Caregivers Handbook

for Advanced-Stage Huntington's Disease

Huntington's Disease Association of Ireland

CAREGIVERS HANDBOOK FOR ADVANCED STAGE HUNTINGTON'S DISEASE

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Preface

This handbook was a collaborative effort of the Huntington Society of Canada and the foundation for the care and cure of Huntington's disease. It is a collection of care principles developed by a team.

Ralph Walker and Liz Mueller originally conceived this handbook. Ralph served as the Executive Director of the Huntington Society of Canada and was a past President of the International Huntington Society. Liz Mueller was the Director the Foundation for the Care and Cure of Huntington's disease. The Foundation funded many projects aimed at educating healthcare professionals about Huntington's disease (HD). It's founder, Dennis Shea was an Irish American who had a special regard for HDAI.

The text of this handbook is a North American effort. Ralph and Liz assembled a team made up of a doctor, nurse, social worker, physiotherapist, a speech therapist, a special education teacher and nursing home administrator from across Canada and the United States. It was an honour to be on that team. All the resources for caring for people with HD on our continent were readily available to us.

As a group of healthcare professionals, most of what was to be included were lessons that we had learned from family caregivers and people with HD. For example, for years families had been telling us that people with HD had easier days if they followed an established routine. Or that fatigue had a dramatic impact on how well people can eat, balance and speak. As a team member I learned more than I contributed and made friendships lasting for many years.

I hope this collaboration complements your efforts as Irish family carers. Most of you have lived its contents from front to back. Perhaps it will clarify some aspects of HD for you. Sometimes nothing may be more important in the day to day life of a family than figuring out strategies to avoid angry outbursts or find ways to shower someone more efficiently. Until there is a cure there are only these small victories to celebrate along the way. Please take a moment to recognise your contribution in the HD challenge.

A Note on Pronouns: For the sake of simplicity, we have chosen to use the pronoun "he" to refer to patients with Huntington's disease. This is not intended to suggest that all Huntington's patients are male. Males and females have an equal chance of developing HD.

Acknowledgements

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Carol Moskowitz, RN-C, New York, NY and Martha Nance, MD, Minneapolis, MN edited the manuscript and made significant contributions to both text and tone. My son, Jimmy, managed word processing and computers, and my wife, Maureen, reviewed many drafts. Shirley Procell of Bossier City, LA contributed her prayer. Thank you, team!

A joint initiative of the Huntington Society of Canada and the Foundation for the Care and Cure of Huntington's Disease, this publication represents the final project overseen by former Executive Director of the Huntington Society of Canada, Ralph Walker. Sadly, Ralph died suddenly in Spring 2002. His determination to fill the void of information about HD has been a profoundly generous contribution that resonates around the world. The collection of educational materials produced by the Society is a national treasure graciously shared with people seeking information about HD on every continent.

This was also the final project for the Foundation for the Care and Cure of Huntington's Disease (FCCHD). In keeping with the wishes of its late founder, Dennis Shea, the Foundation ceased operation in June 1999. As Program Director of the Foundation, Liz Mueller has spent more than a decade bringing people from diverse backgrounds together to collaborate on two missions: curing HD, and improving the quality of care given to those who have HD. She's been a fine coach, confidante, collaborator, contributor, and colleague to many.

All of us who worked on this handbook are grateful to have collaborated with Liz and Ralph on this project. The Huntington Society of Canada wishes to thank The Trillium Foundation for funding the publication of this book. On a more spiritual level, our collective gratitude to the hundreds of people who suffered through Huntington's disease and taught us the lessons contained herein along the way. We pray for a cure for HD. Until there's a cure, there's only care. May our effort improve the care of those who suffer HD until that happy day! Hopefully, any day now...

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1. INTRODUCTION

WELCOME

The fact that you're reading this handbook shows a commitment to the person with Huntington's disease (HD) in your care. He is a very special person. He was probably doing well in life before HD began to show itself. Most likely, he had begun a career and family. But as his HD progressed, he faced challenges that we probably will never have to face. The list of things he has already lost to this relentless disease would force most of us to abandon all hope. He's lost his job, his friends, his independence, and his ability to care for himself. He may feel guilty that he's put his children and his grandchildren at risk for HD. And he may see his disability as a burden he has imposed on both you and his family. And now he's in your care.

You, the caregiver, are a very special person too. Whether you are in a long-term care facility or at home, the challenges of caring for this person with HD may seem daunting at times. Most health care professionals-nurse aides, nurses, social workers, rehabilitation therapists, and psychologists-finish their careers never having cared for a single person with HD. Simply by caring for one person with HD, you are about to gain a wealth of knowledge and experience. By becoming a partner in his care, by understanding how HD progresses, and by recognising how it affects his mood, thinking and movement, you can affect his life in a very positive way. Maybe to a degree far greater than anyone else you've ever had in your care. He's in good hands!

PURPOSE OF THIS HANDBOOK

The aim of this handbook is to familiarise you with HD, so that you can recognise its symptoms and know what to expect as it progresses. Included are some general principles of care and tips that other caregivers have found helpful. By anticipating the problems you'll face as the disease progresses, you'll be better prepared to find solutions for them.

Families touched by HD often say that understanding HD is the hallmark of a good caregiver. That is, if you understand that it is a disease of families, a disease of both body and mind, and a challenging disease with no cure and no easy answers, you'll do well. If you persevere through his adjustment to you, recognise and support him through his anger and his sadness, and communicate with him, then the good days will outnumber the not-so-good and you'll be an excellent caregiver... because you will really understand HD.

2. AN OVERVIEW OF HUNTINGTON'S DISEASE

Huntington's disease is a neurological disorder, a disease of the brain. 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, moods, and thinking processes. But because the damage caused by HD is only inside the brain, the person with HD may look relatively able-bodied until the later stages of the disease. Caregivers often mistakenly assume that changes in the person are due to lack of motivation, laziness or worse, and not to the disease itself.

HD is a genetic disease that you get by inheriting a defective gene for HD from one parent. Every child of a parent with the defective gene for HD has a 50/50 chance of inheriting the gene, no matter how many children that parent has. If you do not inherit the HD gene, you cannot get the disease and you cannot pass it on to your children or their descendants. If you do get the HD gene, you will eventually get the disease. HD always manifests itself if you live long enough. It never skips a generation.

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 caregiver to a parent, brother, sister, son, daughter, aunt, uncle, grandparent, or grandchild at one time or another.

Similarly, this person in your care most likely saw one of his own parents suffer with HD. Just as the winter snow was always deeper and the summer longer through a child's eyes, this horrible disease probably appeared worse from a child's perspective. Feelings of guilt or sadness may be stirred in the person with HD. He may think about how he might have already passed the gene on to his children. Or he may worry that his brothers and sisters will get HD, too.

His family may have struggled to care for him at home for as long as possible. They are very knowledgeable about his preferences, and have learned how to meet his care needs. They may be very helpful to you. When family members visit him in the nursing home, they may be silently thinking of the day when one of his children may need this same kind of care. Visits by children, brothers, and sisters can be as difficult for them as they are for the person with HD. For at-risk relatives, each visit can be another confrontation with the disease they might get themselves.

Although people can first exhibit signs of HD at any age, most people first show them when they are in their 30s and 40s. Thus, HD is described as an "adult-onset disease." In the prime of his life, he may discover that he has HD, that each of his children may have inherited the gene, and, if so, will eventually develop the disease. More than likely, he had settled on a career and was, in many ways, a well-adjusted adult. These achievements will now have to be given up, one after another, along with his plans and dreams, as HD runs its destructive course.

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. It usually takes at least 15 years for the disease to run its course, sometimes longer. Particularly during the last half of those years, the affected person will need help with household chores and personal care. As a caregiver, 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.

There is also a juvenile form of HD. Occurring in about one in ten people with HD, it looks different from the adult version. Usually the person is stiff, rigid and slow, may have involuntary movements that look more like tremors, and may experience seizures. Juvenile HD worsens more rapidly than adult-onset HD.

3. WHAT ADVANCED-STAGE HD LOOKS LIKE

Health care professionals look at HD as a disease made up of three disorders - a movement disorder, a cognitive disorder, and an emotional disorder.
Some people with HD have a very severe movement disorder but very little cognitive impairment. Others have profound cognitive changes but few movement problems. The emotional disorder is often depression, which comes and goes throughout the course of the disease. We'll look at each one of the "trio of disorders" in the following pages.

Every person experiences the beginning of HD in a unique way. Some first notice small subtle movements. Others find themselves becoming forgetful, and still others become depressed. Every person experiences the progression of HD in a unique way, too. For example, one might have a rapid deterioration in cognitive function and less decline in the control of movements. Another person may have rapidly increasing difficulty with movements, but no significant change in cognitive functioning during the same period of time. Therefore, no two people with HD will present the same caregiving challenges. But in the most advanced stages, all symptoms converge in a predictable manner.

