

Understanding Behaviour in Huntington Disease: A Guide for Professionals

Third Edition

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Note of Appreciation to HDSA and authors

The Huntington Society of Canada would like to extend its sincere appreciation to the Huntington's Disease Society of America, and Dr. Johnson and Dr. Paulsen for their permission to adapt the information in this resource for those living in Canada.

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The Huntington Society of Canada would like to extend its sincere thanks to Meghan Andrews for her permission to use some photographs from her book entitled *Huntington Disease: Trials and Triumphs*. Meghan's book captures the "strength, courage and positivity in each of the families" she showcases. HSC chose some of her photos for this book that reflect those same qualities in families who are faced with this debilitating disease.

Note of Appreciation for Canadian Reviewers

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Preface

This book was written for health care professionals providing care to people affected by Huntington disease (HD). With the permission of the authors and the Huntington's Disease Society of America, it has been adapted to reflect the Canadian picture of HD.

Since *Understanding Behaviour in HD* was published in 2000, much has been learned about Huntington disease. Using the most recent findings, this guide outlines major behavioural patterns seen in persons with HD and explains our current understanding of their causes. Examples from the field are provided, as well as strategies for managing them.

One major set of findings has been the detection of HD behaviours in persons at-risk for, but not yet diagnosed with HD. Studying this pre-diagnosis, or "prodrome," period is providing new insights into the impact of the earliest stages of the disease on behaviour. The chapter on the stages of HD has been significantly revised to incorporate this new information.

Understanding behaviour in a person with HD requires placing our current clinical knowledge of HD within the larger context of the person's life. Factors other than HD can be the cause of responsive behaviours. Look for these triggers when considering management options. Because the causes of difficult behaviour can be complex, examples from the field are provided throughout the book to illustrate the importance of taking a broad view.

Addressing behaviour in HD is a dynamic process, as symptoms change over time as the person passes through different stages of the disease. Symptoms also vary from person to person, even within a family. For example, one person may develop a severe mood disorder, requiring multiple hospitalizations but have little motor disability at that time. The person's sibling may have debilitating motor symptoms at the same disease duration but no mood disturbance. While there are broad patterns of behaviour in HD, interventions need to be tailored to the person, as one person's symptoms may not necessarily resemble those of other relatives.

Information presented in this guide represents the best efforts of the authors to provide recommendations and strategies that are based on published guidelines and reviews, tempered by the authors' professional and clinical experience. As our understanding is constantly being improved and refined, it is recommended that the reader periodically visit the website of the Huntington Society of Canada (HSC) for information on the current best practices at www.huntingtonsociety.ca

Chapter 1

Huntington Disease: An Overview



I. Huntington Disease: An Overview

Huntington disease (HD) is a genetic neurological condition that presents three groups of symptom features: movement disorders, psychiatric/behavioural disorders and cognitive disorders. Symptoms in one area will affect the other areas. For example, changes to cognition will have an effect on behaviour.

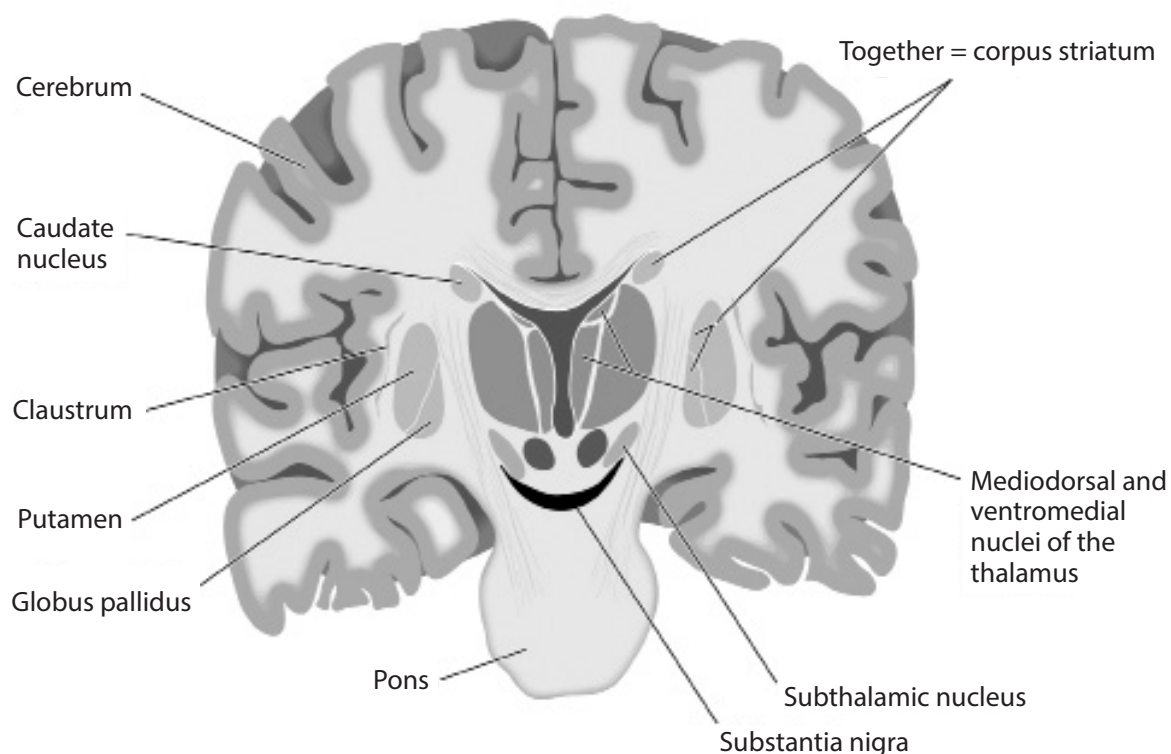
Common symptoms of HD are uncontrollable movements, abnormal balance when walking, slurred speech, difficulty swallowing, thinking difficulties, dysregulation of emotions and personality changes. Symptoms typically begin between the ages of 35 and 55 and progressively worsen until the affected person is unable to live independently. Presently there is no cure for HD and no treatment to slow the disease, but some motor, behavioural and psychiatric symptoms can be treated, and strategies for care in one area can sometimes help manage others.

Current estimates are that about 1 in 7000 persons in Canada has HD, 1 in 5500 is at-risk for HD, and 1 in 1000 is impacted by HD. Each child of a person with HD has a 50% chance of inheriting the disease from his or her affected parent. Although HD remains a rare disease, it is one of the more common hereditary diseases.

The Genetics of HD

HD is a hereditary neurodegenerative disorder caused by an expansion of a repeating CAG triplet series in the huntingtin gene on chromosome 4, which results in a protein with an abnormally long polyglutamine sequence. HD is one of a larger family of polyglutamine repeat disorders, all of which are neurodegenerative.

HD is an autosomal dominant disease, which means it affects males and females with equal likelihood. Each child of an affected person has the same 50% chance of inheriting the abnormal huntingtin gene, and therefore developing the disease one day. Inheriting a normal huntingtin gene from the unaffected parent does not prevent or counteract the disease-causing effects of the abnormal gene. In those rare cases where a person carries two abnormal copies of the gene, the person will develop HD, and each child has a 100% chance of inheriting an abnormal gene.

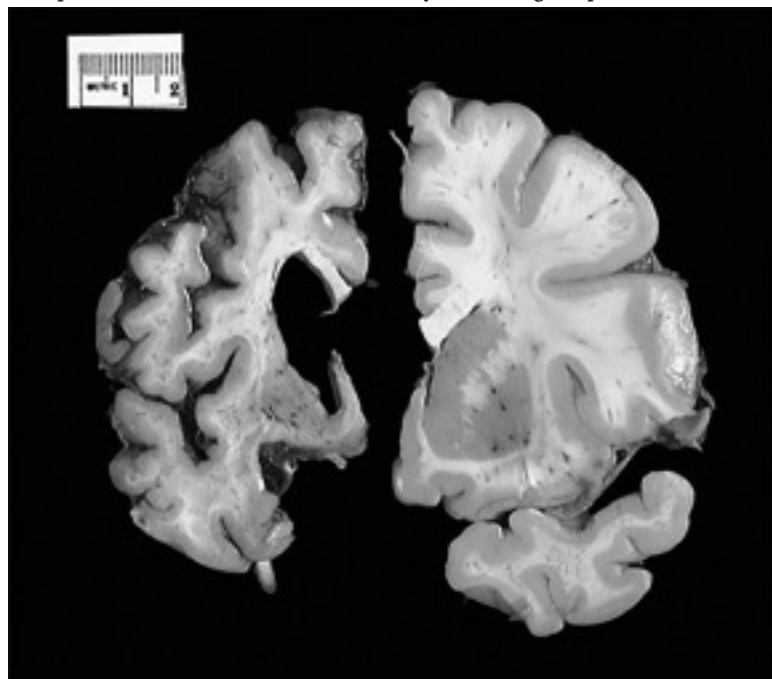


The normal function of huntingtin is not known, but the expanded polyglutamine sequence in the huntingtin protein is in some way toxic to brain cells. Certain neurons appear to be especially vulnerable. Atrophy is most marked in the corpus striatum of the basal ganglia, including the caudate and putamen. In later phases of the disease, other regions of the brain are also affected.

The Brain and Behaviour in HD

It is well established that different parts of the brain are responsible for different aspects of our behaviour. Damage to any region will cause disability in the associated function.

The part of the brain most affected by HD is a group of nerve cells at the base of the brain known collectively as



Brain with damage caused by HD on the left and a healthy brain on the right

the basal ganglia. Although other parts of the brain are also affected by HD, the basal ganglia appear to be the most heavily damaged. Within the basal ganglia are the caudate nucleus and the putamen.

The caudate nucleus is a collection of neuronal bodies that connect to many parts of the brain. The caudate organizes and filters information that is sent to the frontal lobe, which is considered the seat of executive functioning. The executive functions of the frontal lobes involve the ability to differentiate among conflicting thoughts; determine good and bad; better and best; same and different; future consequences of current activities; working toward a defined goal; prediction of outcomes; expectations based on actions; and the ability to suppress socially unacceptable urges.

The deterioration of connections within the caudate results in behavioural changes and the inability to control emotions, impulses, thoughts or movements. Damage to this area of the brain in HD is readily visible in MRI scans.

Damage to the caudate makes it difficult for persons with HD to prioritize tasks, stay focused and handle simultaneous stimuli. Persons with HD may exhibit a lack of self-awareness and the inability to evaluate their own behaviour, including a reduced ability to experience embarrassment, guilt or shame. Persons with caudate damage can be unaware of mistakes or inappropriate behaviours that are evident to others, which may result in getting stuck on one idea or activity in a show of perseverative behaviour. Many of these behavioural concerns are discussed in depth in Section III, Common Behaviour Concerns in HD, page 17.

Chapter 2

The Stages of **Huntington** Disease



II. The Stages of HD

In recent years, the growth in our understanding has changed how the progression of HD is characterized. Based on the new research, the stages of HD now include the time before a clinical diagnosis of the motor disorder. These precursor phases, which are typified in this handbook as At-Risk for HD, Person who is Gene Positive, and HD Prodrome, are now seen as preceding the five stages of Diagnosed HD.

