

■ *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At different times, you'll find he is distracted, confused, uncooperative, angry, and withdrawn.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 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 recognized 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 ORGANIZING 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; zipping 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 organized the information and actions required to complete them. Some folks have difficulty organizing 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 bureau. Posting schedules of daily activities and the time to do them may help organize 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 folks 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, minimizes 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.

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.

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.

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 realize 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 folks 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 (or kitchen)!"

SWEARING & RACIST 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 bitter racial slurs and profanity directed at his caregivers. Even though we can understand that the foul language is fuelled by Huntington disease, it still stings to be the target of racial 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. Racial 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, organizing 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.