MOVEMENT CHANGES

By the time a person with adult-onset HD comes to a long-term care facility, the movement disorder is usually quite apparent. But some years before, it began with small changes in eye movement and involuntary movements of the fingers and face. The symptoms progress to a point where all muscles are affected, and walking takes on a "dance-like" quality. People often try to camouflage and control the movements. As the involuntary movements, often referred to as chorea, become more exaggerated, what was once "dance-like" now looks "drunk-like". 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. Beds may need padded side rails to prevent the patient from bruising himself or falling out.

At the same time, it becomes increasingly difficult for the person with HD to speak and be understood. Nearly all people develop a swallowing disorder, need a special diet, and need assistance eating. At some point, a decision will have to be made whether or not to insert a feeding tube. This decision involves the affected person and family members, and it is most helpful if it is made well in advance. In addition, most residents eventually need adapted beds and wheelchairs to accommodate their severe involuntary movements, impaired balance, and changes in posture.

COGNITIVE CHANGES

The cognitive disorder is less apparent than the movement disorder, but more disabling in many ways. Long before he came to you for care, he struggled with subtle changes that affected his work and family. Most likely, his ability to organise and plan his work day slowly began to erode, and routine tasks, previously performed effortlessly, became more complicated to complete. As cognitive function continued to deteriorate, he may have become quite inflexible, wanting things done a certain way. People around him may have noticed these small but significant changes in temperament. Long-term relationships may have been jeopardised. He may have been unable to see changes in himself, and vigorously denied their existence.

Now, in your care, his thinking is slower, initiating action is more difficult, learning new things is not as easy as it once was, and judgement is impaired. He may have developed difficulty waiting for things he wants immediately and become unreasonably demanding of his family, friends, and caregivers. Now these problems challenge you as you assist him in his daily activities.

EMOTIONAL CHANGES

The emotional disorder is primarily made up of the depression that runs throughout the course of HD. Having seen his parent suffer with HD, knowing that only further decline and dependence is at hand, and recognising all that he's already "lost" to HD, it's easy to see why he might have a reactive depression.

Idiopathic depression, one that is not triggered by life's events, is also common. Some people with HD who are depressed appear irritable or angry. Some deny depression because they lack insight. Others are unconsciously protecting their feelings.

Even in the most advanced stages of HD, people who show classic signs of depression can respond well to medication. Suicide in HD occurs more often than in the general population. Depression paired with a lack of impulse control makes suicide a major risk for patients in all stages of the disease.

IN SUMMARY

There's no typical person with HD. Each individual has complex, unique needs. Some needs can be met easily. Others will require clever or creative solutions. And still others will require an ongoing trial-and-error approach. Taken together, though, you'll become well-versed in this person's care, and your rewarding days will far outnumber your challenging ones!

4. KEEPING COMMUNICATION LINES OPEN

A STRUGGLE FOR CONTROL

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. He has already lost his ability to drive a car, to manage his finances and to relate to his family as he did in the past. As the disease progresses, more losses will occur. If this person saw his parent or another relative in the later stages of HD, he has an idea of what is in store for him. No wonder he may seem angry or depressed or uncommunicative much of the time!

For the person with HD to keep control of what's done to and for him requires enormous effort. To express his feelings and needs is a struggle of monumental proportions. 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 organise information. Undertaking even the simplest movement is hampered by trouble getting started. His capacity for hope that he can maintain some control is compromised by periods of depression and even more losses.

If you remember the struggle he goes through, you will understand that every time he freely chooses which shirt he'll wear, what time he'll get out of bed, or how he'd like his eggs cooked, it is a significant victory in his great struggle to control his world. Help him do it!

SPEAKING...AND LISTENING!

Obviously, nothing is more important in your relationship with the person for whom you're caring than communicating with one another. This becomes more and more important as it becomes more and more difficult. The movement disorder affects speech in several ways. 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. Just as the movement disorder affects speech, the cognitive disorder affects the content of what is said. The ability to form ideas, organise thoughts, and present them in an orderly sequence is compromised in HD. Some people have difficulty starting a conversation, staying on the topic, or switching from one topic to another. Some may get stuck on one topic and have difficulty getting off it.

As clear speech becomes more difficult, it takes great effort for people with HD to carry on a conversation. They will have a tendency to 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 on his behalf. 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 "interpreter" with others.

It can be humiliating and frustrating for the person with HD, and embarrassing for you, when you have difficulty understanding his words. One way to show him respect is to put the burden of understanding firmly on you. Ask him for clarification. Ask his permission: "Do you mind if I repeat your words to you from time to time so you will know how I'm doing?"

Communication boards are commonly introduced to people who are having difficulty being understood. As well-intended as they may be, boards are not often adopted by people with HD as an alternative form of communication. Speaking, as impaired as it is, is easier than learning to use the unfamiliar board. As with other adaptive devices such as helmets and wheelchairs, introducing the communication board 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 his interests, hobbies, family, career, and preferences. Since non-family caregivers may first meet him when he has difficulty expressing himself or recalling events from the past, the album serves two purposes.

First, the album is a communication aid which allows him 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 the sports teams he rooted for in past years can be the basis for hundreds of conversations in your years together.

Please remember that people with HD can comprehend our speech and understand all that's going on around them to a far greater degree than most people may at first suspect. How effectively they communicate through spoken words is not an accurate predictor of how well they understand what you say.

Family members and caregivers agree that people, even in the most advanced stages of HD, somehow manage to communicate with their caregivers very effectively through facial expressions, eye gazes, and other subtle movements that may only be understood by those closest to them. Look and listen carefully!

IN SUMMARY

HD impairs communication in many ways. Speech is affected. The ability to organise thoughts and present them in an orderly way is compromised. Sometimes individuals with HD speak to you through the nonverbal messages of anger, withdrawal, and short temper. Remember that the messages are there for you. To communicate you will need to develop skills in decoding both verbal and nonverbal messages to you and others around.

Caregiver tips

Have the person with HD:

- Slow down, especially if his speech has a "racing" quality to it.
- Repeat/rephrase.
- Say the main word.
- Spell the word.
- Write the word, even if he can write only a few letters.
- Show you.

You, the caregiver should:

- Try to rephrase the main idea.
- Use short sentences.
- Ask for feedback.
- Allow plenty of time.
- Wait... for up to a few minutes... for a reply.
- Try not to repeat or rephrase a question while you're waiting for a response.
- Use touch to help keep him focused on the conversation.
- Ask for help from others when needed.
- Never pretend to understand!
- Consider using a simple communication board.

The family can:

- Make a scrapbook or memory book.
- Tell you about facial expressions or phrases that they understand.
- Continue to call or write even if he cannot respond clearly.

5. EATING AND NUTRITION

PREVENTING WEIGHT LOSS

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 for both of you. 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. No wonder many people with HD say they are always hungry!

In long-term care facilities, nutritionists should consider double and triple portions for people with HD. In fact, free access to food may be the order of the day.

When he asks for more food, some say reflexively, "But you just ate!" It may be more appropriate to say, "Oh, you just ate, but can I get you something else?!" Someone in the later stages of HD who is overweight is very rare. Take his requests for more food or supplements as seriously as you would anyone else who is very, very hungry.

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 patient is awake at night. This prevents constant hunger and may help to minimise gulping. Another strategy is to increase caloric intake by creating a diet of high-calorie foods. Imagine: A person with HD can have a weekly menu of dishes that most people prefer, but choose to avoid because they're so high in calories!

Caregiver tips

- Give frequent meals and high-calorie snacks and drinks to prevent weight loss.
- Help the person with HD eat until full. A calm relaxed environment with good ventilation helps.
- Ensure eating is slow and deliberate.
- Help the person sit in an upright position when eating and digesting food.
- Beware hot drinks. A decreased sensation of heat can cause burns!
- Ensure mouth is kept in a healthy condition.

SOME HIGH CALORIE FOOD

If each mouthful of food is so difficult to chew and swallow, then maximising the number of calories in each bite can only help. There are anecdotal reports that those who reach an ideal body weight report feeling better in general, may metabolise medication more smoothly, and maintain function longer than those who have had significant weight loss. 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, common in long-term care facilities for many years, are now available in most local pharmacies. Family and professional caregivers have cleverly invented "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 are useful. Adding full fat butter or cheese to creamed potatoes, vegetables or soups also help. Extra sugar, honey or cream in porridge, readybrek or fruit smoothies help increase calories. Please contact your pharmacy or HDAI for further information on diet and nutrition.

Super Shake (Single Serving)

- Glass of full fat milk
- Ice-cream 2 scoops
- Ovaltine 2 teaspoons
- High-protein, high-cal additive* 1.5 scoops
- Fruit add to taste

In a blender, blend milk and high-protein additive together; add Ovaltine. Blend in the 2 sccops of ice-cream (or more) and fruit to preferred thickness.