Several assessment methods exist for measuring the progression of HD symptoms. One of the most commonly used is the Total Functional Capacity Rating Scale¹ (Figure 1, page 13) of the Unified Huntington Disease Rating Scale (UHDRS)¹. This scale rates the person's level of independence in five domains: occupation, managing finances, performing domestic chores, performing activities of daily living, and setting for level of care (living independently, nursing home). This score is used with the Shoulson and Fahn Staging Scale (Figure 2, page 14), to determine the stage of Diagnosed HD using a I–IV scale, with a lower number indicating a higher level of function.

Precursors to HD

At-Risk for HD

Persons who have a biological parent with a diagnosis of HD who have not been tested are considered at 50% risk for HD. For persons to be considered at-risk, no signs or symptoms of HD are present.

Person who is Gene Positive

Persons who have undergone a predictive genetic test for the HD-causing CAG repeat length and are found to have an expansion greater than 35 repeats are considered gene positive. They have the gene for the disease, but they show no current signs or symptoms of HD. These individuals are pre-prodromal.

HD Prodrome

The prodrome of HD is a newly described phase that has arisen from the detection and characterization of certain cognitive and behavioural symptoms in at-risk persons who are years from the appearance of the motor symptoms that are currently used for a clinical diagnosis of HD. Medical dictionaries define a prodrome as a clinical or physiological indicator that precedes the onset of disease. In the last decade, observational studies on the earliest stages of HD have revealed that the prodrome of HD includes some cognitive and behavioural symptoms indicating the presence of a disease process prior to the development of the full clinical syndrome. The prodrome may appear up to 15 years before the onset of motor symptoms. **This is significant because several studies have suggested that cognitive and behavioural impairments are greater sources of impaired functioning than the movement disorder in persons with HD, both in the workplace and at home.**

Two central indicators of prodromal HD are CAG repeat length and current age. Statisticians have developed formulas to estimate prodromal phases using these two indicators. Prodromal HD phases have been described as “estimated years to motor diagnosis” or “probability of motor diagnosis within the next five years” and the estimates have been referred to as “HD burden,” “disease burden” or “genetic burden.” Scientists debate about the best term for these estimates, since no currently used term is precisely accurate. Regardless of the label used, all research in prodromal HD now uses this “burden score” to estimate the participant's HD prodromal phase.

HD Prodrome A: LOW-FAR. The prodromal phase of HD can be said to begin when any sign or symptom of HD is noted in a person in the At-Risk or Gene Positive groups. In studies that separate the prodrome into two groups, this phase is referred to as pre-HD A. In studies where the prodrome is divided into

three groups, this phase is referred to as “far from onset” (> 13 years) or “low probability of diagnosis within 5 years” (< 60%). Research findings for this phase suggest that slight brain volume loss has begun, with the most prominent changes in the basal ganglia. The rate of volume loss over time is about 4% per year. Motor ratings can vary widely in this phase, with most having few or no motor symptoms and some showing motor abnormalities that are inconsistent or not yet severe enough to warrant a diagnosis. Cognitive difficulties are likely present but may not be noticeable to either the person with the gene expansion or to employers or family members. Performances are slowed and require greater effort. Fatigue is likely to be present. Emotion recognition may become impaired. Changes may be observed on the amount of time taken to complete tasks. Mood, anxiety and obsessive thinking may all be mildly elevated. Progression in this phase is very slow and rarely noticeable.

HD Prodrome B: MEDIUM-MID. The prodrome phase referred to as having “medium probability of diagnosis within 5 years” (60–85%) or being at the “midpoint towards motor diagnosis” (7–13 years) is typically the phase with the most variation in disease presentation. There appears to be a point in time when HD takes over, accelerates or becomes more aggressive, and it seems to occur 8 to 15 years prior to receiving a motor diagnosis. As a result, some persons in this phase are beginning the more rapid progression seen in the HD Prodrome C phase (described below) and some continue to progress slowly and more similarly to those in the HD Prodrome A phase. Cognitive and motor progression continues in the HD Prodrome B phase, and additional declines may include smell recognition and the perception of time.

HD Prodrome C: HIGH-NEAR. The prodrome phase referred to as “high probability of diagnosis within 5 years” (> 85%) and “near motor diagnosis” (< 7 years) is the phase with the most pronounced rate of decline in all areas studied. This phase is the one that should be used for testing new treatments, since change over time is significant and the measurement of every domain (motor, cognitive, MRI scan) is robust due to advancing disease. MRI volume changes are over 4% per year and changes in cognitive and motor scores are great. Most cognitive measures show decline during this phase, with the most pronounced impact evident in time needed to complete tasks, smell identification and time estimation, and with noted decline also evident in visual perception and decision-making.

Diagnosis and Post Diagnostic Stages of HD

Although HD is characterized by behavioural, cognitive and motor symptoms, the clinical diagnosis is based on the motor disorder. The main measure of motor impairment is the motor assessment section of the UHDRS, which is administered by a trained examiner. The first part of the motor exam consists of five domains of motor impairment, with individual items each rated on a 5-point scale ranging from 0 (normal) to 4 (most severe impairment). The five domains of the motor exam are: eye movement, chorea (jerky movement), dystonia (muscle spasm and twisting), bradykinesia (slowness in movement), and rigidity (stiffness). The sum total of all the 31 items is referred to as the Total Motor Score (TMS). The second part of the motor assessment consists of the Diagnostic Confidence Level (DCL), which is a single item with a 5-category ordinal rating scale. The examiner selects a response to the question: “To what degree are you confident that this person meets the operational definition of the unequivocal presence of an otherwise unexplained extra-pyramidal movement disorder (e.g. chorea, dystonia, bradykinesia, rigidity) in a subject at-risk for Huntington disease?” The rating categories are 0 = normal (no abnormalities); 1 = nonspecific motor abnormalities (less than 50% confidence); 2 = motor abnormalities that may be signs of HD (50–89% confidence); 3 = motor abnormalities that are likely signs of HD (90–98% confidence); 4 = motor abnormalities that are unequivocal signs of HD (99% confidence). Research ratings from the PREDICT-HD study suggest that persons at-risk for HD move through these ratings at different rates, but the average times reported are: up to 21 years in DCL = 1, nine years in DCL = 2, and about three years in DCL = 3 before receiving a DCL = 4, which provides a formal diagnosis of HD.

Once a person has been diagnosed with HD, it is common to assess the progression of HD with the Total Functional Capacity (TFC) rating scale of the UHDRS (Figure 1). This scale rates a person's functional capacity and level of independence in five domains: occupation, ability to manage finances, ability to perform domestic

chores, ability to perform personal activities of daily living, and setting for level of care. The TFC assessment is determined by a personal interview, with input from the family when available.

The TFC scores each domain on a range of 0 to either 2 or 3 (e.g. “Occupation: 0 = unable, 1 = marginal work only, 2 = reduced capacity for usual job, 3 = normal”). TFC total scores range from 0 to 13 with greater scores indicating higher functioning.

Note on the TFC in the HD PRODROME:

Two studies published by the Huntington Study Group suggested that functional capacity measures were not sensitive for HD in the prodrome. Over 88% of persons in the prodrome of HD scored at ceiling, showing no functional decline despite significant change in MRI volumes, cognitive decline, psychiatric and motor symptoms. These findings suggest that more sensitive measures of functional capacity will be needed to assess the impact of symptoms in the HD prodrome. (Beglinger et al., 2010; Paulsen et al., 2010).

Figure 1. Total Functional Capacity Rating Scale (Source: UHDRS)¹

Domain	Ability	Score
Occupation	Unable	0
	Marginal work only	1
	Reduced capacity for usual job	2
	Normal	3
Finances	Unable	0
	Major assistance	1
	Slight Assistance	2
	Normal	3
Domestic Chores	Unable	0
	Impaired	1
	Normal	2
Activities of Daily Living	Total care	0
	Gross tasks only	1
	Minimal impairment	2
	Normal	3
Care level	Full-time nursing care	0
	Home of chronic care	1
	Home	2
TOTAL	Range 0 - 13	

The Stages of Diagnosed HD

Many clinicians use the Shoulson and Fahn rating scale² (Figure 2) to stage diagnosed HD and follow its progression. This rating scale groups total TFC scores into five stages of disease, with lower stages indicating more intact functioning.

Figure 2. Shoulson and Fahn Staging Scale²

TFC Total Score	Stage
11 - 13	I
7 - 10	II
3 - 6	III
1 - 2	IV
0	V

HD Diagnosed STAGE I (0–8 years since motor diagnosis): Maintains only marginal engagement in occupation, having part-time voluntary or salaried employment potential, AND maintains typical pre-disease levels of independence in all other basic functions, such as financial management, domestic responsibilities, and activities of daily living (e.g. eating, dressing, bathing); OR performs satisfactorily in typical salaried employment (perhaps at a lower level) and requires slight assistance in only one basic function: finances, domestic chores or activities of daily living. Progression on this scale is about 1 point per year for both Stages I and II (Marder et al., 2000).

HD Diagnosed STAGE II (3-13 years since motor diagnosis): Typically unable to work, requiring only slight assistance in all basic functions: finances, domestic chores, daily activities; OR unable to work and requiring different levels of assistance with basic functions (some are still handled independently). Progression on this scale is about 1 point per year for both Stages I and II.

HD Diagnosed STAGE III (5-16 years since motor diagnosis): Unable to engage in employment AND requires major assistance in most basic functions: financial affairs, domestic responsibilities, and activities of daily living. Annual progression on the TFC is .38 point per year for this stage.

HD Diagnosed STAGE IV (9-21 years since motor diagnosis): Requires major assistance in financial affairs, domestic responsibilities, and most activities of daily living. For example, comprehension of the nature and purpose of procedures may be intact, but major assistance is required to act on them. Care may be provided at home but needs may be better provided at an extended care facility. Annual decline on the TFC is .06 point per year for persons in this stage.

HD Diagnosed STAGE V (11-26 years since motor diagnosis): Requires major assistance in financial affairs, domestic responsibilities, and all activities of daily living. Full-time skilled nursing care is required.

These stages offer very broad guidelines, and symptoms can vary a great deal in the course of the illness. For instance, one person remained in Stage I for 15 years and was able to remain employed (in a reduced capacity) at his regular job despite significant chorea.

Special Notes

Goals of Treatment for Persons with HD

Caring for persons with HD is both challenging and rewarding. Since there are no treatments that can slow, halt, or reverse the course of the disease, **the goals of treatment are to reduce the burden of symptoms, maximize function, and optimize quality of life.** By paying careful attention to changing symptoms and maintaining good communication with the affected person, their family members and other health care professionals providing care, it may be possible to lessen many difficult behaviours.

