since so much of the brain's activity passes through this area, the death of these cells affects virtually everything about a person—including movements, emotions, and thinking processes. However, the lack of obvious signs that a person has HD (until the later stages of the disease) may make it more challenging for carers to remember that changes that are seen in a person over time are a result of the disease and not necessarily other factors.

HD is a genetic disease that a person gets by inheriting the gene for HD from a parent. Genes are paired units of hereditary information found inside each cell in the body that direct our growth, development and function. We inherit one member of each gene pair from each parent. Genes are made up of DNA, and DNA molecules consist of four bases, known as C (cytosine), A (adenine), G (guanine) and T (thymine). The gene that causes Huntington disease is called the HD gene. There is a region in the HD gene in which a sequence of three bases – cytosine, adenine and guanine (CAG) is repeated many times. For individuals with Huntington disease, the CAG sequence has increased (expanded or mutated) into a range considered abnormal or disease-causing. An HD gene expansion is passed on in families in a dominant manner (which means that both male and female children of a parent with the gene for HD have a 50% chance to inherit the expansion).

Genetic testing involves the examination of an individual's DNA, which is obtained from a blood sample. The process for genetic testing (ie. length of time and steps involved) will vary slightly between different regions in Canada. Testing is done in a specialized laboratory to determine the number of CAG repeats in both copies of the HD gene. As everyone has two copies of the HD gene, it is common for a person to have two different CAG repeat sizes (one inherited from their mother and one inherited from their father). The number of CAG repeats determines whether a person is gene-positive or gene-negative. Test results for Huntington disease can be complex, and it is recommended that they are reviewed with a genetics specialist in order to fully understand what it means for an individual and his/her family.

RESOURCES

For more information, please consult HSC Fact Sheets on *Genetic Testing and HD* and *Genetic Discrimination* at www.hdfactsheets.ca.

As a genetic disease, HD is referred to as "a disease of families." In many families touched by HD, more than one family member may have HD at the same time. Many relatives are at-risk of later developing the disease. Nearly every member of the family has been a carer to a parent, brother, sister, son, daughter, aunt, uncle, grandparent, or grandchild at one time or another.

Similarly, the person in your care most likely saw a parent suffer with HD, which can cause concern about symptoms that he/she saw and found distressing. A person may also have fears about passing on the disease to children or learning that siblings will also get the disease. Families may struggle to care for a loved one at home for as long as possible. The family is very knowledgeable about the

person's preferences and knows how to meet care needs. For a healthcare professional, this family knowledge can be very helpful. However, it is important to keep in mind that at-risk relatives will be confronting the disease that could become their reality - every time they visit the person with HD.

Although people can first exhibit signs of HD at any age, most people first show symptoms when they are between the ages of 35 to 55. Thus, HD is described as an "adult-onset disease." In the prime of life, one may be diagnosed with HD and learn that any children will be at-risk of developing HD also.

As a progressive disease, HD begins very subtly and only the person with HD, close friends or relatives, and the trained eye of a physician can detect its earliest signs. It progresses in stages, slowly advancing for many years. In the final stages of the disease, the affected person will need help with household chores and personal care. As a carer, you can help by anticipating changes in function that may trigger new concerns and preparing in advance for each new set of challenges that you and the person for whom you are caring will encounter.

Juvenile HD

There is a rare, juvenile form of HD which manifests before the age of 20. Approximately 10% of all HD diagnoses are Juvenile HD (JHD). JHD looks different from the adult version. Usually the person is stiff, rigid and slow, may have involuntary jerking movements, and may experience seizures. Juvenile HD progresses more rapidly than adult-onset HD.

Late-Onset HD

In addition, there is a late-onset form of HD which involves a diagnosis after age 60. Knowledge of the typical age of onset (ages 35 to 55) sometimes leads physicians to miss the diagnosis, because doctors incorrectly believe the person is too old to develop HD. Late onset consists of about 10% of all HD diagnoses.

HD Mask

As Huntington disease progresses, the disease places a mask on the person who is suffering from HD. This is sometimes referred to as the Huntington's Disguise or HD Mask (as noted in Jim Pollard's book, *Hurry Up and Wait!*).

The HD Mask makes it difficult to "read" the person's emotions and impossible to know how a person is truly feeling. The weakness and changes in facial muscles can create an appearance of boredom or disinterest. Cognitive changes also contribute to difficulty with organizing thoughts and delayed responses to questions. These combined physical and cognitive features are what make up the Huntington's Disguise (sometimes also referred to as "flat affect".)

It is important for carers and friends and family to see behind the disguise and continue to interact as in the past. The person you once knew is still the same person and needs your help to be seen beyond the Huntington's Disguise.

Symptoms of HD

Health care professionals look at HD as a disease made up of a triad of symptoms - movement (physical), cognitive (thinking), and psychiatric (emotional). Each person with HD will experience the progression and severity of symptoms in a unique way.

Some of the symptoms include the following:

- Movement (physical) symptoms: involuntary movements (chorea), diminished coordination, difficulty walking, talking and swallowing, weight loss
- Cognitive (thinking) symptoms: difficulty with focus, planning, recall of information and making decisions; impaired insight
- Psychiatric (emotional) symptoms: depression, apathy, irritability, anxiety, obsessive behaviour

Movement Changes

As the disease progresses, the movement symptoms usually become more apparent. However, some years before, it began with small changes in eye movement and involuntary movements of the fingers and face. The symptoms progress and as the involuntary movements, known as chorea, become more exaggerated, a person's gait now looks "drunk-like". Voluntary movements are also affected. This can mean an action like stretching an arm or lifting a leg can happen unexpectedly or in an untimely way. Speech becomes affected as words are slurred. As balance deteriorates, falling occurs more often, and the affected person becomes unable to walk safely without assistance.

The lack of muscle control also has an effect on the ability to speak and to swallow food. It becomes increasingly difficult for the person with HD to speak and be understood by others. Impaired muscle control affects the ability to swallow properly, and this causes an increased risk of choking, prompting the need for a special diet (minced or puréed food), and the need for assistance with eating. In addition, most individuals eventually need adapted beds and wheelchairs to accommodate their severe involuntary movements, impaired balance, and changes in posture.

Cognitive Changes

The cognitive symptoms are less apparent than the movement or physical symptoms but more disabling in many ways. Subtle changes that affected work and family began many years before. In many cases, the ability to organize and plan the work day slowly began to erode, and routine tasks, previously performed effortlessly, became more complicated to complete. As cognitive function continued to deteriorate, other people may have noticed increased inflexibility and small but significant changes in temperament. Long-term relationships may have been jeopardized because the person with HD may be unable to see changes within and may vigorously deny their existence.

Now, the person's thinking is slower, initiating action is more difficult, learning new things is not as easy as it once was, and judgement is impaired. Having difficulty waiting for things and becoming unreasonably demanding of family, friends and caregivers will present challenges for the people who are assisting in the person's daily activities.