**Food supplements are prescribable under the medical card in order to prevent/treat the disease related malnutrition which often occurs in Huntington's Disease..*

IN SUMMARY

Eating is one of the primary pleasures in life. Against all odds, most people with HD struggle to eat independently, then with assistance, for as long as they are able. However, with difficulty controlling the movements to get the food to their mouth, involuntary movements that interfere with eating, altered or puréed food, drool, bibs, sudden inhalation and possibly coughing, a meal can be a messy affair. A committed caregiver can make a big difference by taking the time to help the person with HD take in as much food as possible, as safely as possible.

6. PREVENTING SWALLOWING PROBLEMS

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. Those with HD are at serious risk of choking, aspirating, and even suffocating.

Preventing these problems in advanced HD is an ongoing challenge to a caregiver. 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 therapist can make recommendations regarding positioning the patient, texture of food, and other issues that will make swallowing easier.

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 today. 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 or catalogues.

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 tongue or mouth on hot foods. Some people with HD tend to "stuff" food; that is, place more food than they can possibly chew and swallow into their mouth as quickly as possible. This behaviour greatly increases the risk of choking and aspiration and should be discouraged. Providing or feeding them 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 his mouth for adequate nutrition, and a large amount of food is wasted in the struggle to feed himself. Unaware of how nutritionally inefficient his eating has become, he may see your intervening to feed him most of his meal as a final loss and a symbol of his dependence on others for his sustenance. By assisting him with small parts of the meal earlier than he really needs your help, he may become accustomed to your help and be more willing to accept it when it is absolutely necessary for his safety and nutrition. For example, spooning a thick shake into his mouth at the end of a tiresome meal or placing a few pieces of a snack into his mouth at various intervals throughout the day may gradually help him to accept this degree of assistance.

A final thought: Since this person may be hungry, tired, irritable, and unable to wait, it may be wise to help him eat first if you have several people to assist at the same meal, even though he may take longer to assist than others in your care. If you can provide the person with HD with a comfortable experience eating, meal after meal, then you are an excellent caregiver!

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 coughing after swallowing a mouthful of food and waiting through that tense moment for him to stop and take his next breath to assure you that he is not choking. Never consider coughing during a meal as a routine part of eating.

Coughing is a defensive reflex to prevent choking. Consider it Mother Nature's alarm that there is a serious problem to be addressed immediately. Report coughing while eating to a nurse immediately for assessment.

Choking, indeed, is a very serious risk factor. Be aware of this every time you help someone with HD to eat a meal. 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. Choking and aspiration pneumonia are not uncommon causes of death in people with HD. Individuals with swallowing problems need to have their temperature and lung sounds monitored regularly for signs of pneumonia.

Learn the Heimlich manoeuvre so you'll be prepared to respond to a choking incident. Make sure everyone who assists this person to eat is practiced in the manoeuvre. It may be reassuring to explain or demonstrate it to him if he has previously had a serious choking incident. Listen very carefully to the instructions you are given on how to help this person eat his meal. Take no shortcuts; take your time. Check for proper positioning every time you put food in his mouth. Eliminate as many possible distractions in the room as you can. Double-check the texture of the food that's been specially prepared for him. Be 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 helping him eat takes too long or is too tiring for him, arrange to have him eat less food more often throughout the day.

Warning signs of swallowing problems

Any one of these signs could indicate a serious problem with swallowing. Consult your physician or nurse immediately.

- Clearing the throat frequently.
- A voice that sounds wet or "gurgly".
- Spoken or nonverbal expressions about fear of eating, swallowing, or choking.
- A delay in swallowing after food has been chewed.
- Holding food or liquid in the mouth without swallowing it.
- Exaggerated movements of the jaw, lips or tongue.
- Tilting the head back to eat or drink.
- Swallowing several times on one bite.
- Food or liquid falling out of the mouth.
- Food left in the mouth after swallowing it or finishing a meal.
- Coughing during or after the meal.
- Fatigue or exhaustion after or during the meal.
- Significant weight loss over time.

CREATING CULINARY MASTERPIECES WITH PURÉED FOODS

Physicians or speech/language therapists may recommend that people with serious swallowing problems and an increased risk of choking eat a diet of purée consistency. At home or in long-term care facilities this is typically done by placing each item of a meal into a food processor and blending it beyond recognition, except for its basic colour. As if the anxiety of choking were not enough, looking forward to a daily menu of mush that looks like commercial baby food only adds further insult to injury. However, there is an alternative. You can plan and prepare an entire menu cycle of moulded dishes, casseroles and loaves that taste, smell and look appetising, but are the consistency of purée.

During the holiday season, department and specialty stores sell plastic candy moulds to make lollipops or chocolates in your kitchen. Like those moulds of bunnies, Santas, and ghosts, moulds of chicken legs, pork chops, broccoli florets, pear halves, and fish filets are also available. A selection of these will make your meals much more attractive.

For example, cook a chicken, remove its meat, place it in a food processor, and blend it to purée consistency. Add bread crumbs, egg whites, or a commercially available thickening product. Then place this chicken mixture on a plastic sheet with the multiple chicken legs moulded into it and freeze it. When chicken is on the menu, pop one leg from the mould, baste it, and heat it in a convection oven. It maintains its moulded shape and your kitchen smells like you're cooking... chicken!

With gravy and garnish, it looks and smells just like the unaltered chicken the rest of the family is having for dinner. It has the consistency of a chicken pâté. It looks so real, it's not uncommon for nurses' aides to return moulded food to the kitchen because it looks like kitchen staff forgot to purée it!

By planning a menu of these moulded dishes and loaves (meat loaf, for example) and casseroles (tuna casserole, for example) and paying close attention to its required consistency, you can serve this puréed cuisine as an alternative to "baby food" in a three-section plate, originally designed for infants.

Caregivers tips

- Eating should be slow and deliberate.
- Be sure the person is positioned properly.
- Choose food of appropriate texture and temperature.
- Learn the Heimlich manoeuvre.
- Report any coughing or choking incident to your supervisor.
- Make sure he takes small bites and sips.
- Alternate solids and liquids.
- Have him "dry swallow" or "double swallow" between bites.
- Have him sit up after eating.

FEEDING TUBES

As swallowing becomes increasingly impaired, eating by mouth compromises adequate nutrition. At this point some people with HD may choose to receive their nutrition through a gastrostomy tube. Although it is a relatively minor surgical procedure, placement of a feeding tube has greater implications than simply enhancing nutrition.

Deciding whether or not to have a feeding tube forces the individual and his family to confront difficult emotional or spiritual issues about extending life, the quality of life, and providing basic sustenance to prevent starvation. These are very personal decisions, and your understanding and support are needed.

Placement of a gastrostomy tube (commonly called a "g-tube"), a peri-epigastric tube (commonly called a "PEG"), or a jejunostomy tube (commonly called a "j-tube") may not mean that it is no longer possible to eat by mouth.

It is often good practice to continue to take some favourite foods orally. Remember, too, that placement of a feeding tube can be a short-term intervention to help build body weight so that the individual can resume eating primarily by mouth.

If he has a feeding tube, the spot where the tube is placed is particularly prone to infection. Look at this area closely whenever you feed or change him. Pay close attention to washing, rinsing, and drying the skin around the tube when you assist him with bathing. Remember to wash your own hands. Follow any special instructions you may be given to keep the tube and the area around it clean. Be sure to report any signs of infection to a nurse so they may be assessed. Be sure that he is always positioned so that his head is above the level of his stomach to prevent regurgitation or aspiration.

People with severe chorea may find that the area around the tube becomes sore or tender from the repeated involuntary movements of the arms and legs touching or pushing against the area around the tube. Some may find the site so irritating that they tug at the tube, which loosens it. They may injure themselves or even remove the tube. To protect them from accidentally irritating the area or to prevent having to replace the tube, a doctor may order a binder to wear over the site. It's important to put the binder on correctly. If the binder does not fit snugly and smoothly, it will further irritate the skin rather than soothe the discomfort.

When considering a feeding tube

Many people with HD and their families struggle with the decision of whether or not to use a feeding tube. It is never an easy decision, and it is best made well in advance of a crisis. Here are some considerations about feeding tubes.

A feeding tube may be called for if:

- there is a nutritional crisis.
- there is a hydration crisis.
- there is repeated aspiration pneumonia.
- there is a severe swallowing problem.
- there is great fear of choking or aspirating.
- it makes continuing an active life easier.
- there are other conditions, disorders, or complications.

Placement of a feeding tube may be appropriate if previous attempts at continuing to eat by mouth have included:

- changes in position.
- changes in the consistency of the food.
- a speech therapist's swallowing evaluation.