The Caregiver as a Source of Information

Persons with HD will often be accompanied by a caregiver to doctor visits. **A caregiver can be a crucial source of information about behaviour in the person with HD, particularly as the lack of "self-awareness" is a symptom of the disease.**

There can be a tendency in medical professionals to discount the reports of the caregiver as second hand or exaggerated, which is unfortunate, as the caregiver may see tendencies and behaviours that will not be seen by the doctor during the appointment. Persons who are gene positive in the prodrome tend to be reliable reporters in the very early phases, such as Prodrome A, but research suggests that companion reports may prove more reliable in persons who are of medium and high probabilities for motor diagnosis (Duff et al., 2010). The caregiver may not feel comfortable discussing certain issues, such as irritability, cognitive issues or sexual problems, in the presence of the person with HD. Therefore, an effort should be made to periodically speak to the caregiver alone, either during the visit or later by phone.

Alternatively, a small minority of caregivers will present incorrect information for non-altruistic or manipulative purposes. Additional assessments may need to be performed to confirm information provided by caregivers.

- 1 Huntington Study Group (Kiebertz K, primary author). The Unified Huntington's Disease Rating Scale: Reliability and Consistency. *Mov Dis* 1996;11:136-142.
- 2 Shoulson I, Kurlan R, Rubin A, Goldblatt D, Behr J, Miller C, Kennedy J, Bamford K, Caine E, Kido D, Plumb S, Odoroff C: Assessment of functional capacity in neurodegenerative movement disorders: Huntington's disease as a prototype, *Quantification of Neurologic Deficit*, T Munsat (ed), Butterworths, Stoneham, MA., pp. 271-283, 1989.

Chapter 3

Common **Behaviour** Concerns in **Huntington Disease**



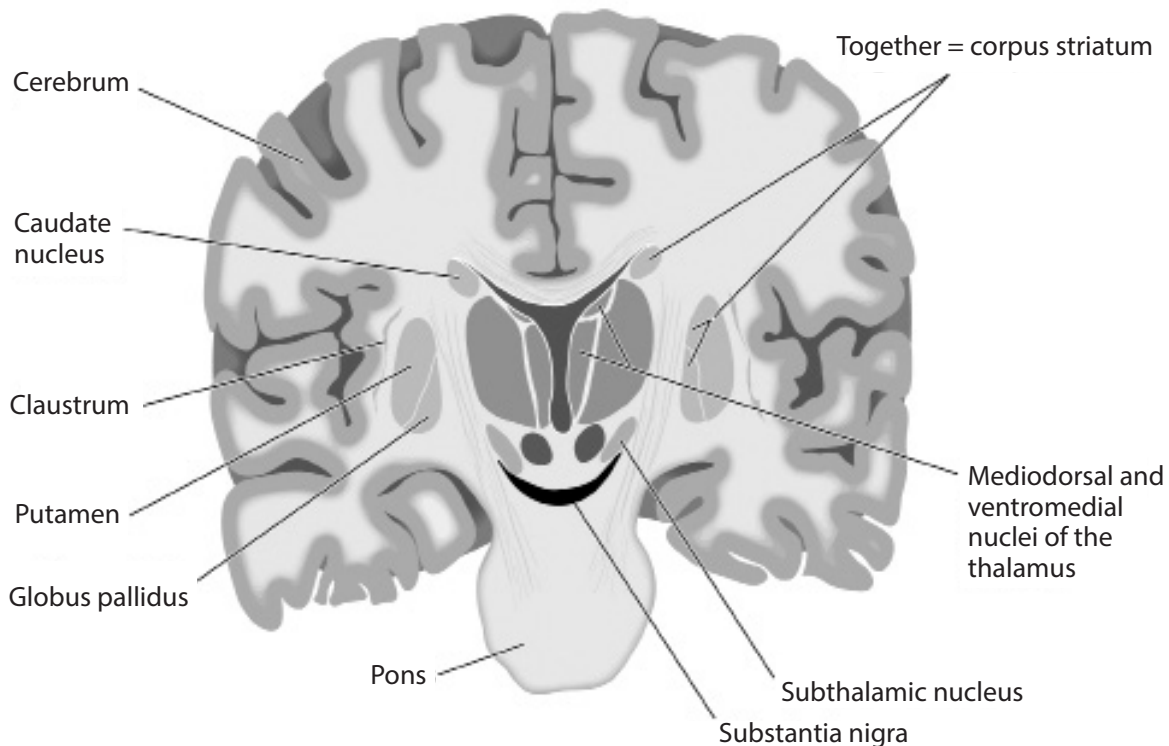
III. Common Behaviour Concerns in HD

Communication

Communication, or the transfer of information from one person to another, requires a complex integration of thought, muscle control, and breathing. Huntington disease (HD) can impair all three of these functions. Although there are many potential causes for difficult behaviours in HD, one of the biggest reasons for any problem is poor communication. Anger, irritability, disorganization, depression, anxiety and perception can all be made worse by communication difficulties. A recent study reported that a primary concern for persons with HD was the increased effort and concentration demanded to communicate and that high speed and initiation of output were the chief detriments to conversing (Hartellius, Jonsson, Rickeberg, & Laakso, 2010).

There are two main aspects to communication: getting the information IN (listening, processing and understanding) and getting the information OUT (expressing). Communication with a person affected by HD can be a difficult task because both aspects are impaired by the disease. The most prominent language difficulties in persons with HD are:

- Understanding (taking in language and processing it)
- Initiation (starting conversation)
- Organizing speech (processing and conveying thoughts)
- Articulation (speaking clearly)



Understanding

The ability to organize sensory input is affected by HD and leads to difficulty understanding or acting upon information. For example, a person with HD may understand each unique word without difficulty, but complex sentences and paragraphs become mixed up as the frontal lobes of the brain struggle to organize and prioritize the incoming information. The frontal lobes of the brain are richly connected through the basal ganglia (striatum, putamen and caudate) and HD specifically interrupts the frontal lobe function. **Care should be taken to simplify complex concepts or directions without talking down to the person with HD.**

Initiating

The ability to start talking is severely impaired by damage to the caudate in the brain. A person with HD may exhibit a lengthy delay when answering questions or may rarely begin a conversation.

Several common scenarios include:

- The listener assumes that the person with HD cannot answer the question and continues with yet another question
- The listener assumes that the person with HD cannot answer the question and makes the decision him or herself
- The listener becomes frustrated and/or fatigued and gives up
- The person with HD becomes frustrated and/or fatigued and gives up

Delays in answering can be problematic. Family members, caregivers and medical professionals may assume that the person with HD no longer has preferences (e.g. for food or activities) or is not motivated to participate, both of which may be inaccurate. **During medical appointments, it is essential to provide the person with HD adequate time to respond to questions.**

Organizing speech

HD affects a person's ability to structure thoughts and produce an organized output. Without the ordering and prioritizing function of the frontal lobes, thoughts and speech can become disorganized, tangential, random or incoherent.

Articulation

Difficulty in speaking clearly is one of the hallmark symptoms of HD. Slurred speech is often noted as well as difficulty maintaining a consistent volume. As HD progresses, phrase length decreases and pauses in speech output are extended (Rohrer, Salmon, Wixted, & Paulsen, 1999).

Possible Causes of Problems with Communication

The ability to communicate includes physiological, psychological and social components, and the progression of HD affects them all. Not only must the person be able to communicate but must also have the desire to speak and be a patient listener willing to hear what is said.

Psychological factors, such as mood symptoms, depression or anxiety, can make it harder for a person to communicate or decrease the person's desire to do so. Social factors can also influence communication. People may avoid speaking to a person with HD or may not allow time for the person to respond. People (even in medical settings) can be rude or insensitive. The person with HD may also isolate him or herself to avoid the difficulties of communication.

Whereas psychological and social factors influence communication indirectly, the physiological changes in the brain, caused by HD, contribute directly to problems with communication. The primary area of the brain that is damaged by the disease is the caudate nucleus. Articulation is impacted when damage to the caudate causes a breakdown in the neuronal pathways or circuits that relay information through the caudate. The motor circuit tells the body how to move specific muscles at precisely the right time. Should one muscle group fire too early or too late, speech articulation will be garbled. A consequence of the deterioration of the caudate is that some motor movements occur randomly (chorea). This impacts articulation because talking requires a complex series of motor movements of the lips, mouth, tongue, and controlled breathing. Without the caudate to regulate all of the different aspects of speaking, speech becomes disorganized, poorly timed, and not adequately supported by breath.

The damage that occurs in the caudate is due to the degeneration and death of the medium spiny neurons (brain cells). As neurons die, information cannot be passed along the circuit as easily. This reduces the brain circuit's ability to communicate, and the initiation of speech becomes more and more impaired. The person with HD will experience increasing difficulty simply finding the words to express thoughts and may take much longer to respond.

Organization of sensory input also becomes more complicated as the caudate loses the ability to regulate information coming into the brain. Although the person with HD can hear normally, the components of the information become jumbled and mixed up without the caudate and frontal lobes to keep them separated. What might seem to be a simple request from a family member or caregiver may be too complex for the person with HD to start and complete without assistance. Instructions may be too complicated or not detailed enough, as people often underestimate the complexity of a given task. For instance, the simple act of getting dressed involves many steps (e.g. decide what to wear, consider the weather, select clothes, assess colors and patterns for matching, and then put them on in the correct order – pants before shoes).

Finally, as the caudate deteriorates, access to the frontal lobes, which help to sequence, order, and prioritize, is compromised. The ability to organize and regulate outgoing information is disrupted. Thoughts and speech are garbled.

Examples

- A woman with HD reported going to a liquor store to buy a bottle of wine as a gift for the host of a dinner party and was denied service because the owner said she “already sounded drunk.”
- The wife of a person with HD stated that she was becoming increasingly frustrated because her husband “wasn’t answering” her when she asked him questions. When asked for an example, she said that when she asks him what he wants to eat for dinner, he does not respond.
- While asking his wife with HD about her day, a man becomes frustrated because she doesn’t answer. Eventually, he stops asking and begins talking to their children. Several minutes later, his wife interrupts and answers the question.
- A woman reports to her husband’s doctor that he doesn’t make sense. “Sometimes he will start talking and it’s like he is trying to talk about everything at once.”
- While talking to her brother in public, the speech of a woman with HD varies from almost a whisper to almost a shout. He states that he gets frustrated and finds that he is often either telling her to speak up or to “keep it down.”

- A woman in a skilled nursing facility frequently goes to the nursing station to talk to the staff member present. Over time, her speech has become quite difficult to understand. If the staff member does not respond, the woman becomes increasingly agitated, which then makes her speech even harder to understand.
- A man who is in the later stages of HD is bedbound and remains quiet almost all day, every day. Occasionally he will cry out, as if he is trying to scream, but the sound is garbled. Staff is becoming worried, as they are unable to determine any possible trigger, and note that there is no pattern related to feeding or toileting.

Strategies

Over time, the communication and speech difficulties associated with HD will impact a person's level of function in many areas. Physiological and psychological issues will make it increasingly difficult to participate in important discussions and decisions. Encourage the person with HD, their family and caregivers to establish good communication practices early in the progression of the disease.

Speech therapy can be a useful tool for persons with HD who are having trouble with articulation. Loss of regulation of the mouth and tongue, as well as breathing, can lead to speech deficits. Techniques utilized in speech therapy can help to address these concerns.