Psychiatric Changes

The psychiatric symptoms associated with the physical changes in the brain could include anxiety, irritability, depression, apathy, perseveration, thoughts of suicide, psychosis, obsessive compulsive disorder and sexual disorders, among others. Emotional symptoms of HD can begin many years before a formal diagnosis of HD is confirmed through a neurological assessment. In the very early stages of the disease, a person who has not been diagnosed and is considered to be "at-risk" may exhibit behaviour that can present many challenges to a person's relationships, employment and finances. When these changes and behaviours begin in a person "at-risk", it is important to seek help from a physician. There are medications that can help with some of the psychiatric symptoms.

Suicide

The definition of suicide is the self-inflicted ending of a person's life on purpose. Suicide is different than Medical Assistance in Dying (MAiD) for many reasons. MAiD will be covered in the section on Advanced-Stage Medical Issues.

Suicide is a concern for people who have HD as well as other people associated with HD, including family members and carers. For people in different stages of the disease, depression, combined with a lack of impulse control (i.e. impulsive behaviour), make suicide a major risk at various times. Depressed people who have feelings of hopelessness and who are prone to act impulsively because of the disease, may be at greater risk of carrying out a suicide attempt.

Other symptoms associated with depression, including insomnia (difficulty sleeping) can contribute to someone considering suicide as an option. The Canadian Mental Health Association (CMHA) states, "People who think about and attempt suicide don't want to die; they want a way out of their deep, psychological pain. They describe feelings of being overwhelmed, being stuck, of not being able to see a way out."

CMHA cites the following warning signs of suicide:

- Change in outlook on life, for example, if someone feels hopeless and like a burden
- Talk about killing oneself and having a specific plan is a major warning sign – Call 911!
- Change in mood, from happy to sad or even from sad to happy, and anything in-between
- Increase in drinking alcohol or taking drugs

If you are concerned about someone, talk with the person. According to the CMHA, talking with a person is critical in changing the outcome and doesn't put the idea of suicide in a person's mind; it is a relief to be able to discuss it. If a loved one shares with you that he or she is having thoughts of suicide and has a plan to carry it out, take the comments seriously and call 911 or your local crisis hotline, or take the person immediately to the emergency department of your local hospital.

RESOURCES

For more information on suicide, visit www.suicideinfo.ca/resources, www.cmha.ca/news/you-can-help-someone-who-is-thinking-about-suicide or read the Depression and HD fact sheet at www.hdfactsheets.ca.

Communication in HD

Keeping Communication in Perspective

The person with HD who enters a long-term care facility or who is in your care at home has already experienced a tremendous number of losses. The ability to drive a car, manage finances and communicate easily with family and friends has been lost or diminished. As the disease progresses, more losses will occur. If this person saw a parent or another relative in the later stages of HD, he/she has an idea of what may lie ahead in the disease progression. It is understandable why anger, depression and lack of communication might be happening.

For the person with HD to express feelings and needs is a struggle. There are many ways we exert some degree of control over our surroundings. However, HD has entangled every one of them. Difficulty in speaking makes it hard to clearly state wishes and needs. Anticipating the future and planning any activity is hindered by the inability to organize information. Undertaking even the simplest movement is hampered by trouble getting started. If carers remember the struggle, they will understand that every time a person freely chooses how he/she would like the eggs cooked, it is a significant victory in a tremendous struggle to control his/her world.

Speaking and Listening

Communicating with one another in any relationship is very important. With HD, this becomes more and more important as it becomes more and more difficult. The movement disorder affects speech in several ways. Often in the mid-stages of HD, people lose precision in making sounds, control of the volume of the sounds they make, and coordination of the speech and breathing mechanisms. This creates speech that is varied in volume, interrupted by grunting or breathing sounds, and hard to understand. In the most advanced stages, people express their range of needs and emotions with a few intelligible words or sounds.

Cognitive symptoms affect the content of what is said. The ability to form ideas, organize thoughts and present them in an orderly sequence decreases in HD. It can mean that a person with HD may have difficulty starting a conversation, staying on the topic, or switching from one topic to another. A person may get stuck on one topic and have difficulty getting off it.

As clear speech becomes more difficult, it takes great effort for a person with HD to carry on a conversation, and he/she may rely on a very small vocabulary of more easily understood words. This allows you to take on a more active role in a conversation, picking up on those key words, anticipating the idea, and expanding on it to keep the conversation going.

At the point where it is extremely difficult to be understood, some people simply stop talking. Your familiarity with a person's likes, dislikes, career, interests, hobbies, and relatives will keep the conversation going or allow you to become his/her "interpreter" with others.

It can be humiliating and frustrating for the person with HD, and embarrassing for you, when you

have difficulty understanding the words. One way to show respect is to put the burden of understanding firmly on you. Ask for clarification and ask for permission: "Do you mind if I repeat your words to you from time to time so you will know how I'm doing?"

Sometimes, tools can be used to help with the communication process. Tablets and other electronic devices as well as communication boards are commonly introduced to people who are having difficulty being understood. As well-intended as they may be, these devices are not often adopted by people with HD as an alternative form of communication. Speaking is still easier than learning to use the unfamiliar tools or board. A low-tech option for communication is the simple use of hand gestures. Using a "thumbs up" (meaning a yes response) or "thumbs down" (meaning no) approach for simple closed-ended questions enables the person with HD to more easily convey an answer. As with other adaptive devices, introducing the communication device early, before it is actually needed, gives the user more time to learn how to use it, practice with it, grow fluent in its use, and possibly adopt it.

Some families find it helpful to assemble a book or picture album full of photographs that represent the person's interests, hobbies, family, career, and preferences. Since non-family caregivers may first meet the person when he/she has difficulty speaking or recalling events from the past, the album serves two purposes. First, the album is a communication aid which allows him/her or you to point to pictures when you don't understand each other.

Second, it serves as a treasury of interests, children, grandchildren, relatives, hobbies, achievements, pets, home or apartment, and favourite sports teams so you can better know who this person is. Simply knowing favourite sports teams can be the basis for many conversations in your years together.

Please remember that a person who has HD can comprehend our speech and understand all that's going on around him/her to a far greater degree than most people realize. The level of communication through spoken words is not an accurate indication of how well he/she understands what you say.

Family members and caregivers agree that a person, even in the most advanced stages of HD, still manages to communicate with carers very effectively through facial expressions, eye gazes, and other subtle movements. Getting to know the person will help with communication.

RESOURCES

The Huntington Society of Canada has fact sheets on *Communication and HD* and *Tips When Working with Individuals Affected by HD* which provide strategies for communicating effectively. Find these fact sheets at www.hdfactsheets.ca.

Eating and Nutrition

Strategies for Weight Loss Prevention

Providing adequate nutrition can be the single greatest clinical issue in caring for a person with HD. Maintaining body weight will be a constant challenge. It is estimated that some people with HD, particularly in the more advanced stages, require a diet of up to 5000 calories a day just to maintain their weight. It is recommended that a registered dietician be consulted to assist with weight maintenance questions.