Placement of a feeding tube may be appropriate after the following interventions have been tried without success:

- medication to make swallowing easier.
- interaction between the person and those helping him to eat.
- adjustments to the environment in which he's eating.
- achievement of a greater degree of relaxation while eating.
- consideration of any other psychological factors.

In some cases placement of a feeding tube may be detrimental. The "right" decision requires that everyone involved make every effort to make their contribution as informed as possible.

7. PREVENTING FALLS

As HD progresses, the muscles that support an upright neck and trunk grow weak. As a result, people with HD tend to hold their head in a forward or hanging position and slump their shoulders. 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. Because most of the medication used to treat chorea can cause or worsen rigidity, many physicians prefer not to treat chorea unless it is very severe.

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. Although orthotics may have been helpful earlier, high-topped sneakers with a wide heel or light-weight work boots are more likely to help in later stages. Avoid high heels, sandals and other shoes that offer little support. Check for wear regularly. Shoes in poor repair cause falls.

Three of the most common places that falls occur are in front of a chair, a toilet, or a bed. Turning to sit down requires him to place all his weight on one foot, to balance, and to pivot on it for a moment. If the ability to balance on one foot is impaired, the person may fall. Similarly, if judgement or spatial awareness is impaired, he is likely to misjudge his distance from the bed, chair or toilet. He may turn to sit and miss it completely, falling to the floor. Teaching simple strategies to compensate for these problems helps reduce falls or prevent them altogether.

One such strategy is to teach a simple procedure called "touch-turn-sit". Instruct the person to bend over slightly and actually touch the chair, toilet, or the side of the bed before turning to sit on it. Touching the chair assures him that he is close enough to sit in it.

Touching also provides support for better balance. Here is an easy procedure to prevent falls. When he is getting up out of a chair, teach him to place his hands on his knees and lean forward. This, in effect, brings his trunk forward - a better position for getting up.

Caregivers tips

Consider these environmental changes to prevent falls:

- Stabilise furniture so that it cannot move.
- Use chairs with armrests and high backs.
- Clear rooms of any unnecessary furniture.
- Remove scatter rugs and high-pile carpeting.
- Remove tables and lamps from frequently used household pathways.
- Pad furniture or doorways if they're hit often.
- Use sliding doors.
- Round off sharp corners of furniture or fixtures.

You can also reduce or prevent falls if you:

- don't call a person with HD from behind, causing him to turn quickly and lose balance.
- don't interrupt him suddenly.
- don't give a medication while he is standing.
- don't try to stop him from "bouncing off the wall." (Some individuals with dystonia walk quickly up on their toes. This gives the appearance that they are about to walk into or bounce off the wall. Rarely do they ever touch the wall. However, well-meaning people often actually cause falls in their efforts to protect them from hitting walls.)

As he struggles to maintain his independent mobility in the face of balance problems and the other complications caused by the movement disorder, he will inevitably fall. As often as falls may occur, they can never be accepted as commonplace events. Even minor falls can lead to cuts that require stitches, or cause painful bruises, broken bones, or injury to the brain. Anyone who has fallen should be examined carefully. Changes in movement, mood, thinking, or neurological function could be a warning sign of a subdural hematoma, or bruise on the brain, even if the changes develop a few days after the fall. Notify your supervisor or a physician immediately if you see these kind of changes in someone who has fallen.

ABOUT HELMETS AND OTHER PADDING

It may be difficult to convince someone to wear a hard bicycle or hockey helmet for protection from injuries to the head due to falls, especially if no one else around him is wearing one. Sometimes a soft head protector, made of leather bands with a Velcro chin strap, is more easily accepted. Although certainly not fashionable, they offer some protection from striking the head while falling. Sadly, it's often right after a fall that someone is most willing to try wearing one. Thinking about head protection before falls become severe may help to avoid serious injury. Setting up a schedule to wear one for a very short period each day is a way to introduce it. To allow the person wearing the helmet to maintain some control over his day, he can select which time to wear it.

Some people repeatedly injure their heads with falls; others seem to repeatedly injure one elbow or both knees when they fall. Wearing hockey-type protectors for the joint or joints may help to prevent traumatic joint swelling.

ABOUT WALKERS

Although for many people with moderate or severe chorea a walker is not helpful, these devices can work well for those who are able to maintain a firm grasp. With a stable base, the rollator or rolling walker has been especially helpful to many folks because it allows a physiotherapist to set its hand grips in a way that shifts the patient's centre of gravity forward. Although they are simple devices, many physical and dynamic factors need to be considered in selecting one. Consult a physiotherapist to evaluate which type of walker best matches his abilities and disabilities. Occasionally some may benefit from wrist or ankle weights to decrease chorea.

Some people with HD get around by themselves in "merrywalkers". They are best used in homes or facilities with ample space to accommodate their considerable size.

NUTSHELL CASE STUDY

Introduce assistance before it's needed

When Andrea first came to her nursing home about three years ago, she had significant balance problems from her HD and had seriously injured herself falling. But she was determined to continue walking... alone! Andrea'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 programme of teaching her how to wear a helmet and use a walker before she absolutely needed one. Andrea practiced her "workout" every day as her "coaches" would encourage her to practice her balance exercises with a helmet on. They taught her how to use a walker and practiced those skills before she needed them. One day she had a serious fall. It was suggested that she wear the helmet full-time. The idea stuck; she agreed. Three months later another fall prompted the introduction of the walker on a full-time basis. Already familiar with it, she accepted it readily. Over the next six months she had far fewer falls.

As soon as Andrea 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 Andrea began to fall frequently in the evening. She agreed when it was suggested that she use the wheelchair just at supper time and in the evening. 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.

ABOUT FATIGUE

We know when we're tired. And the people around us know when we're tired as well. Recognising our fatigue, we might yawn and say, "I'm exhausted. I've been on my feet all day." Or eyes may droop so that a friend says, "You look beat." We walk a bit slower and talk a bit more softly. It usually happens at the end of the day or after an extended period of work. You usually don't have to know someone well to know when they're tired.

In the more advanced stages of Huntington's disease, fatigue affects people dramatically, but it is often difficult for caregivers to recognise it for several reasons. We are not accustomed to seeing someone very tired early in the day. The person with HD may not be able to communicate how he feels in words, but his behaviour or his motor function may get worse.

People with HD use great effort for simple things like walking and standing and may become fatigued early in the day. After a half-hour struggle to chew and swallow safely, trying to sit up straight, and frightened all the while of choking, breakfast can be exhausting. When a person who has problems with balance, poor posture, and severely impaired walking falls down one afternoon it comes as no surprise. It is not always obvious, however, that the primary reason for the fall was fatigue. If there is a pattern to behaviour or motor problems that a particular person shows, consider whether fatigue is playing a role. Offer a nap or rearrange the daily routine to better fit the person's needs and abilities.

Caregiver tips

To prevent fatigue:

- Schedule and allow rest periods throughout the day.
- Offer the use of a wheelchair at the time of day when the patient usually gets tired.

ABOUT WHEELCHAIRS

The importance of choosing the right type of wheelchair cannot be overstated. Selecting an appropriate wheelchair prolongs mobility, prevents deformity, conserves energy, and allows the individual with HD to do many activities without help. A poorly selected wheelchair can discourage mobility, contribute to deformity, and jeopardise safety.

Many people with HD have an easier time propelling a wheelchair with their feet than they do with their hands. If this is the case, your physiotherapist will likely recommend a "hemi-height" or "drop seat" wheelchair so that the person can firmly plant his feet on the floor.

It may be very difficult to find an appropriate wheelchair or "seating system" for some individuals. They have involuntary movements, rigid or fluctuating muscle tone, unstable posture, and an inability to modulate the force of their movement. A range of specialist seating is available from suppliers and manufacturers who will undergo assessments of people with HD to address the unique needs of the individual. Such specialist equipment may include deeply angled seats, good padding, strong, lightweight frames and tilt positioning to reduce the formation of pressure sores.

Selecting a wheelchair

Selecting an appropriate wheelchair is a team effort involving the person using it, his caregivers, his physician and his physiotherapist. To determine the best wheelchair, physiotherapists make these considerations:

- Does the chair restrain the person as little as possible?
- Does it allow enough room to move freely and without injury?
- Are its hard surfaces and sharp edges padded?
- Does it allow the user to get into and out of it easily?
- If appropriate, does it provide independent mobility?
- Does it offer solid steady support for the feet?
- Does its height allow it to be used at a table or with a lap tray?
- Will involuntary movements cause the chair to tip over?

Adapted from Lori Quinn, Ph.D., PT

AVOIDING RESTRAINTS

The long-term care industry has made great strides in reducing the use of restraints in the last five years. However, the use of restraints is not uncommon among residents with HD in long-term care facilities, even though it may be particularly risky in this group of residents. Restraints do not prevent involuntary movements but can lead to injury of the limb, chest or abdomen, or even strangulation. Adam and Brenda serve as two examples.