The strategies for dealing with communication issues listed below are basic, and when used, help address the four underlying factors leading to problems with communication: articulation, initiation, understanding and organizing.

- When asking questions, allow the person with HD enough time to answer or express him or herself. It is not necessary to speak slowly. Keeping directions or commands to one or two steps can be helpful as working memory is often diminished by HD.
- Advise family members and caregivers to practice patience when requesting a response from the person with HD.
- Offer cues and prompts to help the person with HD to start speaking or answer a question, but pay attention to how the person with HD is responding to this assistance—if it is creating tension, try giving a little more time for him or her to answer.
- When asking a question, offer limited choices instead of asking open-ended questions. Questions can be phrased in a “yes or no” or a “this or that” format.
- If the person with HD is confused by a conversation, modify what is being said. Simplify the conversation and make it short.
- If the family member or caregiver does not understand what the person with HD is saying, encourage them to ask him to repeat what is not understood. If this does not work, try alternative methods, such as asking the person with HD to spell the word that wasn't understood or to give the first letter.
- When delivering a task or a set of instructions, break it down into small steps. Remember, many seemingly basic tasks are actually very complex activities. Modify the steps of a request as the person with HD becomes more impaired.
- As communication becomes more impaired for the person with HD, consider using simple words and short sentences. Be careful not to talk down to the person with HD or use baby talk.

- In late stages of HD, alphabet boards, yes-no cards or other technical communication devices can be considered. Due to motor symptoms in HD, these tools are not appropriate for all, so be conscious of the person's symptom spectrum.
- Once a person with HD loses the ability to verbally communicate, do not stop talking to them. This only intensifies feelings of isolation that accompany the loss of the ability to speak.

Communication problems become more pronounced as HD progresses. In the earlier stages, a person with HD might notice difficulty with organization of speech and thoughts. As the disease progresses, the person might experience problems with initiation, understanding and articulation. These aspects of communication continue to deteriorate over the course of the disease. Recent reports emphasize the importance of early assessment and the introduction of assistive devices throughout the disease course (Saldert, Fors, Ströberg, & Hartelius, 2010), (Ferm, Sahlin, Sundin, & Hartelius, 2010).

Some professionals have described the person in the later stages of HD as “locked in,” meaning that he or she is unable to communicate despite having a relatively normal understanding of his or her surroundings. **A person with late-stage HD may not speak, but may still hear what is said and be able to take in information and process it, albeit at a slower rate.**

Learning and Memory

Learning is the ability to acquire new or modify existing knowledge, behaviours, skills, values or preferences. This process often involves combining multiple sources or synthesizing different types of information. Memory is the ability to recall previously learned information. Persons affected by HD will experience problems in learning new information or in recalling encoded information. The reason for this deficit is that HD disrupts the search mechanism used to find the desired information – it is still there, it is just harder to retrieve, unlike Alzheimer's disease, where memories are truly lost.

Another aspect of memory impacted by HD involves implicit memory, the type of memory used to collect and sequence a series of coordinated skills or movements, such as tying your shoes or riding a bike. Implicit memory often involves motor memory of how to perform an action or activity. Explicit memory, which has to do with specific facts or information, like names or dates, is more reliable for the person with HD.

Possible Causes

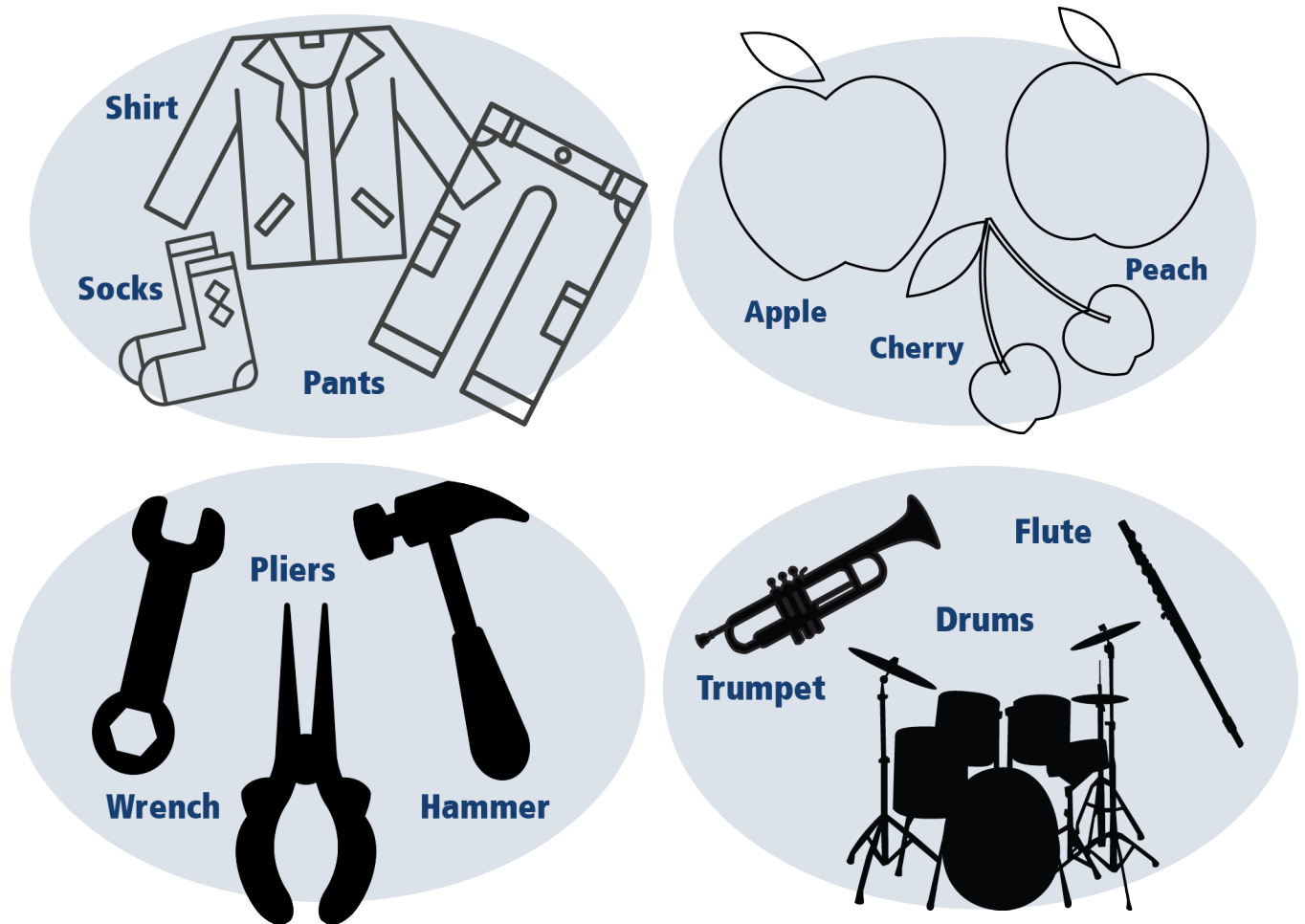
Problems with learning appear to be due to the disruption of the circuits connecting the frontal lobes and the caudate in the brain. The frontal lobe of the brain is the area responsible for higher order functions, or executive functions, of thinking and emotional regulation. Because of the disruption in this area, a person with HD can experience impairments in the ability to organize and sequence the information to be learned. When information is not organized in an efficient manner, retention and recall of the information is very difficult.

Learning may also be impaired as the damage to the caudate makes it more difficult for the person with HD to use divided attention. Divided attention is necessary to attend to more than one thing at a time. When the caudate becomes unable to filter or regulate information as it travels through the brain, it becomes impossible to use divided attention because all the information tries to come through the circuit at the same time, causing overload.

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

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

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



There are different aspects of memory impacted by HD. Recognition, or the ability to identify previously learned information, is less impacted than a person's free recall, the ability to easily elicit information from stored memory. In addition, explicit memory (recollection of names, dates) is less affected than implicit memory (recall of a collection of coordinated skills, movements). Long-term, older memories are less impaired than short-term memories. The literature shows that memory is one of the most prevalent cognitive domains studied in HD and accounts for about one-quarter of the published cognitive literature in HD (Rohrer et al., 1999; Dumas, van den Bogaard, Middelkoop, & Roos, 2013).

When it comes to learning and memory in HD, it can be helpful to compare some of the problems of persons affected by HD with those affected by Alzheimer's disease. Even though both HD and Alzheimer's disease are commonly referred to as dementia, the following table points out the numerous differences between these two disorders. For instance, persons with Alzheimer's disease have severe problems remembering information that was learned previously. A person with HD will often be able to recall the word or fact when offered an either/or choice from which they can recognize a piece of information.

Ability	Huntington disease	Alzheimer's disease
Speed of processing	Slow, but relatively accurate	Slow, often inaccurate
Speech output	Slurred and slow, but accurate	Normal in clarity and rate; often the incorrect word
Learning new information	Disorganized and slow, but can learn	Rapid forgetting, defective storage of information
Free recall of memory	Impaired, cannot find the right word; can recognize with choices, benefits from cues	Impaired, memory store is defective; cannot recognize, cues don't help
Motor memory	Impaired, cannot learn or recall motor memories	Intact, can learn and retain motor memories

Both the ability to learn and the ability to remember can be impacted by factors other than HD, such as psychiatric symptoms and environmental influences. For example, depression and anxiety can impact a person's ability to encode new information. In addition, a stressful environment may make it more difficult for a person with HD to process new information. Finally, other cognitive changes can complicate both learning and memory, like organizational difficulties or impulsivity.

Examples

- A woman with HD tells her doctor that she is having a hard time learning a new accounting system that her company switched over to recently. Although many people at the firm report difficulties with the new system, she feels it is taking her an exceptional amount of time to figure it out.
- At his last clinic visit, a third grade teacher with HD stated that "each year it gets harder to remember all of the kids' names."
- A woman reports her husband with HD will go to the store to buy two or three things, then call home to be reminded what he was supposed to buy.
- When doing the mini mental status exam, a woman with HD is not able to remember the three memory items, but identifies all of them correctly when they are presented in multiple-choice form.
- A woman with HD who lives in a care facility says, "I don't know," almost instinctively when asked questions about her day. If another question is not asked right away, she will eventually offer additional information on her own.

Strategies

There are a number of methods used to work with changes in learning and memory associated with HD. Some of the recommendations are mentioned in the section about communication, as techniques and interventions are similar across behaviours. The following are strategies for addressing issues with learning and memory to share with family and caregivers.

- Breaking a task into simple steps can improve performance in a person who has difficulties with learning.
- Checklists and written reminders can be helpful for both learning and memory. A visual reminder can help direct the process. Sticky notes, dry eraser boards and written lists can be used as visual cues to

spur an action. If using a written list, recommend that the caregiver review the list with the person with HD to be certain each item is understandable.