In long-term care facilities, nutritionists should consider double and triple portions for people with HD. Take requests for more food or supplements seriously.

Constant hunger can make it difficult to wait for lunch and dinner. Perception of time may be altered. It may be helpful to serve five or more "mini-meals" throughout the day and while the person is awake at night. Having snacks available and helping the person stay well hydrated may prevent constant hunger and may help to minimize gulping. Another strategy is to increase caloric intake by creating a diet of high-calorie foods.

If each mouthful of food is so difficult to chew and swallow, then maximizing the number of calories in each bite can only help. Unfortunately, the involuntary movements that may knock food to the floor, the swallowing disorder, and the great concentration needed to chew safely complicate getting those calories into the digestive tract.

People often supplement their meals with high-calorie drinks. These commercially manufactured supplements (e.g. Boost, Ensure) are commonly used in long-term care facilities and are readily available for purchase by consumers. Family and professional caregivers have cleverly invented homemade "super-calorie" foods to quickly boost calorie intake at a single meal. High-protein, high-calorie powders that add calories to shakes, puddings and other foods can be used to make regular foods much more calorie-laden.

About Swallowing

Swallowing is a very complex activity. It involves coordinating the opening and closing of the mouth and lips and chewing while inhaling and exhaling. Food needs to be mixed with saliva, moved to the back of the tongue, and sent on its way down the esophagus by the swallow reflex. A person who has HD and is in the later stages of the disease is at serious risk of choking and aspirating.

Preventing these problems in advanced HD is an ongoing challenge to a carer. Stuffing too much food into the mouth, gasping for air, gulping liquids, and poorly coordinating the complex movements needed to bite, chew, move, and swallow food increase the likelihood that food will unintentionally be aspirated. A speech language pathologist (SLP) can make recommendations regarding positioning of the person, texture of food, and other issues that will make swallowing easier. It is important to

consult with the professionals to learn proper techniques and strategies for making eating as enjoyable and safe as possible.

Proper positioning assures that the person is comfortable, reduces involuntary movements, inhibits reflexes, and accommodates any postural changes caused by dystonia. A "chin-tuck" manoeuvre can help to direct food toward the esophagus. Sitting upright with support for the head and neck can help to avoid the hyperextension of the neck that increases the risk of choking.

As a general rule, thicker and colder liquids are easier to swallow. Thin liquids are the most difficult because they are virtually impossible to control within the mouth. Water may be particularly dangerous!

However, liquids from coffee to orange juice to soft drinks can be combined with commercially available thickeners, which change the texture without significantly changing the taste. Drinking through a straw nearly always makes it easier to swallow liquids, especially thin ones, by limiting the amount taken at a time and by directing it to the back of the mouth. Check the length of the straw; one that is too long can injure the back of the throat or cause choking.

There are many different styles of "sport" bottles, cups, and mugs available. Many of them are insulated to keep drinks hot or cold and have flexible straws attached. Since they have been designed to facilitate drinking liquids in a moving car or while engaged in outdoor athletic activity, many of them have grips that make them easier to hold, straws or "sippy" spouts that guide the liquid to the mouth, and covers that prevent spills. Many people with HD find one that is particularly effective and comfortable and carry it with them throughout the day. Cups with spout-style covers are also available in medical supply stores.

It's a common safety practice to ask the person you're helping to do a "dry swallow" (that is, a swallow with no food or liquid in the mouth), after each time food is swallowed. Pay close attention to food temperature; many people with HD have an altered sense of temperature and may burn their tongues or mouths on hot foods. Sometimes a person with HD will "stuff" food by placing more food than he/she can possibly chew and swallow into the mouth as quickly as possible. This behaviour greatly increases the risk of choking and aspiration and should be discouraged. Providing or feeding a person with a teaspoon will encourage small amounts per mouthful.

It's a difficult period when chorea progresses to a degree that the person with HD isn't regularly getting enough food into the mouth for adequate nutrition. Attempts to intervene and help the person may be seen as a final loss and a symbol of dependence on others for eating. By assisting with small parts of the meal earlier than is needed, he/she may become accustomed to your help and be more willing to accept it when it is absolutely necessary for safety and nutrition. For example, spoon-feeding a thick shake at the end of a tiresome meal or placing a few pieces of a snack into his/her mouth at various intervals throughout the day may gradually increase acceptance of this assistance.

Coughing, Choking and Aspiration Pneumonia

If you've helped someone with a swallowing disorder to eat, you know that it is often a difficult task for both of you. You might recall him/her coughing after swallowing a mouthful of food and waiting through that tense moment to ensure there is no choking. Coughing during a meal is not a routine part of eating. Coughing is a defensive reflex to prevent choking. When coughing episodes begin to occur, carers should ask for a professional assessment by a Speech Language Pathologist (SLP). If an assessment is done early and a baseline is established by the SLP, it will be easier to monitor the progression of swallowing difficulties and address any changing needs through regular re-testing.

Choking is a very serious risk factor. Most people with HD develop a swallowing disorder, or "dysphagia", at some point in the course of their disease. Often the first sign is a serious unanticipated choking episode. Individuals with swallowing problems need to have their temperature and lung sounds monitored regularly for signs of pneumonia.

Learn the proper technique (through a CPR/First Aid course) so you'll be prepared to respond to a choking incident. Make sure everyone who assists this person to eat is practiced in the technique. It may be reassuring to explain or demonstrate it if there has been a previous choking incident. Listen very carefully to the instructions you are given on how to help this person eat, and ensure that the appropriate chair and recommended adapted utensils are used. Take no shortcuts; take your time. Check for proper positioning every time you put food in the mouth. Eliminate as many distractions in the room as you can. Double-check the texture of the food that's been specially prepared and ensure that certain liquids are thickened.

Remember, this person may be very hungry and very tired and want to race through the meal. Take your time for safety's sake. If eating takes too long or is too tiring, arrange to have him/her eat less food more often throughout the day.

A Note About Feeding Tubes

As swallowing becomes increasingly difficult, eating by mouth may not provide enough calories and nutrition to sustain a person. At this point, some people with HD may choose to receive their nutrition through a feeding tube (called a gastrostomy tube). It is most helpful if the decision about feeding tubes can be made well in advance, with the assistance and support of family members.

Although it is a relatively minor surgical procedure, placement of a feeding tube has greater implications than simply increasing calorie intake and enhancing nutrition. Deciding whether or not to have a feeding tube forces the individual and the family to confront difficult emotional or spiritual issues about extending life and quality of life. These are very personal decisions, and your understanding and support are needed.

RESOURCES

Find fact sheets on *Eating and Swallowing* and *Feeding Tubes* at www.hdfactsheets.ca.

Balance and Issues with Falling

As HD progresses, changes in muscle tone cause some people to have asymmetrical posturing in their trunk, arms, or legs. This creates the appearance that they're leaning forward, backwards or to one side. The normal, unconscious reflexes to prevent falling become slower. It gets increasingly difficult to keep from falling or to avoid injury during a fall. Walking is more and more difficult as balance becomes progressively impaired. Although it may appear that the involuntary movements cause falls, research has shown that this may not be the case. Instead, falling is more likely to occur as people with HD develop stiffness and rigidity and as their balance deteriorates. Consult a physician, physiotherapist (PT) or occupational therapist (OT) for strategies to help prevent falls and to learn the best ways for helping a person to move around in the day (from bed, to chair, to toilet).