NUTSHELL CASE STUDY

Alternatives to restraints

Adam has involuntary movements and keeps sliding down and out of his wheelchair. To help him remain seated in the chair, his nursing home staff restrain him with a seat belt. A more thorough assessment of his movement disorder might suggest a high-back seat, increased seat depth, foot supports, arm rests, and padding to prevent him from sliding out of the chair without the risk of a belt restraint.

Brenda's balance is impaired. She has a deficit in spatial awareness and impulse control disorder. She's always hungry and never sits still. She constantly stands up from her wheelchair and quickly falls down. Brenda's nurses and therapists initially recommended a waist restraint to keep her safe in her chair and to prevent her from falling. On further reflection, though, they came up with alternatives to restraint. Anticipating Brenda's needs and wants, they arranged a daily routine by scheduling periods out of her chair to walk with supervision, to eat more frequent smaller meals, and to get out of her chair to relax on a couch or bed. They also use bed and chair alarms to alert them when she gets up so they can quickly respond to her.

ABOUT BEDS

Although most people with HD have no involuntary movements while sleeping, many have difficulty remaining in their beds. Deficits in spatial awareness make it difficult to sense the edge of the bed. Although bed rails provide protection for some, in other cases, they may serve as nothing more than objects to bang up against, or they may serve merely as obstacles to climb over when the patient feels an urge to go to the bathroom in the middle of the night.

For those who walk, "low beds", which are between 12–20 cm off the floor, may be safer. A thin, high-density mat can be placed on the floor next to the side used most often. In some cases one side rail may be left up and padded with several centimetres of foam, leaving the other one down for easy access.

In the presence of very severe chorea when side rails are necessary but traditional side rail pads are inadequate, an alternative method of padding may be necessary. A foam mattress overlay (Ultraform, for example) can be easily cut in half lengthwise with an electric knife. This creates two side rail pads with enough padding left over to pad the head and foot boards. The pads, covered with thin plastic and a sheet, are secured to the side rails by clip-style buckles with belts of webbing. This should protect the patient from bruising or abrasion.

A few individuals seem to "vault" out of their beds. This is caused by an inability to regulate or "modulate" the force of voluntary movement.

The large muscle groups in the legs are used while turning over or adjusting the position in bed. Large poorly modulated extensions and contractions of these muscle groups can result in the individual flipping out of bed. There are no traditional beds available that allow a great deal of freedom of movement and protection from serious injury from the vaulting. One possible solution to this problem is to build a specialist bed with high padded walls for all-round protection against injury or entrapment. It has fall-down sides ,to allow access for transfers and care.

These beds are often criticised for their odd boxy appearance and the degree to which they shelter the person from stimulation in the room around them. However, experience has shown that people with HD who use them are grateful for a good night's sleep, the chance to roll over and change position without "vaulting" out, and the opportunity to sit up without crashing into a side rail.

Other possible adaptations are simply placing mattresses on the floor; using a double or queen-size bed rather than a single or twin bed; and using chairs, lounges and beds made of moulded foam.

8. 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 programme can help to address all areas of decline and help him become stronger, improve balance and posture, and feel more in control of his 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 him maintain his 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.

Sample exercise plan

All exercises should be done slowly, five to ten times each.

Arm Exercises:

1. Lie on your back with your legs straight. Stretch your arms overhead, hold the position momentarily, then relax.
2. Lie on your back with your legs straight and arms at your side. Make a fist, strongly straighten your arm, raise it about 30 degrees, hold it, open your fingers, then slowly lower your arm to the floor.

Breathing Exercises:

1. Close your mouth. Inhale through your nose while expanding your chest and abdomen. Hold for a few seconds. Exhale through the mouth as completely as possible.
2. Do it again; this time exhale through your nose and make the sound "mmm".
3. Now again, exhaling through your mouth while making the sound "ahhh".
4. Again and cough two times.
5. Again and this time swallow after you exhale.

Trunk Exercises:

1. Lie on your back with your knees bent and feet flat on the floor. Lift your hips up, hold the position, lower yourself down slowly, then relax.
2. Roll onto your stomach, then push yourself up on your hands and knees. Raise one arm forward, reach out and hold the position. Now lower your arm and raise the opposite leg up as straight as possible. Hold the leg up, then lower slowly. Repeat it with the other arm and leg.
3. Begin on your hands and knees, then lower your hips so that your shoulders, hips and knees are in a straight line. Now lift your feet off the ground, bend your elbows, and lower your upper body to the floor and back up in a modified push up.

Gross Motor and Balance Exercises:

1. Sit on the floor with your legs crossed. Try to keep your knees as low as possible. Now reverse your legs.
2. Sit on the floor with your back and legs straight. Reach for your toes. Hold that position. Repeat.
3. Standing with your feet six inches apart, shrug your shoulders up toward your ears, hold that position, then relax.
4. Walk forward with one foot in front of the other as if you were walking on a straight line. Now try it going backwards.
5. Stand on one foot. Count the number of seconds you can do it. Now do the other foot.

NUTSHELL CASE STUDY

Physiotherapy helps!

Tom reluctantly came to a nursing home because he was no longer able to live alone and had no family members who could help him. He looked depressed, unkempt, and undernourished. He kept to himself as much as possible, avoiding staff and fellow residents as best he could. 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 Tom and chatted briefly with him in his room.

When the coach learned that Tom, 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 Tom 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 pulmonary status. After eight weeks of "workouts" in the gym, Tom 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 programme. Three months later his strength, balance, gait, and respiratory status were all improved.

9. PERSONAL CARE

If you can't lay out your toothbrush, toothpaste, a glass for rinsing, and a towel to dry your mouth, then brushing your teeth can be a very intimidating activity. We often overlook the fact that before we begin to brush our teeth, we make sure we have everything we need and that we plan to follow a very set sequence of steps. Because these abilities to plan and organise such daily activities are often "automatic", we often take them for granted. These are the very skills that present problems as HD progresses. The cognitive disorder of HD presents more problems with these activities of daily living than the motor disorder does.

People who face these difficulties in bathing, dressing and grooming react very differently. Having lost interest in themselves, some "give up" and easily let those caring for them "do everything". Others plow ahead undaunted by the many challenges of caring for themselves, unfazed by an occasionally misbuttoned blouse or a t-shirt put on over a sweater. Either way, needing someone else's assistance is yet another significant loss to which the individual with HD must adjust.

BATHING

Many people with HD are 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 before an unfamiliar caregiver can be humiliating. Perhaps the feeling of water is no longer pleasing and being splashed is very unpleasant. Maintaining both your modesty and balance while trying to help your helper to help you is certainly a tiring job!

Even though it is difficult, it is important for people with HD to bathe often. Many perspire profusely. Increased hunger and thirst require that they eat more food and drink more liquids, which leads to more frequent urination. Involuntary movements, dystonia, and problems with balance cause spills that soil both clothes and the people who wear them. Changes in the tone and weakness of the facial muscles, as well as less effective and less frequent automatic swallowing, can cause some people to drool excessively. This frequent need for bathing means arranging times for bathing in the daily routine.

Try to keep the shower or bath as brief as possible by gathering everything necessary before beginning. 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, minimising 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

Extra attention needs to be given to the bathrooms used by people with chorea, dystonia, and problems with balance. "Flopping down" onto toilet fixtures loosens the hardware that holds the seat to the bowl, as well as the wax seal and fittings that hold it firmly to the floor. 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 odor problems. Floors wet from urine or water are another hazard that contributes to falls. The widely available padded toilet seats may be helpful since they cushion the impact of "flopping down" onto the seat, and the padded cover may protect his back and the toilet tank as well.

DRESSING

Clothes should be changed and washed often, too. It's helpful if they're easy to get off and on and are durable enough to withstand many, many washings. They 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. Patterned shirts and blouses camouflage spills and stains; single-colour tops accentuate them. Fashionable patterned scarves may catch drool as effectively as institutional bibs. Many people with HD feel hot and prefer to have the thermostat turned down or fans blowing in their rooms. They may even feel comfortable wearing light cotton clothes in the wintertime.

Be sure to allow him to complete as much of his dressing routine as he can without assistance. Giving more assistance than is actually needed is a widely accepted, usually unnoticed, unintentional but harmful act against a person. It robs him, little by little, day by day, of what little independence he has remaining. It leaves him increasingly dependent and docile. Over time, you may teach him to be helpless. "Learned helplessness" is the inevitable consequence of caregivers unwilling to make or take the time to allow people to do what they can do for themselves.

Sometimes one particular caregiver or family member can get the resident to do things that no one else can; respecting this preference by allowing that caregiver to work with the resident as often as possible can be a comfort to the resident, family, and staff.