- Technology may be able to assist with reminders. Caregivers can send a text message or set up a calendar or alarm on a tablet or cell phone. Difficulties with movement and cognition may make some technology too frustrating to be effective.
- Encourage the person with HD to practise with the new information. Repetition can help with understanding and retention.
- Offer simple alternatives or a limited list of choices to assist with recall. Either/or questions often work well. Avoid asking open-ended questions.
- Keep day-to-day activities as routine as possible. Routines and schedules give a person with HD the opportunity to practise a sequence of behaviours or activities and become accustomed to them. An example of a routine is getting up every day at the same time, taking a shower, and having breakfast. The more the person with HD does this sequence, the more likely he is to learn it and repeat it in the future.
- A person with HD may have a more difficult time retrieving information, but that does not mean that he or she can't. It cannot be said enough - when asking a question, give the person enough time to respond.

Perception

Perception is the ability to interpret and organize sensory information in order to understand and interact with the environment. Research has shown that difficulties with perception can arise in the early stages of HD, approximately 8-15 years prior to the movement disorder.

Symptoms of perceptual problems may not be easily evident in office visits, although they may interfere significantly with the person's home and work life. **Reporting from family and caregivers may be the way that the physician becomes aware of these impairments.**

Perceptual problems that are seen in the prodrome of HD, and which can cause problems over the course of the disease, include impairments in the following:

- Emotional recognition
- Perception of time
- Smell identification
- Spatial perception

Emotional Recognition

The earliest cognitive impairment detected in people in the prodrome stage of HD is the ability to recognize emotions. Persons with HD begin to inaccurately identify which emotion is being communicated in a facial expression. Research has shown that persons with HD when asked to identify whether a facial expression represented fear, sadness, or happiness exhibited significant impairment. It is hypothesized that this early impairment may be associated with growing difficulties in social relations. Understanding of emotions and memory for emotions is intact; it is the identification of emotion, based on the complex processing of the face that becomes difficult. Decreased emotional recognition will impact a person with HD's ability to communicate, as this directly affects the person's ability to read non-verbal cues. A review of research around emotion in HD

found that early defects of emotional recognition (primarily for negative emotions) were replicated across studies (Calder, Keane, Young, Lawrence, Mason, & Barker, 2010) and were shown to progress steadily throughout the disease course (Paulsen, Smith, & Long, 2013).

Perception of Time

Research has suggested that persons with HD have difficulty in estimating time well before the motor diagnosis. When persons in the HD prodrome were asked to keep a metronome going after the cued tapping was discontinued, their internal “clock” or timer was inconsistent. Family members and caregivers often note that the person with HD, who was once punctual, is now often late and underestimates how long activities will take. Numerous studies have demonstrated a timing defect in HD that progresses with disease course (Rowe, Paulsen, Langbehn, 2010). These findings are also consistent with animal models of striatal degeneration (Jin & Costa, 2010).

Smell Identification

Persons with HD, as well as persons in the prodrome of HD, performed in the impaired range on a test of smell identification. Although they were able to detect the smells, they were less able to identify what they smelled. Performances on traditional memory tests were intact although smell identification was impaired. The olfactory system appears to be impacted early in the progression of HD. Problems with smell identification are often noted in relationship to hygiene and, in some cases, personal safety, for example forgetting to turn off a gas stove. Impairments in smell have been known for nearly two decades (Nordin, Paulsen, & Murphy, 1995) and performance losses in this circuitry have received a great deal of attention as a marker for many neurodegenerative diseases.

Spatial Perception

The mental manipulation of personal space becomes impaired as HD progresses. For instance, the judgment of where the body is in relation to walls, corners or tables may be impaired, resulting in falls and accidents. This decreased spatial perception is directly related to increasing risk of falls and injuries from running into objects (Stout, Queller, Baker & Borowsky, 2013; Paulsen et al., 2013)

Possible Causes

Each of the four perceptual problems associated with HD discussed above can be related to changes in the brain. As discussed in other sections, the primary area of damage in the brain is the caudate nucleus. However, there is evidence that HD has a wider impact on the brain, even in the prodrome stage of the disease. Emotional recognition is likely affected by changes to the cingulate cortex (Reading et al., 2004) and insular cortex (Rosas et al., 2005). Changes in the ability to perceive time is linked to a neurophysiological system including striatal medium spiny neurons, the dopamine system and the supplementary motor area (Mattel & Meck, 2004). Smell identification difficulties are linked to changes in the olfactory system (Nordin, Paulsen, & Murphy, 1995). Finally, spatial perception limitations are related to changes in both the caudate and cortical structures. As more research is done on perceptual changes due to HD, the complex neurophysiological systems impacting perception deficits will be better understood.

Examples

- The husband of a woman with HD reports that, unless he says something directly, she does not pick up on his nonverbal cues and facial expressions. “I had no idea you were angry!”
- A man with HD reports an increase in general clumsiness and frequency of running into things.
- A woman with HD is upsetting people in a waiting room because she is not respecting their personal space.

- The son of a woman with HD states that he is worried about her cooking for herself. “Not because she might burn herself, but because she has left the gas on a couple of times and claims she can’t smell it.”
- The husband of a woman with HD complains about his wife’s tardiness. “She tells me she will be ready to go in 30 minutes and it takes her at least an hour.”
- A man with HD reports coming down with food poisoning after eating some lunch meat that had gone bad. His wife was shocked that he had eaten it because she said it had a strong, unpleasant odour.

Strategies

The changes in perception experienced by a person with HD can have an effect on a number of areas, including personal relationships and increased safety concerns. Behavioural strategies may improve a person’s functional capacity. Assess the impact that perception changes have on a person’s overall function and treat the behavioural symptoms that might arise. The following are some strategies to share with family members and caregivers.

Emotional recognition problems can be especially detrimental to relationships.

- The person with HD may no longer be able to understand emotions but can respond to what you tell them. Clearly state your feelings in calm language.
- Engage in working dialogues with the person with HD. This will involve active listening, clear expression of thoughts and feelings, and a mutual focus on the issue at hand.
- Consider couples or family counselling as a tool to practise and improve communication strategies. Establishing a foundation of clear communication early in the disease will make it easier to interact as the disease progresses.

When dealing with time perception issues, many of the techniques recommended for learning, memory and organization apply.

- Try to allow the person with HD extra time to complete tasks and avoid short deadlines. Take into account the level of function when requesting something of the person with HD. Avoid creating time pressure when possible.
- Map out timelines and create lists for the person with HD. Reminders such as whiteboards, calendars, tablets and phones can provide visual triggers to help the person with HD maintain a schedule.

Issues with smell identification are usually noted in relation to hygiene and safety.

- If the person with HD is not aware of his or her own body odour or uncleanliness, gently remind or encourage him or her to wash. Present this concern calmly and respectfully, because often the individual is unaware and may become embarrassed.
- In later stages of the disease, the person with HD may not only be unaware, but may not care about hygiene. Setting regular times to bathe and change clothes can be helpful.
- A concern that arises related to smell identification is safety when using the stove. Whether it is solely related to not being able to smell the gas or is compounded by attention and memory concerns, this problem can be addressed by putting up a visual reminder, for example a sign reading, “Make sure the burner is turned off.”

Spatial perception problems are primarily addressed by making changes to the environment in which the person with HD lives. The following recommendations can be suggested to family members or caregivers:

- Minimize clutter in the home. Remove obstacles and decrease visual stimuli so it will be easier for the person with HD to navigate his surroundings.
- Keep clear pathways through rooms. Allowing adequate space for the person with HD to move safely through the home will decrease fall and injury risks.
- Consider carpeting the floor. Carpeted floors provide a softer landing for falls than wood or tile floors. In addition, the surface offers greater traction. Avoid unanchored area rugs, which present a tripping hazard.
- A person with HD may not be able to accurately judge where he or she is in space and may collide with the furniture. Pad sharp corners to reduce the risk of injury.

Executive Functions

Executive functions are a group of cognitive functions that are critical to a person's ability to care for him or herself. These functions are instrumental in a person's ability to manage work and family life, as well as manage activities of daily living.

Executive functions fall into at least four categories: organization, self-regulation, attention and problem-solving. These four categories include a long list of specific skills, many of which people take for granted. Skills that fall under the umbrella term of executive functions include the following: attention, concentration, thought organization, planning, sequencing, prioritizing, initiation, follow-through, mental flexibility, problem-solving, creativity, abstract thinking, decision-making, judgment, and controlling one's feelings. Due to changes in the brain caused by HD, many of these skills decline over time.

As discussed earlier, changes in executive functions occur when the frontal lobes, or their connections with the caudate, are damaged. When this occurs, even simple tasks can become difficult and frustrating. The literature in executive functions in HD is vast and includes over 160 published research studies (Dumas, van den Bogaard, Middelkoop, & Roos, 2013; Paulsen, 2011; Paulsen, Smith & Long, 2013).

There are many different skills that fall under the umbrella term of executive functioning, but some of the concerns related to executive functions that are most commonly reported by persons living with HD and their families include:

- Apathy and diminished initiation
- Difficulties with organization
- Impulsivity
- Irritability and anger
- Denial and unawareness
- Perseveration

Each of these concerns will be discussed in greater detail in the following sections.

Apathy and Lack of Initiation

Apathy is synonymous with inattention, indifference, and lethargy. Apathy is present when the person with HD seems to have less concern for things about which they used to demonstrate a great deal of care and concern. A person with apathy might appear to be disengaged or not emotionally invested in what is going on around them. Apathy is prevalent in HD and impacts a majority of persons at some time during the disease. One study reported that over 20% of persons with HD showed apathy only and 32% showed depression only, whereas 38% had both apathy and depression at the time of study (Levy et al., 1998). It appears that apathy progresses with disease severity and is associated with cognitive decline in HD.

A lack of initiation often accompanies apathy but can also occur by itself in the absence of apathy. The ability to initiate behaviours, conversation, or activity is a very complex function which is frequently compromised in persons with various types of brain dysfunction, including mild head injury, Parkinson's disease, multiple sclerosis, stroke and, of course, HD.

Possible Causes

The circuits in the middle and bottom sections of the frontal lobes are connected to the limbic system, which is the “emotional lobe” of the brain. As the caudate nucleus degenerates, these circuits can become weaker, causing the frontal lobe to be disconnected from the feelings of the brain.

Apathy can easily be misread as depression. A person who is depressed may also have reduced concern and energy for aspects of life. However, a person with HD can be apathetic and not be depressed. **It is important to try to differentiate primary apathy from depression because treatments can differ, particularly if using medications.** Apathy may indicate the need for a thorough psychological assessment for depression.

Examples

- A man with HD reports that he is having a hard time getting projects completed at work. He denies any difficulty in understanding the tasks or being able to organize the information but states that he has a problem “getting them started.”
- A husband reports that his wife, who shows symptoms of early stage HD, “isn’t doing anything at home.” He states that they talk regularly about her getting involved in an exercise program, but she hasn’t made it to any on her own. He does note that if one of her friends takes her, she reports that she enjoys the class.
- A woman reports that her husband with HD is lazy. “I tell him every day when I go to the office that he should do some of the chores around the house, especially now that he is no longer working. He never does.”
- The nurse at a long-term care facility tells a woman with HD that her family is coming to visit and she needs to get out of bed. When he returns 30 minutes later, she hasn’t moved from bed yet.