Consider footwear when trying to prevent falls. As strength in certain foot muscles decreases, sensory changes in the feet may also be taking place. This can lead to abnormal foot placement when walking and, consequently, tripping. Consult the physiotherapist (PT) or occupational therapist (OT) for recommendations on the best type of shoes to wear. Shoes in poor repair can cause falls, so check them regularly. Loose shoelaces are also a tripping hazard. A special type of elasticized shoe lace (called a "spring lace") can help with shoe tying to minimize the possibility of tripping.

As careful as we are, falls may be inevitable. Even minor falls can lead to cuts that require stitches, or cause painful bruises, broken bones, or injury to the brain. Sudden changes in movement, mood, thinking, or neurological function could be a warning sign of an injury to the brain, even if the changes only develop a few days after the fall. If you see these kinds of changes in someone who has fallen, notify a physician immediately and request an examination.

Fatigue will also play a role in the tendency for a person to fall. Since "routine tasks" take more effort and time, this can be very tiring and will make a person more fatigued earlier in the day. Keep this in mind for fall prevention, and rearrange the daily routine to better fit the person's needs.

A Note About Equipment

There are many devices, aids and pieces of equipment that can help a person maintain independence and dignity while coping with this disease. Consult an OT or PT for help in determining what will work best for the situation.

Sometimes a soft head protector, made of leather bands with a Velcro chin strap, can be used for head protection.

Wearing protectors (e.g. padded mitts, knee and elbow pads used by athletes) for the joints may help to prevent traumatic joint swelling that can happen from falls and can also protect against injuries sustained from severe involuntary movements.

A physiotherapist (PT) or occupational therapist (OT) can provide the best advice on whether a walker will be suitable for a person's abilities and can also assist with the decision around a wheelchair.

The importance of choosing the right type of wheelchair cannot be overstated. Selecting an appropriate wheelchair prolongs mobility, conserves energy, and allows the individual with HD to do many activities without help.

As the disease progresses, the selection of an appropriate bed becomes very important. Consult an OT or PT for suggestions.

RESOURCES

HSC has a fact sheet on *Home Safety* which includes tips for preventing falls and strategies to keep people safe at www.hdfactsheets.ca.

SUSAN'S STORY

When Susan first came to her long-term care home about three years ago, she had significant balance problems from her HD and had seriously injured herself falling. However, Susan was determined to continue walking alone! Susan's gait and strength improved for a while but, given the severity of her problems, it was inevitable that she would soon need assistance walking.

Physiotherapists began a daily program of teaching her how to use a walker before she absolutely needed one. Susan completed her "workout" every day as her "coaches" would encourage her to practise her balance exercises. After a few falls, the physiotherapists were able to encourage use of the walker on a full-time basis. Already familiar with it, Susan accepted it readily, and the number of falls decreased over the next six months.

As soon as Susan accepted the walker, her coaches started to teach her how to use a wheelchair as part of her daily workout, long before she actually needed it. She especially enjoyed doing "wheelies" when visitors were watching. Eighteen months later, Susan began to fall frequently in the evening. When it was suggested that she use the wheelchair just at supper time and in the evening, Susan agreed. Again, she had far fewer falls.

Progressively introducing assistive devices earlier than actually needed prevents their introduction from becoming a symbol of yet another loss of function. The same principle can be used with eating assistance as well as wearing adult incontinence pads.

Exercise and Fitness

As the disease progresses, the individual with HD will decline in health and lead a more sedentary lifestyle. Although the disease process can't be altered, a routine exercise program can help to address all areas of decline and help the person maintain strength, improve balance and posture, and feel more in control of his/her body. With aerobic activity such as pedalling a stationary bike, it is possible to improve breathing, which in turn helps with breath control for talking and eating. Improvement in deep breathing can help maintain ability to cough effectively, which in turn helps prevent choking and aspiration pneumonia. People who regularly exercise are able to clear secretions more efficiently when they do have colds or pneumonia.

RESOURCES

Consult a physiotherapist (PT) for the most appropriate exercises and refer to the HSC fact sheet *Exercise and HD* at hdfactsheets.ca. Also refer to the *Physiotherapy Clinical Guidelines (EHDN)* and the *Occupational Therapy for People with Huntington's Disease: Best Practice Guidelines*.

ROBERT'S STORY

Robert reluctantly came to a long-term care home because he was no longer able to live alone and had no family members who could help him. He looked depressed and undernourished and avoided interaction with staff and other residents. His depression was treated with medication and counselling, and in several weeks his mood, appearance, and nutrition were all improved. Every day a "coach" from the physiotherapy staff visited Robert and chatted briefly with him in his room. When the coach learned that Robert, though not particularly athletic, enjoyed bicycle riding, she invited him to the physiotherapy "gymnasium" to ride the stationary bike. After three more weeks of the coach's daily visits, Robert rode the bike in the gym. After two visits to the gym, he agreed to a physiotherapy evaluation.

He had no significant chorea but his gait was affected by difficulty with balance. His poor posture, due to weak upper back muscles, and his lack of endurance compromised his heart health.

After eight weeks of "workouts" in the gym, Robert achieved his therapeutic goals as well as his personal goal to ride the stationary bike for twenty minutes without shortness of breath. He smiled, talked with fellow residents, and began to participate in other therapeutic groups. He was a changed man! Formally discharged from physiotherapy, he continued his daily workout in a group exercise program. Three months later his strength, balance, gait, and respiratory status were all improved.

Personal Hygiene

Any kind of daily hygiene like brushing our teeth involves a specific sequence of steps. Once these steps are learned, they become so automatic in our day that we forget that we are even planning and organizing before we do these tasks. We take the steps for granted. These are the very skills that present problems as HD progresses. The cognitive symptoms of HD can present more problems with these activities of daily living and personal care - than the movement symptoms.

Although this may not affect some people, others may react with a decreased interest in self-care. If this is the case, the carer can help to establish a self-care routine and anticipate potential problems. Set clear expectations. Give no more assistance than needed to start or finish an activity. The disinterest is driven by the disease, not a lack of concern about appearance or hygiene. Providing extra help and support in a low-key fashion can be very beneficial. There are other areas of personal hygiene which can provide challenges for the person with HD and the carer.

Bathing

People with HD may be reluctant to bathe in a tub or shower. There are many hard surfaces, protruding fixtures and close quarters to be considered. It may have been the site of previous falls. Standing naked in front of a carer is uncomfortable. Perhaps the feeling of water being splashed is unpleasant.