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. His anxiety can intensify his chorea and make it more difficult to help him. Toothbrushing will be a comfortable activity if the person needing help and their helper take a few moments to relax together and to position themselves in such a way that the helper can gently stabilise the head.

The importance of oral and dental care increases as the disease progresses into its more advanced stages. He may breathe in his own saliva. Effectively cleaning the mouth minimises 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 not uncommon. 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.

IN SUMMARY

HD damages the brain so profoundly that it changes movement, emotions, and thought. These changes affect the person with HD in ways that are difficult to understand. He may resist taking a shower, changing his clothes, and brushing his teeth. He may see your prompting as nagging, your direction as patronising, and your insistence as antagonism.

A decreased interest in self-care is typical in HD. Establish a self-care routine. Understand the problems. Set clear expectations. Give no more assistance than needed to start or finish an activity. His disinterest is driven by the disease, not a lack of concern about his appearance or hygiene. He really needs your help and support in this area!

10. UNDERSTANDING COGNITIVE CHANGES

At different times, you'll find he is distracted, confused, uncooperative, angry, and withdrawn. He may demand things from you immediately. He may angrily challenge you as you try to protect him from injury. He may refuse to do therapeutic exercises with you. He may even yell and threaten when you ask him to do the simplest things.

When you're trying your hardest to give him the best possible care in very challenging circumstances, it's difficult not to take it all personally. 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's disease. They are not caused by a dislike for you, by a spiteful attempt to make your job more difficult, or because he's a bad person. He is not the problem. The behaviour that comes from changes in his brain is the problem... for both of you.

A critical component of your care is to look carefully at his actions and to try to determine their cause. Often, what is labeled as "inappropriate behaviour" is an attempt by the individual, through great impediments caused by the disease, to express his needs or preferences. The better we are at understanding them, the smoother your caregiving relationship.

NUTSHELL CASE STUDY

"Would you kindly warm this up a bit?"

Patricia has HD and lives in a nursing home. For several days, she threw her breakfast tray onto the floor every morning. The nurse and aide caring for her saw her "agitation" and attributed it to HD. The nurse reported Patricia's misbehaviour to her physician, who ordered an antipsychotic drug, Haldol, for the "agitation". However, further investigation by an aide more familiar with her showed that she had a complaint that she couldn't express verbally: her coffee was cold. Given fresh hot coffee, the problem was resolved without medication.

The best caregivers understand that "inappropriate" behaviour may be an attempt to express needs or preferences despite the many impediments and all the impairments caused by HD.

Here are some of the ways that changes in the brain affect this person in your care. By understanding these changes, you may be able to better "read" his needs and preferences, and also find new ways to do the things he 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 he responds. You may ask, "Would you like to go shopping today?" Five seconds later you've still not received an answer and you've gone on to someone else. But ten seconds from when you asked him, he may say, "Yes!" Too often caregivers mistake the delayed response to mean "No!" No response may not mean "No!" Allow more time than usual for him to respond. Once you've recogniSed a delay in responding, you will be able to wait more easily. You may also find that you can anticipate responses with surprising precision!

Despite all the challenges 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 you and I, there is often an urgency on our part to do it for them. By understanding his cognitive deficits and anticipating processing delays, you can wait for him to respond and allow him to participate in his own care. Those who don't understand the deficits and delay can actively rob him of his independence or teach him to be helpless.

NUTSHELL CASE STUDY

Difficulty getting started

Mark wakes up and sits on the side of his bed. "Good morning, Mark! Breakfast is ready downstairs." Five minutes later he is still sitting there. "Mark, it's time to wash up and eat breakfast!" Five more minutes later he's still there. You approach him, hand him the washcloth and toothbrush, motion toward the bathroom door and say, "Here you go, Mark, start by washing up!" Five minutes later he has washed his face, brushed his teeth and is dressing himself.

Sometimes initiating an activity - just getting it started - is very difficult. Just like Mark did, people may need a "jump start" from you. Do the first few steps of an activity with them or for them and you may find they complete the rest of it without your help. By allowing them to complete it themselves, you are actively helping them maintain their independence.

DIFFICULTY LEARNING

There is a myth that people with HD cannot learn new information. If he has learned your name and can find his own room, he has already disproved that myth! As HD progresses, it is certainly true that learning new complex notions and concepts becomes progressively more difficult. If people tend to learn by doing, it may take them many more repetitions or opportunities to learn. If they tend to learn by trial and error, they may not learn from their mistakes the first time they make them. But believing the myth that people with HD cannot learn new information can become a cruel self-fulfilling expectation. Try to give him all the opportunities he needs in order to learn new information.

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 ORGANISING ACTION

Many of the activities we engage in every day involve long sequences of smaller activities. Peeling, slicing, boiling or frying vegetables; cooking meat in an oven; and setting a table are all parts of preparing a meal. Choosing clothes; putting on underwear, socks, shirts, pants and a sweater; buckling a belt; closing zippers; and buttoning shirts are all parts of dressing. These sequences of activity become "second nature" or "automatic" and we think little about them when we can do them. Unconsciously, we've organised the information and actions required to complete them. Some people have difficulty organising these sequences of activity at some point in the course of their HD. This may explain why some 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 on a wardrobe. Posting schedules of daily activities and the time to do them may help organise the day.

NEED FOR ROUTINE AND CONSISTENCY

Picture this. Someone smiles and tells you, "It's time for breakfast!" You've eaten breakfast every day of your adult life, but you're not exactly sure what this means. You're partially dressed, so you quickly run through what you have to do before eating. It's confusing. You can't think as quickly as you once did, and you stand there for a while, just thinking and trying to get going. A noise in the other room distracts you. Now you can't remember what you were trying to do in the first place! You just can't figure out what to do next. You're distracted, confused, and annoyed. Again. At noon you hear someone shout, "Lunch is ready." And it starts all over... again.

If you're confused and don't know what to do next, sometimes you do nothing. When you're confused, sometimes it's easy to get angry. When you're confused, you don't know what to expect next. In fact, not knowing what's happening next is exactly what's confusing you! It's a relief to know what to do next. 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 for him to know what's next in his day. This routine helps him to predict the day, gives him confidence that he can do whatever is asked of him, builds trust between him and you, minimises distractions that can disrupt daily activity, and makes it easier for him to perform at his best. You've established a routine in which he can succeed.

In nursing homes, where there are shifts and many personnel 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 his daily schedule, noting the importance of consistency to him and his dependence on his routine. If you're unable to keep to his daily routine, then let him know the day before. In this way he will not be surprised. He will have enough time to dwell on the change and the change will not become a disruption. Whatever routine you establish, be sure it's easy to follow. Once it's in place, it's difficult to change it. A caregiver who establishes a daily routine for him provides the best care. There is comfort in consistency, and power in routine.

A NEUROLOGICAL LACK OF SELF-AWARENESS

It is disturbing to see someone with severe chorea or impaired judgement get into a car and drive off. In the more advanced stages it is just as disturbing to see him with severe chorea and profoundly impaired balance get up out of his wheelchair and try to walk, only to fall down. He tries to light a cigarette, even though he can't hold the cigarette still in his mouth or get the lighter close to the end of the cigarette. It's easy to say these people are in "denial" about their disability, using a psychological defense mechanism in which they refuse to accept their limitations.

This becomes a great source of concern and anxiety to those around them, but the behaviour may actually be due to a neurological lack of self-awareness, an inability to accurately perceive themselves. Someone with readily apparent chorea will often tell you that he is unaware of it! When you "confront him" with examples of his own disordered movement or disability, you are asking him to look at something he just can't see.

Interestingly, some individuals, even as they deny that they have chorea or HD, will accept treatments or remedies for their symptoms or problems. Thus it may not be necessary to "convince" the person that he has HD in order to care for him.

POOR JUDGEMENT

Caregivers are often concerned when they see someone with HD using poor judgement. They often become involved in "power struggles" as they try to dissuade him from doing something they prefer that he not do.

You need to know when to "back off". As difficult as it may be, if an individual's poor judgement does not hurt anyone, you might consider allowing him to do what he wishes to do. You may be allowing him to come to your point of view for himself.

NUTSHELL CASE STUDY

Maureen's cold bath

Maureen has had HD for ten years. She lives in a nursing home. One night she wanted to take a bath. Several residents on her floor had just completed taking showers that evening. The hot water was not keeping up with demand and was running from the faucet at room temperature. As she gathered her shampoo, towel and bathrobe, her nurse aide came to assist her by drawing the water for her tub. The tub half-filled, her aide turned off the water and put her hand in to check the temperature. "Brrrrr! That's cold!" she told Maureen. "Too cold for a bath! You'll have to wait until there's more hot water!"