Strategies

There are several recommended medical treatments for apathy. In some cases, stimulants are useful; however, there is abuse potential of these medications and the possibility of increasing irritability and agitation. Antidepressants may offer some benefit but are more helpful if there are concurrent symptoms of depression. Please refer to HSC's *A Physician's Guide to the Management of Huntington Disease* (third edition) for additional information on medications. For non-physicians, consult with a medical doctor regarding pharmacological interventions.

Behavioural interventions can be successful. Typically, once a person has started an activity, he or she will participate. Often it is the person doing the initiating—the spouse, friend or caregiver—who needs support and encouragement. The following are some strategies to share with family members and caregivers.

- Apathy and a lack of initiation are part of the disease. Telling the person with HD that he or she is lazy will only increase tensions. Try not to interpret a lack of initiation or activity as laziness.
- Educate other people about the inability of the person with HD to begin or initiate behaviours and encourage them to include the person in activities.
- Seek counselling to address the stress and frustration of dealing with an apathetic person.
- Gently guide behaviours, but emphasize that when the person with HD says “no,” it means no.
- Use calendars, schedules, and regular routines. These can provide a reminder about activities. Cell phones, smart phones and tablets can be used to set reminders or alerts. Phone calls and texts from family members and caregivers can encourage participation.
- Sometimes lessened initiation is accompanied by mild organizational difficulties. A suggested activity may require a series of tasks that are overwhelming or difficult to organize. The person with HD may need extra help in order to participate.

Organization

Organization involves the planning, sequencing and prioritizing of information. Difficulties in this area can affect a person’s attempts to finalize a project at work, to complete a list of household tasks, or to apply for social security benefits. As HD progresses, difficulties in organization will impact a person’s abilities to conduct basic activities of daily living, as many everyday activities require a complex set of steps. Examples are tying one’s shoes or making lunch.

These same organizational impairments affect problem-solving, logical thinking and deductive reasoning. When the brain cannot sequence bits of information, many aspects of intellectual, social and personality functions are impaired.

Possible Causes

Organization is an executive function, or a higher order cognitive process, which is impacted by changes in the frontal lobes and its many connections with the basal ganglia (caudate and putamen). While connected to many areas of the brain, the caudate has the most connections with the frontal lobes. Therefore, as cells in the caudate nucleus die, neuronal transmission to and from the frontal lobes is affected.

Examples

- A person with HD reports that it is getting harder to plan the day; for example, “It’s difficult figuring out each of the kids’ schedules and who must be where at what time.”
- Work assignments have become more difficult for a person with HD, especially when she has to meet with people from a number of different departments who are all collaborating on a specific project.
- The wife of a person with HD reports that when she tells her husband to do things around the house, it now takes him a lot longer and sometimes he doesn’t finish, no matter how easy the task may be.

- A new person with HD has come to clinic with a folder that is packed with loose papers. When trying to provide his medical history, he repeatedly looks through the papers, but never seems to find what he is looking for.
- A woman with HD who lives on her own reports she filed for disability six months ago. With the assistance of a social worker, it is determined that her application was not filed because it did not include the basic information requested. The woman says she may have received a reminder letter or two, but she hadn't really gone through her mail in a while.
- A caregiver reports that the person with HD is unable to make simple decisions. "The other day I told her to hurry up and get dressed. Ten minutes later, she was still standing in front of the closet and hadn't done anything."
- A man with HD had been at the care facility for about four years when the staff noticed that he was not answering questions. "He will answer you. It's just that he does it after you have already moved on to something else." During the exam, it became obvious that the man was having a harder time formulating his responses.

Strategies

There are no medications specifically for organizational difficulties. Problems with organization can be addressed with behavioural techniques, using common tools as reinforcers. Techniques will vary based on the stage of disease and level of function of the person. The following are some strategies to share with family members and caregivers.

- Limit the amount of information the person with HD is presented at one time to allow for slowed cognitive processing speed.
- Maintain regular schedules and routines as much as possible. Consistent and structured schedules can make it easier for a person with HD to stay organized and engage in daily activities.
- Use calendars, white-boards and to-do lists that can be easily referred to during the day. Calendars and lists should be kept in one place in the home.
- Review schedules and routines frequently and keep them up to date. Set a dedicated time each week to look over the person's schedule and calendar with the entire family and caregivers.

Impulsivity/Disinhibition

Impulsivity and disinhibition are seen when the person with HD has difficulty regulating or controlling emotional responses and impulses. Impaired impulse control may be the reason that some persons affected with HD easily lose their temper, begin to drink too much, or have inappropriate sexual relations. Disinhibition usually presents as the person having trouble controlling a sudden desire to do or say something that comes to mind, even when the behaviour is hurtful, repetitious, or socially or sexually inappropriate. Also, disinhibition can sometimes contribute to illegal behaviours, such as stealing.

Possible Causes

Damage to the caudate nucleus or circuits connecting the caudate and the frontal lobes can contribute to impulsivity. One of the primary functions of the caudate is to regulate the information received from other areas of the brain. As the caudate is affected by HD, the control mechanisms of the brain break down. Without the

caudate, the brain cannot regulate how much movement, feeling, or thinking is required in a certain situation. This can lead to the person with HD receiving an overwhelming flood of input. The brain may become over-ridden with messages to act, resulting in responses that are not appropriate for the situation. Disinhibition has been reported as one of the first symptoms impacting those with prodromal HD (Paulsen et al., 2013).

Environmental factors can also contribute to disinhibited responses. Home environments without routine (e.g. meals served at different times every day, activities not planned but occurring spontaneously) may provoke greater confusion or anxiety which in turn may lead to a greater number of outbursts and responsive behaviours. Noise and distractions can also be triggers. Mild feelings of confusion, annoyance, frustration, irritability or anxiety may be expressed as strong feelings such as anger, rage or fear.

Examples

- A young man with HD and his wife report an increased frequency of arguments at home. She reports that “It’s like he doesn’t even think before he speaks!”
- Over the last few months, a woman with HD noted that she had been drinking more. When questioned further, she noted she will sometimes just think about having a drink and then find she has finished a six pack of beer in an hour.
- A woman with diagnosed HD has had a history of being argumentative with her eldest daughter. While at a stop light on the way home from a shopping trip, they began arguing in the car. “That’s it, I’m out of here!” she shouted and then opened the car door, jumped out and started walking away.
- The police were called about a man with HD who had been throwing rocks at his neighbour’s house and screaming profanities. When they arrived at the scene, the man was back inside his home. He acknowledged what he’d done but stated that he couldn’t stop himself.
- A woman with HD in a long-term care facility had a personal support worker assigned to her at dinner time because she would quickly try to shove all of her food into her mouth, leading to an increased risk of choking.
- A man in the later stages of the disease was no longer able to walk on his own, but that did not stop him from trying to get out from his wheelchair to try to get something he wanted, creating a dangerous risk of falling.

Strategies

There are a number of ways to work with impulsivity. Several classes of medications have been found to be useful in treating this symptom in HD. These include: selective serotonin reuptake inhibitors (SSRIs), particularly those recommended for treating obsessive-compulsive disorder, and antipsychotic or neuroleptic medications. Please refer to HSC’s *A Physician’s Guide to the Management of Huntington Disease* (third edition) for additional information on medications. Non-physicians are encouraged to consult with a medical doctor regarding any pharmacological interventions.

There are a number of behavioural interventions that can be useful for a person who is dealing with impulsivity. It may be that the impulsive behaviour is a response to something real that needs attention. The person might be upset or anxious about a situation or just physically uncomfortable. The following are some strategies to share with family members and caregivers.

- Assess the situation and look for any possible triggers. Environmental factors like noise or temperature or personal factors (e.g. other health issues, hunger or thirst, need to use the bathroom) can lead to a sudden change in behaviour. If there is an identifiable trigger, see if it can be modified.
- Often impulsivity manifests as temper outbursts (yelling, screaming, hitting). Once the person with HD has calmed down, discuss appropriate methods of communicating his or her needs.
- Learn about active listening, which involves letting the person with HD express his or her feelings rather than rapidly reacting to a behaviour. As the person with HD might not be able to put his or her feelings into words, gently attempting to identify what he or she is feeling, and asking for confirmation, might help to clarify the cause of the outburst.
- Remaining in control may help calm the person with HD. Regulating one's responses can help avoid escalation of the situation. It will also reduce the chances of reinforcing maladaptive attention-seeking behaviour. Practice staying calm in order to avoid reacting emotionally.
- A set routine and a predictable daily schedule can reduce feelings of confusion or fear and may decrease the frequency of outbursts.
- Remember that even though the things being said may be hurtful or embarrassing, generally the person with HD is not doing this intentionally. In many cases, the person with HD may be remorseful afterward.
- Be sensitive to the efforts of the person with HD to apologize and avoid reminding the person of the behaviour after the fact. This lack of control is part of the disease and is not by choice.

Frustration, Irritability, and Anger

Frustration, irritability and anger are behaviours that most people experience regardless of whether or not they have HD. However, these behaviours can become more pronounced in a person affected with HD because of the deterioration of the caudate.

Frustration is an emotion that arises when a person's thoughts or actions are prevented (internally or externally) from reaching some desired outcome. In HD, this is compounded by the disease altering the way a person thinks and moves.

Irritability occurs when a person is easily annoyed, excited, or angered. With that broad definition, irritability can take different forms. Responses can become exaggerated in intensity and duration or be punctuated by episodes of explosiveness, compounded by impulsivity or disinhibition.

Anger is a strong, negative feeling of displeasure or hostility. Anger can lead to outbursts, including aggression. Aggressive behaviours can be particularly disturbing to family members or caregivers of the person with HD because they can be a source of fear and tension in the household or in the care facility.

Possible Causes

Frustration, irritability, anger and temper outbursts are all affected by the changes in the brain due to HD. Feelings of frustration and anger, and their subsequent reactions, are usually triggered by real and legitimate events in life, but the brain cannot control the intensity of the response. Often, anger is an emotion that covers

for another emotion. Typically when a person experiences anger, the underlying feeling is disappointment, grief, hurt, frustration, fear or anxiety. There are other possible triggers for this kind of behaviour, including hunger, pain, boredom, difficult interpersonal relationships, inability to communicate, frustration with failing capabilities, and in particular, minor changes in routine.

Recognize that frustration, irritability and anger are usually expressed in relationship to a true feeling or in response to a triggering event, but with HD they become exaggerated due to a loss of control from the caudate.