For these reasons, it's important to keep the shower or bath as brief as possible. Using a shower chair allows people with HD to focus all their energy and attention on bathing and not on balancing in the shower. Hand-held shower heads allow you to aim the water stream exactly where it's needed, minimizing the movement required and cutting down on the splash when it's held close to the body. People who have difficulty holding onto soap, facecloths, and sponges may still be able to lather up with bath mitts that require no grasping and that fit right over the hand.

Toileting

Because of the involuntary movements and problems with balance, extra care needs to be taken during the toileting routine. "Flopping down" onto toilet fixtures can loosen the hardware that holds the seat to the bowl, as well as the wax seal and fittings that hold it firmly to the floor. This could create a safety hazard. Men who stand to urinate may have difficulty keeping their urine stream aimed in the bowl. Loose seals and urine on the floor contribute to odour problems. Floors wet from urine or water are a falling hazard. Padded toilet seats may be helpful since they cushion the impact of "flopping down" onto the seat, and the padded cover may protect a person's back and the toilet tank as well. Grab bars can also be installed to increase safety.

RESOURCES

HSC has a fact sheet on *Home Safety* which includes tips on safety in the bathroom and general home safety strategies at www.hdfactsheets.ca.

Dressing

Clothes should be easy to get off and on and durable enough to withstand many washings. Clothes need to be loose enough to accommodate movements that are extraordinary in their range and frequency. Track suits are often a good clothing choice for those who have difficulty with buttons or zippers. Choosing patterned or dark colour shirts will help to hide spills and stains. Fashionable patterned scarves may catch drool as effectively as institutional bibs. Many people with HD feel hot all the time, so they may even feel comfortable wearing light cotton clothes in the wintertime.

It is important to allow the person to complete as much of the dressing routine as possible without offering assistance. Providing more assistance than is actually needed robs the person of the independence that he/she still has and creates "learned helplessness" in the person with HD over time.

Sometimes one particular carer or family member has a good rapport with the person and can influence the person with HD to do things that no one else can. Respecting this preference by allowing that carer to work with the person as often as possible can be a comfort to everyone involved.

Dental Care

Having someone else put a toothbrush or anything else in your mouth is an uncomfortable experience. When you can't hold your head still and have difficulty speaking fluently, it's frightening—even more so when that person is someone unfamiliar to you. Anxiety can intensify chorea and make it more difficult to help. Providing dental care can be a comfortable activity if the person needing help and the helper take a few moments to relax together and to position themselves in such a way that the helper can gently stabilize the head.

The importance of oral and dental care increases as the disease progresses into its more advanced stages. A person may breathe in his/her own saliva. Effectively cleaning the mouth minimizes the bacteria that can be aspirated and reduces the risk of infection. Sometimes dipping the toothbrush in mouthwash rather than toothpaste is preferred since it can be difficult to spit out toothpaste. Cleaning the teeth and mouth should be done after main meals and, most importantly, at bedtime.

HD presents a series of unique problems related to dental care. Due to involuntary movement and changes of muscle tone in the mouth, "bruxism" or teeth grinding is quite common. Sometimes people regurgitate food. Over time, stomach acid can damage tooth enamel, weakening the teeth and leaving them susceptible to breakage or decay. Assessing pain in the mouth or teeth can be difficult when the person with HD can't communicate clearly, but it is important.

RESOURCES

The Huntington Society of Canada has a fact sheet on *Dental Care* at www.hdfactsheets.

Understanding Cognitive Changes

We all have bad days! However, when you're trying your hardest to give a person the best possible care in very challenging circumstances, it's difficult not to take it all personally when that person is distracted, angry or generally unco-operative. As unpleasant as it may be to care for someone behaving this way, never forget that the problems you're facing are caused by Huntington disease. The behaviour that comes from changes in his/her brain is the problem... for both of you.

A critical component of your care is to look carefully at the person's actions and try to determine their cause. Often, what is labelled as "inappropriate behaviour" is an attempt by the individual, through great impediments caused by the disease, to express his/her needs or preferences. This is why we like to refer to this as "responsive behaviour". A person is responding in the only way that he/she is able to - in that moment in time. It may take time to begin to understand the needs and preferences of the person for whom you are caring.

RESOURCES

Find tips in our fact sheets *Responsive Behaviours, Tips When Working with Individuals Affected by HD, Cognitive Changes and Communication in HD* at www.hdfactsheets.ca and in the *Understanding Behaviour in HD guide.*

STEVE'S STORY

Steve has HD and lives in a long term care facility. For several days in a row, he threw his breakfast tray onto the floor every morning. The staff caring for him saw his "agitation" and attributed it to HD. The nurse reported Steve's actions to his physician, who ordered an antipsychotic drug for the "agitation". A staff member who knew Steve well and knew how much Steve loved his coffee, suggested that there might be another reason for Steve's behaviour. Further investigation showed that Steve had a complaint that he couldn't express verbally: his coffee was cold. Once Steve was given fresh hot coffee, the problem was resolved without medication.

If carers understand that "inappropriate behaviour" (referred to as responsive behaviour) is often an attempt to express needs or preferences, the needs of the person with HD can be met quickly by addressing the complaint.

Here are some of the ways that changes in the brain affect the person in your care. By understanding these changes, you may be able to "read" needs and preferences of the person more accurately, and find new ways to do the things he/she wants to do, despite the losses.

Slower Thinking

People in the more advanced stages of HD no longer think and process information as quickly as they once did. Simply put, there are fewer healthy neurons available to process information. This often causes a delay in responding to your requests, questions, or comments. In fact, you may learn

that there is a consistent predictable lag of several seconds before a response comes. You may ask, "Would you like to go shopping today?" Five seconds later you've still not received an answer.

But ten seconds from when you asked, the person may say, "Yes!" Too often carers mistake the delayed response to mean "No!" No response may not mean "No!" Allow more time than usual for a response. Once you've recognized a delay in responding, you will be able to wait more easily.

Despite all the challenges that the cognitive disorder presents, people want to continue to care for themselves, dress themselves, bathe themselves, and eat independently. When they don't respond or do it as quickly as we do, there is often an urgency on our part to do it for them. By understanding the cognitive deficits and anticipating processing delays, you can wait for the person to respond and enable participation in his/her own care.

RICK'S STORY

Rick wakes up and sits on the side of his bed. "Good morning, Rick! Breakfast is ready." Five minutes later, he is still sitting there. "Rick, it's time to get up and eat breakfast!" Another five minutes pass, and Rick hasn't moved. You approach him, hand him a towel and his toothbrush, motion toward the bathroom door and say, "Here you go, Rick, start by getting cleaned up." A short time later, Rick has come out of the bathroom and is ready to get dressed for the day.

Sometimes initiating an activity—just getting it started—is very difficult. Just like Rick did, a person may need a "jump start" from you. Do the first few steps of an activity with - or for - the person, and often, he/she completes the rest of it without your help. By allowing a person to complete it on his/her own, you are actively helping the person maintain independence.