Unfazed by the aide's report of cold water, Maureen began to get undressed for her bath. "You can't take a bath, it's too cold!" the aide told her. "I don't care; I just want a bath," she persisted. "Listen, Maureen, you're going to have to wait; it's too cold to bathe!" scolded the aide. Maureen was angry and quickly getting angrier. She began to disrobe and move toward the tub. "No," said her aide, "you can't take a bath now!"

Another nurse aide heard the escalating commotion and joined them in the bathroom. She didn't believe it was worth getting into an argument or worse with Maureen over the water temperature of a bath. She told her fellow aide that she had a good rapport with Maureen and that she'd work it out with her. "I want to take a bath now!" Maureen shouted at her new aide.

"OK," said the new aide, "let me help you get in." Lifting her leg to get in the tub, her toe touched the water. "It's freezing! I can't take a bath in that! I'll wait for hot water!" she exclaimed. Problem solved; confrontation avoided.

DIFFICULTY WAITING

An experienced caregiver observed, "People with advanced HD can't wait." As absolute as that sounds, it's based on a sensitive observation of the difficulty people have when struggling to control their impulses. When they want something, they want it now. Their demands are driven by the damage to their brain caused by the disease. They may be impatient, unrealistic, angry, selfish, and imposing but, because of their impaired ability to inhibit themselves and to control their impulses when they can't do something, they just can't wait.

If someone asks for your assistance, give it to him right away or as soon as practically possible. As disruptive as it may be to you, it'll be more efficient for you in the long run. If you're unable to assist him right away, try to set a specific time when you will realistically be available to help him. For example, you might say, "I'll do that for you in fifteen minutes at four o'clock." Be sure to keep your promise! Do not leave your time frame open-ended by using phrases such as "as soon as I'm finished what I'm doing." This will frustrate both of you as he will inevitably reapproach you many times before you're ready to help him. Asking him to wait is asking him to do something that he may be neurologically incapable of doing. Always make the effort to anticipate what he'll need and eliminate the wait!

MISTAKING THE MOVEMENT DISORDER FOR MISBEHAVIOUR

It is not uncommon for some caregivers to misinterpret some disordered movements as "misbehaviour", or "inappropriate" or even "aggressive" behaviour. Consider these examples: You help Henry walk down the hallway holding onto a gait belt. Suddenly he slumps to the floor. You gently nudge him and ask him to get back up on his feet. As you attempt to assist him, you realise he's become "dead weight". Now back on his feet, together you take several more strides down the hallway... again he slumps to the ground, testing the patience of even the most understanding caregiver. You suspect he's doing this intentionally, perhaps "for attention".

As you watch and help Michelle to eat her lunch, you guide her hand to scoop a spoonful of potatoes off her plate. As she lifts the spoon toward her mouth, she drops it. The potatoes land on her bib, her lap, her tray, and on the floor. You clean them up a bit before you prompt her to scoop another spoonful. Halfway to her mouth, she drops the spoon... again! More on her bib, tray, lap, and floor. None in her mouth. Frustrated by the ever-increasing mess, the "wasted" food, and the suspicion that "she's not really trying," you may presume she's doing it "on purpose" and wonder, "How many more times is she going to drop that spoon?"

However, both Henry's slumping to the floor and Michelle's dropping her spoon are just as likely to be attributed to a phenomenon of the movement disorder called "motor impersistence", an inability to maintain a position. Henry may have been unable to maintain his upright position and Michelle unable to maintain her grasp on the spoon. This condition is an aspect of the movement disorder that is less well known. It is driven by the progressive changes in the brain and not by their personalities, mood, or character.

Similarly, you may be helping Shaun take a shower. Since he's got plenty to do just balancing himself and holding onto the rail for support in the cramped quarters of the shower stall, you gently nudge his elbow upward saying, "Let me lather you up under your arm here!" The next thing you feel is his elbow glancing up off the side of your head. You immediately show your disapproval, call for assistance, and end the shower, assuming Shaun tried to hit you with his elbow!

Another little understood part of the movement disorder is an inability to modulate or regulate the force of one's movement. So if Shaun intended to gently lift up his arm to let you wash under it, but was unable to regulate the force to lift it, his good faith attempt to help you help him looked more like he was trying to hurt you! When you understand these aspects of the movement disorder, you can arrange your position so that you won't be surprised or harmed by these big bursts of movement. Very often they are nothing more than an attempt to cooperate with the caregiver!

GETTING STUCK

It is common for people in the mid- and advanced stages to "lock onto" and "get stuck" on a topic. They might demand something from you or command you to do something... incessantly! It can range from asking you for a cigarette to your taking them to visit a friend... over and over and over again. They may compulsively insist that you help them in absolute disregard of conditions that make it impossible for you to do so. It may even escalate to shouting and swearing. Your explanations appear as unreasonable to them as their request does to you!

It's extremely difficult for him to stop "getting stuck", or "perseverating", just as it is for very young children with the same kind of behaviour. A few principles can help you manage repetitive or compulsive behaviours.

- Once the routine or rules are established, stick to them. If different caregivers respond differently to repetitive demands it is confusing to the resident.
- 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 few 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 you promise that you'll "do it in a minute" when he is stuck on a topic, "a minute" is, literally, 60 seconds. You might try setting a time much later to do it - a time which gives you plenty of time to do it. Agree to meet at that time, making absolutely sure you've got it done! Sometimes he will "lock onto" this later promised date and time and focus less on what he wants. However, a general rule of care is to do something that you're asked to do as soon as practically possible. Even though it may temporarily disrupt your activity and delay your helping someone else, it will be worth it in the long run. Avoid describing this "getting stuck" too casually as "agitation".

When someone is stuck on a topic, avoid saying "No" to them. A refusal risks needlessly angering them. For example, Virginia routinely smokes a cigarette every day after breakfast at 9:00 am. "Can I have a cigarette?" she asks at 8:45. Rather than telling her, "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 cigarette. Why don't you head into the smoking room!"

SWEARING & OFFENSIVE REMARKS

The combination of lack of impulse control and anger over loss of independence in a person with Huntington's may erupt in the form of offensive or prejudiced remarks and profanity directed at his caregivers. Even though we can understand that the foul language is fuelled by Huntington's disease, it still stings to be the target of such slurs. It takes great tolerance on our part to disregard them. Do not endure these words alone. Tell your supervisor what happened so that these hurtful statements can be addressed if they persist. Consistently polite bedside manner will eventually build bridges over his prejudice. Intolerance will be replaced by his trust.

IN SUMMARY

Some days, you may wonder why he stands in befuddlement when told to "get dressed and be ready in half an hour." Or you may look aghast when he takes a cigarette from another person's pack and lights it without ever having asked for it. It's easier to see how problems with balance can lead to falls than it is to see how difficulty thinking can affect behaviour: problems with recall; starting, organising and stopping action; and lack of impulse control. These symptoms of HD present you with unique challenges. By understanding how changes in the brain affect thinking processes, you can begin to find the causes of inappropriate behaviour. You can often find simple solutions to what seem at first to be difficult situations.

11. UNDERSTANDING CHANGES IN MOOD

DEPRESSION

Many people with HD experience depression at some time during the course of their disease. Depression is among the most treatable features of HD, responding well to medication, and in some cases, counselling. But, because some of the more typical signs of depression can also be attributed to the movement and cognitive disorders of HD, 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 appearance, irritability, and motor slowing 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. Here are some signs of depression to watch for carefully. Report these signs of depression to your supervisor or medical and mental health professionals:

- a lack of initiative and withdrawal
- a lack of interest and activity
- irritability
- a sad facial expression
- isolation
- 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
- a general slowing of activity
- talking about or attempting suicide. Any talk about suicide should be treated seriously. Immediately report it to the appropriate person.

ANGER

There are many reasons for a person with HD to be angry. A list of those reasons might begin with having HD in one's family, losing a parent to it, and losing the ability to support oneself. The list could include putting one's children at risk for HD, being dependent on others for care, and losing control of one's day-to-day activity. There is, indeed, much reason for anger. These reasons for frustration and anger, coupled with a neurologically based impulse control problem, create a tendency to become angry quickly, giving him a "short fuse". The anger can be extreme and frightening to those who see it. And it may be directed at you!

This is the time to give him space or a "wide berth"; that is, protect yourself and those nearby. Keep well clear of him. Do not attempt to reason, explain or persuade. This may further antagonise him. Try to figure out what triggered this angry outburst so that it can be avoided in the future. People who watch and wait to intervene learn that people with HD often "cool down" as quickly as they "heat up". When it's over, you need not be surprised if the person apologises to you, explains that, despite a great effort on his part, he lost control. Accept this most sincere apology.

IN SUMMARY

People with HD certainly have reasons to be depressed, but there may be a physical reason for depression. Depressive moods can be expressed as angry outbursts directed at you. Keep your perspective, and try to decipher the angry message. Above all, don't take it personally. No matter how it looks, it's probably not intended that way.