Examples

- A woman who is working as a housekeeper and does not have any motor symptoms of HD reports that she is starting to feel like she is losing her patience with her clients. “It’s not that I am yelling at them or anything, but I am getting so fed up with all of the comments and suggestions they make.”
- A father of three recently went on disability and planned to spend his time helping at home with the kids. As time went on, he noted that they were always running around, chores weren’t getting done, and at times he felt like he couldn’t handle them. He was aware that he was getting frustrated, but he was afraid he might lose it one day and snap, just like he remembered his mother doing when he was younger.
- A young accountant reported increased frustration and irritability at work as she became aware that she was not able to do her job as she used to.
- The wife of a person with HD says that he tends to get extremely upset about the smallest things.
- Although she denies any problems with driving, a woman with HD recently admitted that she gets really angry at other drivers on the highway, yelling in her car and giving people the finger. Sometimes, she will follow someone who has cut her off, crossing lanes and driving aggressively, just to let them know how she feels.
- A man said that his wife had started getting angry at meal time, pushing her plate off the table and refusing to eat. Upon investigation, it was found that she had been having incidents of choking and that her reaction came whenever hard to eat food was served.
- A man living in a care facility slammed his door and turned up his radio as loud as he could whenever the nurse that he preferred went on vacation.
- Without warning, a person in a nursing home took a swing at the personal support worker who was helping him get out of bed. Even though she had worked with him for several months, his behaviour towards her became more and more aggressive. When a new staff member was assigned, he stopped acting out.

Strategies

There are both pharmacological and behavioural recommendations for dealing with frustration, irritability and anger. As there is often an underlying depression or other mood symptom present in irritability and anger, the first line of medication suggested are antidepressants, such as the selective serotonin reuptake inhibitors (SSRIs) and selective serotonin and norepinephrine reuptake inhibitors (SNRIs). In some cases, mood stabilizers or long acting benzodiazepines can be beneficial. Neuroleptics or antipsychotic medications are helpful in cases where the person with HD did not respond to the antidepressant medications. Specific recommendations for medications are unique to each case.

Please refer to HSC's *A Physician's Guide to the Management of Huntington Disease* (third edition) for additional information on medications, and non-physicians are encouraged to consult with a medical doctor regarding any pharmacological interventions.

Behavioural interventions can be useful for dealing with frustration, irritability and anger, with variations depending on the individual circumstances and the stage of disease. The following are some strategies to share with family members and caregivers.

- Close attention should be paid to signals, verbal or nonverbal, that the person with HD is upset or wants something, so that behaviours do not escalate before they receive attention. Verbal cues include stated feelings of frustration and what is being said. Nonverbal cues include volume of speech, speed or intensity of speech, and sometimes when a person stops talking.
- Frustration arises when a person is not able to do something they want to do (either for internal or external reasons). If the person with HD is capable, encourage him or her to be as independent as possible, allowing for appropriate risks without the expectation of failure.
- Be aware of your own level of frustration and that of other caregivers. Assess your expectations regarding the person with HD. Are you unwilling or unable to accept the new limitations of the person with HD?
- Restructuring the person with HD's interactions, expectations and responsibilities can lessen the likelihood of irritability. Changes made gradually or in stages are often easier to accept than one big change. Restructuring may need to take place frequently as more activities become difficult.
- A non-institutionalized adult should be responsible for some things around the house. Tasks should be appropriate to the level of function (e.g. watering the plants in the front yard, sweeping a specific room) but not overwhelming. Retaining some responsibility can help the person with HD avoid frustration.
- A person with HD, who lives in a care facility, may not be as active or communicative as they once were, but they can still be a part of making decisions. Note that it can be difficult for the person with HD when roles change and responsibilities get reassigned.

Anger and aggressive outbursts can be frightening and in some cases can turn dangerous. The coping strategies for dealing with this type of behaviour include an emphasis on safety. The following are some strategies to share with family members and caregivers.

- Try to identify triggers that lead to temper outbursts. Strategize how, whenever possible, to avoid or modify them. Some triggers are easier to change than others (e.g. a particular show on television or the kids running around vs. a specific family member or world events). If the trigger is one that is hard or impossible to change, consult with a mental health provider.

- Try to redirect the person with HD away from the source of anger. Distraction can be a useful tool. Remember, persons with HD tend to become mentally inflexible and are typically comforted by stability. Confrontations and ultimatums should be avoided. Do not point or shake a finger, yell, or show other obvious signs of anger, as this could escalate the situation.
- HD in itself does not cause a person to become dangerous, but a loss of normal regulation of impulses (disinhibition) can contribute to unsafe situations. Take steps to provide a safe environment so that no person is ever in danger. Potential weapons should be removed from the house and emergency numbers should be posted near the telephone.
- **If a situation seems to be getting worse, and the person with HD is unable to stop the escalation, leave the situation, call for assistance (friend, family, neighbour), and call the police. You may be reluctant to involve the police, but if the situation is getting to a point where there is a threat of danger to anyone involved, contacting the authorities is the appropriate action for both you and the person with HD. Safety first.**

Denial and Unawareness

Denial is commonly seen in persons with HD and arises from several different sources. Frequently, denial is presented as a psychological inability to cope with distressing circumstances, such as loss of a loved one (denial that they are gone) or receiving news of a terminal disease (e.g. getting an HD diagnosis). This behaviour often decreases over time as the person begins to face reality.

In addition to this common form of denial, persons with HD often suffer from a lack of insight or self-awareness that causes them to be unable to recognize their own disabilities or evaluate their own behaviour. This type of denial is called organic denial, or anosognosia, and is a condition that may last a lifetime. **Given that denial is often presumed to be under the control of the individual, the term may not be useful for persons with HD. Therefore, the term unawareness is better suited to describe this behaviour.**

Unawareness can initially be beneficial because it keeps the person motivated to try things and to avoid labelling him or herself as affected or impaired. On the other hand, anger may develop from unawareness because the person with HD cannot understand why he or she cannot go back to work or live independently. The person with HD may believe that people are unjustifiably keeping him or her away from activities such as driving, working or caring for children. This type of unawareness can become dangerous if the person with HD attempts to do things independently that are not safe. Often, a person with HD is thought to have poor judgment. In this case, judgment is impaired because of unawareness due to the disease.

Unawareness is not only a problem for the person with HD but also for health professionals, friends, family members and caregivers. Some persons with HD will not acknowledge symptoms and therefore don't report them to their doctor. The doctor might also think that the person is actively denying symptoms, when in fact they are unaware of chorea or cognitive changes.

Lastly, denial can occur when family members and/or health professionals delay making a diagnosis of HD, or keep the diagnosis from the affected individual, because they are concerned that the person cannot handle it. Some people interpret the unawareness shown by a person with HD as a sign that the person does not want to know, will get depressed, or become suicidal if he or she has HD. **There is no evidence to suggest that talking about HD to a person with unawareness will cause depression or suicide.**

Possible Causes

Denial is a normal psychological response to an overwhelming situation. This can be a conscious process where a person refuses to admit that something has occurred, whether it is an HD diagnosis or to say that a person is having trouble at work. Denial is seen as a way to protect the person's self-perception or identity.

Unawareness is due to damage that occurs in the circuits connecting the caudate nucleus and the frontal and parietal lobes. The circuit that relays information from other centres in the brain to the frontal lobes (where appropriate actions are considered and initiated) is interrupted by caudate degeneration secondary to HD. Unawareness of symptoms is a common problem experienced by people with various types of brain disease, including head trauma, schizophrenia, dementia and other neurodegenerative disorders. Literature in HD is robust with numerous publications describing unawareness of motor, cognitive, psychiatric, and functional dysfunction secondary to HD in both prodromal and diagnosed patients (Deckel & Morrison, 1996; Craufurd, Griffiths, & Neary, 1998; Hoth et al., 2007).

Examples

- A woman who exhibits some early evidence of symptoms reports increasing difficulty at work, but states it is because of the changes made around the office including her new computer.
- A man with obvious chorea is getting the results from a confirmatory gene test. When asked if he has any thoughts about what the outcome might be, he states that he is sure it will be negative.
- Even though his doctor told him over two years ago that he had some motor symptoms, a man will not say that he has HD, but that he is at-risk.
- A person with HD is reluctant to schedule another appointment at the clinic. "I don't know why I even need to come in anymore; I do not have HD."
- A woman reports that she is able to still manage things at home, but her arms and legs are bruised and have some burn marks on them.
- When asked if she wanted to try increasing her medication to control her involuntary movements, a woman with HD stated she never even notices moving.
- A man in an assisted living facility only bathes when forced. He doesn't think that he is unclean, despite other residents complaining about his hygiene.
- Whenever there is a new healthcare aide assigned to a man with HD living in a facility, he insists he can feed himself. When he attempts to do so, he eats too quickly and chokes on his meal.

Strategies

Coping with denial and unawareness can be difficult, and there is no specific medical treatment for these symptoms of HD. Treating any underlying mood conditions, like depression or anxiety, may reduce the denial. Behavioural interventions may help with unawareness, whether implemented with the person with HD directly or with family members or caregivers. The following are some strategies to share with family members and caregivers.

- There is no single best way to cope with denial and unawareness. At times it may require creative thinking to get the person with HD to cooperate with a request.

- Avoid interpreting non-compliance with therapy, nursing care or even household arrangements as intentional, as unawareness is often the reason for these behaviours.
- Try to accept that unawareness is a component of HD that is not treatable. Often the person with HD does not seem to change or accept his or her HD.
- When needing compliance from a person with HD who has unawareness, it may be helpful to develop a contract that includes incentives or rewards for compliance that are not tied to the acknowledgment of the HD diagnosis.

Perseveration

A person with HD may perseverate, or become stuck, on one idea or activity. The person may become rigid in thinking and have a difficult time switching from one thought or activity to another or altering routines. Psychiatric rating scales assessing levels of Obsessive Compulsive Disorder (OCD) suggest that OCD-like behaviours, including perseveration, are among the most common early symptoms of HD (Beglinger, et al., 2010; Duff et al., 2010). Some common examples of perseveration in persons with HD include obtaining cigarettes, getting coffee refills, meal times, or using the washroom. A person with HD may become irritable when these requests are ignored or denied.

Perseveration can also be caused by communication difficulties. For example, if legitimate needs are not being met, the person with HD will continue to repeat him or herself in the hope of being understood and assisted.

An obsessive thought or the need to complete a compulsive action has been described by some persons with HD as an itch or a habit that must occur. Some describe it as similar to restless leg syndrome where actual discomfort occurs if the compulsion is not conducted. It can be helpful to allow the behaviours to occur when they are not harmful.

Possible Causes

Perseveration occurs when there is damage to the frontal lobes or the neuronal circuitry connecting the lobes to the basal ganglia.

Examples

- A young man with HD tries to exercise every day. Not only does he go to the gym and run when he can, but he talks about it with his friends and family. Not a day goes by where he doesn't explain his entire routine and share his achievements at the gym.
- After getting a negative review at work, a woman with HD started to think about one particular mistake she had made. As time went on, she continued to think about this mistake, noting she felt upset whenever she did and that sometimes she would have a hard time falling asleep because it was on her mind. At her next review cycle, she brought it up to her supervisor, who said she could barely remember that happening a year ago.
- A woman reports that her husband with HD will not let go of the idea that he will one day go back to work. At times, he will follow her around the house and insist that she take him out so that he can look for work.
- During the last few weeks that she lived on her own, a woman with HD began to obsess about a form she had mailed pertaining to her divorce. After sending it, she began to worry that it contained errors that would impact her children's well being. No one could reassure her and she became so agitated that she would call her son every few minutes to talk about the matter.