Difficulty Learning

There is a myth that a person with HD cannot learn new information. If a person has learned your name and can find his/her own room, the myth has been disproved! As HD progresses, it is certainly true that learning new information becomes progressively more difficult. If a person tends to learn by doing, it may take many more repetitions or opportunities to learn. If a person uses trial and error, he/she may not learn from mistakes the first time.

Because learning can be more difficult, it's helpful to keep your instructions and directions as specific as possible. For example, saying, "Please hang your coat up in the closet" is more easily understood than, "Please put your clothes away."

Difficulty Organizing Action

As discussed in the personal hygiene section, many of the activities we engage in every day involve long sequences of smaller activities or steps. Choosing clothes, putting on underwear, socks, shirt, pants and a sweater, buckling a belt, zipping zippers, and buttoning shirts are all parts of dressing. These sequences of activity become "second nature", and we don't even think about them when we

can do them. Unconsciously, we've organized the information and actions required to complete them. A person may have difficulty organizing these sequences of activity at some point in the course of his/her HD. This may explain why a person might wear a blouse over a sweater, misbutton a shirt, or wear no socks. Writing lists of the steps involved in lengthy or complex activities may be helpful. You may list in order the steps required to get dressed and tape them near the closet. Posting schedules of daily activities and the time to do them may help organize the day.

Need for Routine and Consistency

Although we take it for granted, there is great comfort in knowing what's going to happen next. That comfort comes from consistency.

A consistent sequence of events or "routine" enables many people in the more advanced stages of HD to go about their daily activities without disruption, with greater independence, and in good spirits. Consistency comes from doing the same thing, in the same order, at the same time, in the same way, each and every day. When today's events are the same as yesterday's events and those of the day before, it's easy to know what's next in the day. This routine helps to build trust between the person and carer, minimizes distractions that can disrupt daily activity, and makes it easier for success!

In care facilities, where there are shifts and many staff changes three times every day, sometimes it is difficult to deliver care as consistently as we would like. Consider posting daily schedules in the resident's room and providing specific notations in the chart about the daily schedule, noting the importance of consistency and routine to him/her. If it's possible, ask that a specific team of two or three staff members be assigned to look after your loved one and help them learn the routine. Whatever routine you establish, be sure it's easy to follow.

A Lack of Self-Awareness and Lack of Insight

One of the elements of HD is a lack of self-awareness and insight. This can take many forms. An example is a person with HD who has severe chorea and is in a wheelchair, trying to get up and walk but falling down continuously.

This behaviour may actually be due to a lack of self-awareness, an inability to accurately perceive oneself. This is different than the psychological mechanism of denial – when a person won't acknowledge something.

Someone with readily apparent chorea will often tell you that he/she is unaware of it! The reality of the symptoms of the disease is something that he/she can't see.

Poor Judgment

Carers are often concerned when they see someone with HD using poor judgment. They often become involved in "power struggles" as they try to dissuade a person from doing something which seems to be risky or not wise.

You need to know when to "back off". As difficult as it may be, if an individual's poor judgment does not hurt anyone, you might consider allowing the person to continue. Perhaps eventually, the person will come to understand your point of view.

TONYA'S STORY

Tonya has had HD for eleven years and now lives in long term care. Tonya has always loved the outdoors and often enjoys sitting outside in the sun on pleasant days and listening to the birds singing. One cold and very windy afternoon, Tonya decided that she wanted to go outside and read her favourite magazines. A staff member cautioned Tonya and said, "It's too cold and windy to sit outside today. You won't be able to read your magazines because of the wind. You will have to wait for a nicer day to sit outside!"

Unfazed by the staff member's weather report, Tonya began to gather her magazines and proceed towards the door. The staff person told Tonya again that it was just too cold and miserable outside to sit and read. Tonya insisted, "I don't care; I just want to sit outside in the fresh air".

The verbal exchange continued until another staff person heard the escalating commotion. This staff member didn't believe it was worth getting into an argument with Tonya about the poor weather outside. She told the other staff member that she would work it out with Tonya.

"Let me get the door for you, Tonya", said the staff person. Once outside the door, Tonya looked at the staff member and said, "It's really cold and windy today. I don't want to sit outside!" By allowing the person to follow through on her idea, the staff member avoided a serious confrontation and no harm was done.

Difficulty Waiting

An experienced carer observed, "A person with advanced HD can't wait." As absolute as that sounds, it's based on a sensitive observation of the difficulty a person with HD has when struggling to control impulses.

When a person wants something, he/she wants it now. The demands are driven by the damage to the brain caused by the disease. Due to the impaired ability to control impulses, he/she just can't wait.

If someone asks for your assistance, provide it as soon as practically possible. As disruptive as it may be to you, it will be more efficient for you in the long run. If you're unable to assist right away, try to set a specific time when you will realistically be available to help. For example, you might say, "I'll do that for you in fifteen minutes at four o'clock." Be sure to keep your promise! Asking a person with HD to wait is asking him/her to do something that may be neurologically impossible. Always make the effort to anticipate what is needed and eliminate the wait!

Challenges and Misunderstandings

Sometimes involuntary movements by a person with HD can be mistaken as deliberate or aggressive actions when they are actually a direct result of the disease. Carers may think a person with HD is "playing mind games" with them and "being difficult" on purpose. An example of this is a person continually dropping a spoon while eating. This is actually due to a physical symptom known as "motor impersistence" which is an inability to maintain a position. The person is unable to continuously grasp the spoon, so the spoon falls out of the hand repeatedly.

Another poorly-understood part of the movement symptoms is an inability to control the force of one's movement. If a carer asks a person to lift his/her arm, a person could unintentionally move the arm in a forceful way which appears as a hitting motion towards the carer.

When carers understand these actions are not within a person's control, they can avoid being in the way of big bursts of movement, and minimize being annoyed, or emotionally or physically hurt by these actions.

"Getting Stuck"

It is common for a person in the mid and advanced stages to "lock onto" and "get stuck" on a topic. It's extremely difficult for him/her to stop "getting stuck", or "perseverating". A few principles can help you manage repetitive or compulsive behaviours.

- Once the routine or rules are established, stick to them. If different carers respond differently to repetitive demands, it is confusing to the person.
- Don't promise to do something "in a minute" if you know that you can't keep the promise. If you do make a promise, keep it.
- Keep a schedule and remind the person frequently what time it is and what is happening next.
- Make sure that you meet some of the requests. There may be limited ways for the person with late-stage HD to feel good or to be happy, and to deny those pleasures because of "bad behaviour" or your busy schedule is not good care.

When someone is stuck on a topic, avoid saying "No" to them. A refusal can needlessly cause anger. For example, Robert routinely has a snack at 10:15 every morning. "Can I have my snack?" he asks at 10:00. Rather than telling him, "No, it's not time yet," it may be helpful to suggest, "Yes, you can. In 15 minutes I'm going to give you a snack. Go get yourself settled in the room!"

RESOURCES

For more information on strategies to help with perseveration, review the fact sheet on *Tips When Working with Individuals Affected by HD* at www.hdfactsheets.ca.

Understanding Changes in Mood

Changes in mood, like depression, irritability, anger, anxiety or other emotions should be monitored by a psychiatrist or other healthcare professional.