12. SMOKING

Half jokingly and half seriously, physicians who are experts in caring for people with HD have referred to HD as "one of the smoking diseases." Certainly a very large portion of people with HD smoke cigarettes. People with HD often view it as "one of the last pleasures I have left." Smoking becomes symbolic of independence.

Smokers and their caregivers are faced with a number of problems. Some folks with HD have an altered sense of hot and cold. Their fingers are often burned lighting cigarettes or smoking them down to the butt. Impaired judgement can make them unaware of the danger of burns to clothing, ashtray fires, or lighting the cigarettes of friends who are themselves unsafe smokers. The movement disorder makes it unsafe to use and dispose of matches and lighters. Impulse control problems may drive them to take another person's lit cigarette right out of his mouth.

There are a number of clever devices that allow people to continue to smoke safely. "Smoking robots" eliminate the need to light and handle the cigarette. However, supporting smoking with assistive devices only delays dealing with the inevitable issue of unsafe smoking. The unsafe smoker doesn't want to hear your suggestion to quit or your offer of more supervision. Caregivers are often caught in a power struggle. You may try to responsibly manage his smoking but he can't see the risk of burning himself or even worse!

If you anticipate this months ahead of time, or if you have the opportunity to begin a discussion about limiting or quitting smoking, take advantage of it. If the unsafe smoker has a lengthy spell of illness or a hospital stay that precludes his smoking, use it as an opportunity to encourage him to cut down or quit smoking. Although this may put the caregiver in a patronising position to the unsafe smoker, it may reduce or eliminate unnecessary risk of burns and fire for years to come.

Caregivers tips

- Take seriously the smoker's emotional need to smoke.
- Discuss the use of nicotine patches.
- Build a reward system to encourage him to quit.
- Use large ashtrays that are solid, sturdy, and untippable.
- Limit smoking to a well-protected area.
- Purchase nonflammable clothing, furniture, and floor covering.
- Install extra smoke detectors.
- Use a "smoker's robot". It holds a cigarette to prevent ashes and embers from being dropped.

13. SOME 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, is a classic sign of depression. Little sleep and a very high activity level while awake could be a sign of mania. If you see this, report it to the appropriate health care professional so a complete evaluation can be made.

Due to daytime fatigue in advanced HD, some folks accidentally fall into a cycle of napping during the day and then being unable to fall asleep at night. You can usually find the right balance between conserving energy during the day and being tired enough to sleep through the night. Try to help them maintain their "rhythm of life".

UNEXPLAINED SCREAMING

Certain people with HD persistently scream for reasons that are not readily apparent to their caregivers. Since they're unable to simply say why they're screaming, it's a challenge to their caregivers to figure it out. It could be a need that's been overlooked and gone unmet. They could be in pain or panic; hallucinating or heartbroken. They might be frightened, anxious, grieving or hurt and have just one way to express all these different feelings... screaming!

In the most advanced stages of HD, it may be related to medication or the cramps that come with the changes in their muscle tone. Try to figure out exactly what triggers the screaming. This will mean systematically trying one approach after the other, asking other caregivers and the family for ideas of what may be causing it.

Some people may also make frequent unusual sounds as they encounter problems coordinating their breathing: gasping, sniffing, grunting, slurping sounds, etc. Since these sounds are not under their control, caregivers should graciously tolerate them.

EXCESSIVE SWEATING, TEMPERATURE AND THIRST

People with HD may be more comfortable in surroundings that are cooler than typical, perhaps as low as 18° 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 their physician regarding potential problems with electrolyte imbalance and kidney function.

FREQUENT URINATION AND CONSTIPATION

He will have more and more difficulty thoroughly emptying his bladder, as his muscles become progressively uncoordinated. Increased thirst may lead to increased fluid consumption. This often causes him to sense the need to urinate more often than usual. Problems controlling impulses, coupled with the increased urges to urinate, often lead to him demanding to go to the bathroom over and over, often after just having urinated. Do not remind him that he just went to the bathroom. Do not ask him to wait. He may have an accident or only be further antagonised.

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.

SEIZURES

It is not uncommon for people with juvenile-onset HD to have seizures. Occasionally, those in the most advanced stages of adult-onset HD will have seizures, too. It is more likely, though, that you may see sudden, brief, involuntary jerks involving groups of muscles that are easily mistaken for seizure activity. These large muscle jerks are called "myoclonus" and usually are not treated.

HIGH FEVERS

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

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 his care is critically important to prevent serious progressive deformity. In the advanced stages of HD, his 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 may still have involuntary movements, he cannot change his position. The fluctuations in muscle tone and the involuntary movements make it difficult to prevent and manage contractures. Typical approaches such as orthoplastic splints can easily cause skin problems. More useful, especially for knee and elbow contractures, are newer air-assist splints which use air bladders for support. The "give" in the soft splint prevents skin from breaking down. There are also lightweight, washable foam-core splints that can be helpful in maintaining functional positions of the hands.

Since rigidity is typical in people with juvenile HD, contractures may be a problem earlier in their disease. People with adult-onset HD may have involuntary movements that progress into rigidity. People who develop HD in their early twenties may present with stiffness, slowness, and occasional involuntary movements. Over time all involunatary movements are slowed, and dystonia and loss of motor control dominate.

SEVERE CHOREA

Most physicians and physiotherapists familiar with HD tend not to treat chorea. Many people with HD who have taken medication to suppress their chorea feel that it is easier to live with their chorea than with the side effects of the medication used to suppress it. 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 for athletes can be used.

14. CARING FOR YOU!

Caring for this person with Huntington's disease is a challenge. It challenges family caregivers and professional caregivers alike. The disorders of movement, cognition, and emotion are ever-changing and progress slowly over many years. This greatly challenges the skills of even the most experienced, formally trained caregivers.

It's difficult to go through a progressive disease with anyone; to do it with someone your own age or younger, as is often the case with the person with HD, is even more difficult. It's hurtful to have someone with whom you work very hard direct his anger at you and hard not to take it personally. To have to allow someone whom you're trying to protect to take careless risks is very unsettling. It's also discouraging to realise 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.

Nobody can beat Huntington's disease yet, but 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 realising 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.

A CAREGIVER'S PRAYER

by Shirley Procell

Dear Father in Heaven,
Please help me today
To find courage and strength
And to watch what I say.

Make me gentle and kind
And have a big heart
Give me endurance, fortitude;
Let me be clinically smart.

Let the ones I care for
Know of my love,
Help them today
From Heaven above.
Amen.

P.S.
And oh, please, God,
Just one more request;
One little thing
That you can do best.
Grant us a cure
For this awful disease.
I'm asking for some
That can't drop to their knees;
But they are your children
Suffering their pain.
Asking all this
In God's name.

APPENDIX 1:

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APPENDIX 2:

HUNTINGTON'S DISEASE ASSOCIATION

Huntington's Disease Association of Ireland (HDAI) is a national voluntary organisation established by Huntington's Disease (HD) family members to provide consultation, information and individualised support to those diagnosed with HD, their families and their health care teams. The Association was formally launched in 1985 and was incorporated in 1988. HDAI is a registered charity CHY 10130. HDAI exists to provide a unique service offering comfort, information and support to all those affected by HD.

HDAI offers:

- A national information and support service based in Dublin which provides information and support to families; individuals at risk of HD; carers and health professionals; HDAI liaise with service providers; highlights the needs of members and creates awareness of HD. Confidentiality is respected.
- A Family Support Officer is available to meet family members in crisis.
- Access to counselling for those in need.
- Support group meetings/carers workshops in Dublin, Cork, Mayo/Roscommon and Limerick.
- An annual information meeting and respite weekend available to people with HD and their families.
- Publications including leaflets, booklets and articles covering the many issues specific to HD available for families, social care and health professionals on request.
- Information through a quarterly newsletter and annual magazine.
- The loan of a specialised HD Chair which helps protect against injury related to involuntary movements and debilitation.
- Talks and information seminars on request.
- Therapeutic treatments for patients in the mid-stages of HD.
- HD ID cards provided on request (free-of-charge) to people with HD.

For further information please contact:

Huntington's Disease Association of Ireland (HDAI)
Carmichael Centre, North Brunswick Street, Dublin 7.
Tel: (01) 872 1303. Freefone: 1800 393939.
Email: hdai@indigo.ie
www.huntingtons.ie

Symbol of the International Fight against Huntington's Disease

The main theme of this symbol of the International Huntington's Association is a logo depicting a head and shoulders, representing the threat of Huntington's Disease to both mental and physical capabilities. The reduced size of the inner image indicates their dimunition in the sufferer.

This symbolic design, also reflected in our cover design, appears as the flower of a growing, vibrant plant, and is protected within its leaves. The purpose, growth and development of the International Huntington Association is thereby illustrated, together with the increasing worldwide concern that this disease shall be tamed.

Huntington's Disease Association of Ireland

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