Depression

Depression is a symptom of HD which can occur during the course of the disease. Depression and other psychiatric symptoms are among the most treatable features of HD, responding well to medication, and in some cases, counselling and talk therapy. Some of the more typical signs of depression can also be attributed to the movement and cognitive disorders of HD, so they are often overlooked. For example, a lack of interest, initiative, and concentration may appear to be signs of cognitive decline as well as classic signs of depression. Changes in sleep or appetite, a sad facial expression, irritability, and general slowing of activity could be due to the physical changes brought on by HD or due to possible depression.

Even when people are very physically debilitated in advanced HD, depressive symptoms can respond to antidepressant medication. The following signs of depression should be watched for, and if noticed, should be reported to healthcare professionals immediately:

- lack of initiative
- lack of interest
- irritability
- sad facial expression
- isolation and social withdrawal
- change in sleep pattern, sleeping more or less than usual, difficulty falling asleep, waking up very early in the morning, or waking up several times during the night
- expressions of guilt
- expressions of hopelessness and helplessness
- lack of energy
- lack of concentration
- restlessness or inability to sit still
- general slowing of activity
- talking about or attempting suicide

RESOURCES

For more information on *Depression and HD*, please refer to our fact sheet at www.hdfactsheets.ca.

SANDRA'S STORY

Sandra was in the late stages of HD and as the disease progressed, she had become non-verbal. Sandra's mood appeared to have changed, and she did not appear to be very happy. The family was concerned that Sandra was depressed and wanted to find out if there was anything that could be done to assist her. During one visit to the neurologist, the doctor asked Sandra, "Are you feeling depressed?" Although Sandra was not able to verbally communicate her answer, the answer came. Tears began to flow down Sandra's face, and the doctor was able to confirm with this non-verbal cue, that Sandra was feeling depressed. Appropriate medication to treat the depression was prescribed, and Sandra began to show signs of improvement in her mood.

This example highlights the importance of looking out for signs of depression and advocating for treatment. The example also shows that, despite the challenges with verbally communicating with others, Sandra understood the question being asked, and she was able to respond without speaking any words. It serves as a reminder that, in the late stages of the disease, a person with HD can understand what is being said to – and about him/her – even if he/she is unable to verbally respond.

Irritability and Angry Outbursts

One of the most challenging and common concerns for families affected by HD is irritability and angry outbursts from the person with HD. There could be several explanations for the irritability or outbursts. Irritability may stem from frustration with the loss of certain capabilities, inability to communicate, boredom, pain or unexpected changes in routine. An angry outburst may be caused by the loss of impulse control – which is also part of the disease. This can happen very quickly and is sometimes referred to as having a "short fuse". Irritability and explosive outbursts could also occur because of an unmet need like hunger or thirst, which is a common symptom of HD. The outbursts can be extreme and frightening to the people who witness them or are the target of the confrontation.

If an angry outburst occurs, this is the time to give him/her space and protect yourself and those nearby. Do not attempt to reason, explain or persuade. Since a person with HD is often unaware of his/her symptoms and lacks insight, attempts to change the behaviour at this point generally do not work. Allow the person time to cool off. Once calmness has returned, try to figure out what triggered the outburst so that it can be avoided in the future. If carers learn what the triggers or causes are for the outbursts, it may be possible to prevent some of them and also avoid premature or excessive use of medications to treat the symptoms.

People who watch and wait to intervene, learn that a person with HD often "cools down" as quickly as he/she "heats up". When it's over, you need not be surprised if the person apologizes to you, explains that, despite a great effort on his/her part, he/she lost control. Accept this most sincere apology. This is yet another loss for the individual. He/she may feel guilty and remorseful but is not able to prevent the angry outbursts from happening.

Anxiety/Worries

Some people with HD will have anxiety. This may be the result of physical changes in the brain or because of challenging life circumstances or a combination of both. This can take the form of social anxiety in which a person is fearful of going out in public because he/she is self-conscious about involuntary movements and others' reactions. He/she may be worried for days in advance about what to wear to an event, or a person might become anxious if an unexpected guest comes to visit. Carers can simplify life and establish predictable routines which can often help to ease anxiety for the person with HD. Sometimes anxiety will not be helped with these measures, and after proper assessment of the condition and underlying factors, a psychiatrist may be able to prescribe medication to treat anxiety.

Obsessive Compulsive Disorder

Obsessions are recurrent, intrusive thoughts or impulses, and a compulsion is a repetitive performance of a routine task, like hand washing or turning off the stove. Sometimes, obsessions and compulsions are related. If a person is obsessed with germs, the compulsion of hand washing may follow. People with HD can become obsessed or preoccupied with an idea and/or participate in an activity repeatedly. If these actions interfere with a person's quality of life, it may be necessary to seek assistance from a psychiatrist.

Perseveration

Perseveration means that a person becomes "stuck" or fixed on a specific thought or action and will have difficulty letting go or moving on from those thoughts. It is a result of a combination of reduced mental and emotional flexibility brought on by the disease. Perseveration is a common symptom for people who have HD. Families are encouraged to become educated on ways to distract or diffuse situations, and some medications may help with easing perseveration. Some additional information and strategies for carers are found on page 21 – "Getting Stuck" in this guide.

RESOURCES

Find tips in our fact sheets *Responsive Behaviours in HD* and *Tips When Working with Individuals Affected by HD* at www.hdfactsheets.ca and in the *Understanding Behaviour in HD guide* and in the *Physician's Guide*

Advanced-Stage Medical Issues

Sleep and Sleeplessness

Some people have difficulty sleeping. A change in one's sleep pattern, much more of it or much less of it, may be a sign of a psychiatric symptom that should be evaluated by a psychiatrist or other healthcare professional. Medications may be prescribed to address this issue.

Sometimes people with HD will experience day and night reversal which means being awake at night and sleeping during the day. Due to daytime fatigue in advanced HD, some people may accidentally fall into a cycle of napping during the day and then being unable to fall asleep at night. Strategies may be needed to find the right balance between conserving energy during the day and being tired enough to sleep through the night – to help the person get back on the regular sleep patterns.

Excessive Sweating, Temperature and Thirst

People with HD may be more comfortable in surroundings that are cooler than typical, perhaps as low as 18º Celsius. This may be related to some aspect of the disease that affects metabolism. They may also have episodes of excessive sweating. In some cases, this may be related to certain medications. Others may have a compelling thirst. Those who drink an excessive amount of liquid per day should consult a physician regarding potential problems with electrolyte imbalance and kidney function.

Frequent Urination and Constipation

As muscles become progressively uncoordinated, emptying of the bladder will become more difficult. Increased thirst may lead to increased fluid consumption. This often causes the sensation of needing to urinate more often than usual. Problems controlling impulses, coupled with the increased urges to urinate, often lead to increased demands to go to the bathroom. Accommodate these requests as often as necessary. Sometimes urges happen so quickly that a person is unable to make it to the toilet in time. The use of adult briefs may help to alleviate the embarrassment of accidents. Introducing the use of briefs before they become absolutely essential may make the transition easier.

Constipation is a common problem in the more advanced stages of HD. Filling up on high-calorie low-fibre foods to keep weight on, the loss of some fibre in altered texture diets, and an increasingly sedentary lifestyle can all add to the problem. After a thorough assessment, constipation is often treated with increased fluids, more frequent position changes, and a regimen of stool softeners.

High Fevers

Late in the progression of the disease, a very small number of people experience recurring high fevers, at times reaching 40º Celsius and higher. As in other times of high fever, the person's level of activity will decline. Consult your physician immediately. These high fevers occur despite physicians' best efforts to identify infectious causes. As you work together with the doctor, pay close attention to room temperature and how much fluid is being consumed. Some medications interfere with sweating and the regulation of body temperature and may actually be the cause of the high fever.

Contractures

A contracture is a permanent shortening of a muscle that causes a deformity with or without pain. Providing frequent changes in position and range of motion exercises is important to prevent contractures. The participation of a physiotherapist in care is critical to prevent serious progressive deformity. In the advanced stages of HD, ability to control movement becomes severely compromised. Those who once had involuntary movements may now be rigid and vulnerable to developing contractures. Even though he/she may still have involuntary movements, he/she cannot change position. The fluctuations in muscle tone and the involuntary movements make it difficult to prevent and manage contractures. Consult with a professional for treatment options.

Chorea

Many physicians and physiotherapists familiar with HD tend not to treat chorea until people with HD can't function well on a daily basis. Many people with HD who have taken medication to suppress their chorea feel that it is easier to live with chorea than with the side effects of the medication used to suppress it. It is common for chorea to improve (lessen in severity) in the later stages of the disease.

There are, however, people whose chorea is so severe that it actually causes them bodily harm. In these cases, medication is most helpful. In addition, carefully selected padding of the environment is required. It may even become necessary to pad parts of the body if they are being repeatedly injured. Padded mitts as well as knee and elbow pads (often used by athletes) as well as socks and lamb and sheep skins can be used.

Palliative Care and End of Life Care

According to Canadian Virtual Hospice, "Palliative care is a type of health care for patients and families facing life-limiting illness. Palliative care helps patients to achieve the best possible quality of life right up until the end of life. Palliative care is sometimes considered end-of-life care, with a main focus on comfort. However, it is increasingly recognized that a palliative approach, as part of health care is beneficial early on in serious and chronic illness."

In Canada and around the world, quality palliative care:

- focuses on the concerns of patients and their families;
- pays close attention to physical symptoms such as pain, nausea, loss of appetite and confusion;
- considers the emotional and spiritual concerns of patients and families;
- ensures that care is respectful and supportive of patient dignity;
- respects the social and cultural needs of patients and families;
- uses a team approach that may include volunteers, social workers and spiritual leaders in addition to medical staff.

Medical Assistance in Dying (MAiD)

In June, 2016, the Government of Canada passed legislation that allows eligible Canadian adults to request medical assistance in dying. There is a process to follow and criteria which must be met in order to receive this procedure, and each case is carefully reviewed.

There are two types of medical assistance in dying available to Canadians. The first type must include a physician or nurse practitioner who directly administers a substance that causes death, such as an injection of a drug. This is becoming known as clinician-administered medical assistance in dying and was previously known as voluntary euthanasia.

The second type must include a physician or nurse practitioner who provides or prescribes a drug that the eligible person takes themselves, in order to bring about their own death. This is becoming known as self-administered medical assistance in dying and was previously known as medically assisted suicide or assisted suicide.

RESOURCES

For more information on End of Life Care, please refer to our fact sheet at www.hdfactsheets.ca OR Medical Assistance in Dying (MAiD): www.canada.ca/en/health-canada/services/medical-assistance-dying.html

For more information on palliative care visit www.chpca.net OR www.canada.ca/en/health-canada/services/palliative-care.html

Caring For You - The Carer

Caring for a person with Huntington disease challenges family carers and professional carers alike. The symptoms are ever-changing and progress slowly over many years.

It's difficult to go through a progressive disease with someone. It's also discouraging to realize that, no matter how hard you work, no matter how clever you are, and no matter how deeply you care, the course of this disease will not change.

There are many little victories to celebrate along the way, such as figuring out how to avoid or redirect a person's anger, discovering a new way to approach an old problem that finally works, or realizing that what you had worried about happening was really no big deal at all!

In the face of it all, first and foremost, you need to take care of yourself—not only for you, but for this person that you care so much about. If you are too drained, too exhausted, or too weak, you will not see the heroic struggle made every day by the person for whom you care.

Many wonderful caregivers draw great hope from their partners in this struggle. That hope charges batteries. That hope is the second wind when you need it. But without you there, in good health and in good spirit, there is no hope. Take care of yourself. You are worth it!

TAKE TIME FOR YOURSELF

In order to be able to do the work that you do for others, you need to take time for yourself and "recharge your batteries". Some people refer to this as "filling your cup". What kinds of activities will help you fill your cup? Here are some suggestions:

- staying physically active
- keeping up regular activities
- staying engaged socially
- participating in yoga and/or relaxation techniques
- going to the gym
- reading
- enjoying music
- having a bath
- sitting in the sun
- meditating

RESOURCES

For more information on caring for yourself, the carer, please refer to our fact sheet at www.hdfactsheets.ca. Support groups are also available across Canada. Information is available through the Family Services team: www.huntingtonsociety.ca/family-services-team

Final Thoughts to Leave With You

Yesterday Today and Tomorrow

There are two days in every week about which we should not worry,
Two days which should be kept free of fear and apprehension.

One of these days is YESTERDAY, with its mistakes and cares,
Its faults and blunders, its aches and pains.
YESTERDAY has passed forever beyond our control.

All the money in the world cannot bring back YESTERDAY.
We cannot undo a single act we performed;
We cannot erase a single word we said.
YESTERDAY is gone.

The other day we should not worry about is TOMORROW
With its possible adversities, its burdens, its larger promise.
TOMORROW is also beyond our immediate control.

TOMORROW, the sun will rise,
Either in splendour or behind a mask of clouds,
But it will rise.
Until it does, we have no stake in TOMORROW
For it is as yet unborn.

This leaves only one day – Today
Any person can fight the battles of one day
It's only when you or I add the burdens of those awful eternities – Yesterday and Tomorrow – that we break down.

It is not the experience of Today that drives people mad –
It is the remorse or bitterness of Yesterday and the dread of what Tomorrow may bring.
Yesterday never saw Today & Today will never see Tomorrow.
So therefore let us live but one day at a time.

Author Unknown

NOTES

Huntington Society of Canada
HUNTINGTON
Société Huntington du Canada

HSC Publication
2018

Huntington Society of Canada
20 Erb St. W., Suite 801
Waterloo, ON N2L 1T2
1-800-998-7398
info@huntingtonsociety.ca
www.huntingtonsociety.ca