- The assisted living facility a man with HD lived in decided it was time for him to stop smoking, as his chorea was worsening and he was becoming a burn risk. Even after he stopped, he would go to the desk multiple times per day and ask for a cigarette.
- A woman in a nursing home would only drink her nutritional supplement if it was in a paper cup from a specific coffee shop. If she was given any other cup, she would get upset and act out.

Strategies

Managing perseveration is a particularly difficult challenge. Giving the person with HD a selective serotonin reuptake inhibitor (SSRI) may help with some of the obsessive focus. In cases where the person does not respond to an SSRI, a neuroleptic or antipsychotic can be tried.

While rational arguments do not have much effect, providing the person with a sense of security, and an alternative way to express his or her feelings, may be an effective way to reduce frustration for both the person and caregiver. The following are some strategies to share with family members and caregivers.

- Empathizing with the feelings behind the words or actions can help make the person with HD feel more secure. For example, “It sounds like you are upset about not driving,” or “it seems to me that you are angry that we didn’t go the mall.” Acknowledging the feelings of the person with HD may reduce the tendency for perseveration.
- Consider employing distraction as a method for dealing with perseveration. Keep a list of favoured activities in mind, or have various foods that are of interest to the person with HD on hand. These activities or items can be used to gently shift the person off a perseverative topic. For example, if the person with HD begins to perseverate while doing routine chores, the caregiver should be prepared to switch to another task. Be prepared with several options.
- Humour may also effectively break a cognitive set (a topic that the person is stuck on).
- Remind the person with HD that the topic has already been discussed. If no acceptable conclusion was agreed upon, gently remind the person where you are in the process of problem-solving.
- Accommodation can be a strategy. In the case of the woman who would only take her nutritional supplement in a cup from a specific coffee shop, the staff obtained a package of cups so that there was always one available for her.
- Unfortunately, some perseverations are unsolvable. Try setting limits on the behaviour, or even on how long the person with HD will be allowed to talk about the issue.

Note: It can be effective to have the physician act as the “bad person” and limit activities that the person with HD refuses to limit at the family or caregiver’s request. This allows caregivers to maintain a positive relationship with the person. For example, if a person with HD wants to drive but is clearly unable to do so, recommend that he or she go through the tests at the motor vehicle licensing centre and let that specialist make the decision. In these types of scenarios, the team approach to care enables all team members to fulfill their roles. Discussions within the care team (including the caregivers) ensure that everyone on the team is clear and comfortable with the respective roles.

Depression

Depression is a mood disorder that is common among persons with HD. It can develop during the prodromal stage or when the disease is manifest. In the disease prodrome, symptoms of depression can develop many years before motor symptoms and the highest prevalence of depression during this period may be within one year of clinical diagnosis (Epping and Paulsen, 2011). In the manifest disease, earlier stages have the highest prevalence of depression.

Symptoms of Major Depressive Disorder (MDD)³ include:

- Depressed mood most of the day, nearly every day
- Decreased ability to find pleasure or interest in usual activities
- Significant decrease or increase in appetite or weight
- Fatigue or loss of energy nearly every day
- Changes in sleep – insomnia or hypersomnia
- Restlessness or feeling physically slower
- Decreased ability to think, concentrate, and make decisions
- Feelings of guilt or worthlessness
- Recurrent thoughts of death or committing suicide

Many of the symptoms of HD resemble and may potentially disguise the symptoms of depression. It can be difficult to tell whether a person's symptoms are depression, HD or a combination of both. A person who is at-risk, or in the prodrome stages of the disease, may self-report that they feel depressed. However, as the disease progresses into the diagnosed stages, it is often the observation of a family member or caregiver that alerts the doctor of the person with HD to the presence of depression. Being alert to symptoms of depression is necessary because the incidence of suicide is elevated among people with HD. This will be discussed in greater detail at the end of this section.

Possible Causes

There are two reasons that depression is so common in HD. First, a depressed mood is a natural and understandable emotional response to the diagnosis and symptoms of HD. HD progressively alters a person's role in the family, work and social realms. For example, the person's role in their family may change from breadwinner and household manager to a dependent person requiring supervision. Symptoms of depressed mood as a result of an identifiable cause or reason can be referred to as a reactive depression, similar to the diagnosis of an adjustment disorder. However, this is more complicated in HD because of the progressive nature of the disease. As a person with HD develops more symptoms, or experiences more changes in life, he can continue to experience reactive depression, whether a new onset or the perpetuation of an existing state.

The second reason for depression in HD is that the disease directly alters neurotransmitters in the brain that regulate mood. Consequently, even persons who are naturally optimistic and happy can experience severe depression secondary to the brain changes in HD. To complicate matters even further, the individual who is experiencing depression, due to changes in the brain, can also be reacting to changes in his life.

Examples

- A young man is seen in clinic six months after having genetic testing and discovering that he is gene positive. He reports that his mood has been “up and down,” but that in the last month or so he has been feeling more hopeless about the future.
- A young woman reports that she is not depressed, but feels “kind of angry, like all of the time.” She hasn’t been sleeping as well as she used to and doesn’t have much of an appetite. She reports getting upset with her boyfriend and cries frequently. She goes to her college classes, but has stopped going to the gym and doesn’t see her friends as much as before.
- A woman with HD, who had early evidence of the movement disorder, reports that she just doesn’t have the energy she used to for working around the house. Laundry is left unwashed, the dishes pile up in the sink, and taking the dog out for a walk only lasts about five minutes. Most days, she sits on the couch with the TV on. Sometimes she says an hour or two might pass, and she cannot recall what she has been watching.
- A man with HD, who has been experiencing more motor symptoms, talked about his father’s suicide attempt quite frequently. At his most recent clinic visit, he stated that he wanted to plan “one last trip back east” to where his father had lived. When asked what he meant by that, he switched the topic. When asked directly about suicide, he denied having any thoughts about killing himself. Two weeks after his visit, his wife called and said he had been acting weird and had been saying goodbye to the neighbours in such a way that she was worried.
- A man with HD becomes tearful and agitated every time his family visits him at the nursing home. After the visit is over, he resists getting out of bed and stays in his room, indicating to the staff that he wants the lights off and the curtains drawn.

Strategies

There are a number of ways to work with the symptoms of depression. While each case is unique, several classes of medications have been found useful for treating depression. These include selective serotonin reuptake inhibitors (SSRIs), selective serotonin and norepinephrine reuptake inhibitors (SNRIs), atypical antidepressants, and antipsychotic or neuroleptic medications. Please refer to HSC’s *A Physician’s Guide to the Management of Huntington Disease* (third edition) for additional information on medications. Non-physicians should consult with a medical doctor regarding any pharmacological interventions.

Behavioural interventions are a non-pharmacological approach that can also be successful in helping an individual manage depression.

- Encourage the person with HD to talk about depression with someone he or she knows and trusts - whether a loved one, friend or doctor. Sometimes just talking to someone about thoughts and feelings can help alleviate a depressed mood. Taking this first step is often the most difficult.
- In addition to informal counselling, recommend that the depressed person meet with a mental health professional who can provide insight into depression and develop ongoing interventions.
- Have a conversation with the person with HD about medications. As there are many different medications available that can help depression and no single best recommendation, the person with HD will have to work with his or her doctor to find an effective medication and dose.

- Since the ability of a person with HD to communicate can change over the course of the disease, counselling may be more effective in the at-risk, prodrome, and early diagnosed HD stages. In mid-stage, a mental health professional can help to develop and implement behavioural interventions. In the late stages of HD, regular visits from a mental health professional, social worker, or chaplain can be reassuring and beneficial to a person with HD even if they are unable to talk.
- Suggest attending an HD support group as this may be a useful tool in addressing symptoms of depression. Often times, people living with HD feel isolated and alone which only worsens depression. Meeting other people who have a shared experience and know the disease can provide a social connection and outlet for the person. That said, support groups are not for everyone, and it is up to each person to determine their own level of comfort with this resource.
- Encourage the person with HD, who is feeling depressed, to engage in exercise and other physical activities. There is abundant research on the benefits of exercise and strong evidence that it can help a person's mood.
- Hobbies and other pleasurable activities can also relieve symptoms of depression. Encourage family and friends to include the person with HD in activities. Having a companion in the activity may also increase participation, as difficulty initiating activities is a symptom of HD.
- Reinforce maintenance of routines and schedules. Consistent and regular activities can help provide structure for the person with HD, which in turn can help address depressed mood. Gentle encouragement may increase the likelihood of the person remaining engaged in an activity.
- Monitor and report conversations about suicide and death (see the following sections).

Suicide and HD

While studies have suggested that there is a greater risk of suicide in persons with HD, there has been little research on this topic – especially in Canada. One study sought to determine risk factors for suicidal behaviour (defined as suicide or attempted suicide) in prodromal HD (Fiedorowicz, Mills, Ruggle, Langbehn, & Paulsen 2012). This research concluded that “a history of suicide attempts and the presence of depression are strongly predictive of suicidal behaviour in prodromal HD. As these risk factors are among the most robust risk factors for suicide, established suicide risk factors appear applicable to those with prodromal HD.”

Given the variance in statistics related to suicide rates, it is difficult to determine the rate of suicide in persons with HD or those who are at-risk for HD. As mentioned, findings typically suggest that risk factors are similar to those of the general population (previous attempts, depressed mood). Monitor depression in the person with HD and ask about suicide regularly, as thoughts about death and suicide can be a part of depression. Suicide is of great concern in HD due to cognitive changes in the brain, including disinhibition and impulsivity.

There are a number of risk factors for suicide - some that are obvious and some that are more subtle. Things to look for include the following:

- Depressed mood (see the section on depression for signs and symptoms)
- Expressed feelings of hopelessness
- Expressed desire to be alone
- Withdrawal from activities, friends, family, or society
- Talk about “ending it all,” “not dealing with it anymore,” or generally about death

- Active planning, for example specifically stating what he or she would do
- Stated desire to put one's life in order by saying goodbye to people or giving away possessions
- Changes in behaviour such as increased agitation or sleeping much more (or much less)
- A sudden change in mood from depressed to happy

Suicide Prevention

Based on the above criteria and your clinical experience, if you consider the person with HD to be at-risk for suicide:

- Provide a referral to a mental health specialist. Suicide is preventable and counselling can help a person deal with thoughts about suicide.
- Encourage the person with HD to talk about their feelings with you, family members, caregivers or other trusted individuals.
- Ask the person with HD if they have a specific suicide plan. If they do, do not leave them alone. Call a suicide prevention line or crisis centre, call 911 or take the person to the closest emergency room.
- Coach family and caregivers on how to react to discussions of suicide. It is common for people to get upset if they hear that someone is thinking about suicide. They may try to negate or dismiss the talk. Counsel them to listen supportively and to encourage the person with HD to share what they are feeling, but also to be prepared to call a suicide prevention line or 911 if they feel the person with HD is in imminent danger of hurting themselves.
- If the talk of suicide is not specific, suggest that the person with HD contact a medical professional or mental health provider. For example, "I feel sad and scared when I hear you say that. Let's get in touch with Dr. Johnson, okay?" If the person with HD does not want to talk to a doctor or mental health professional, recommend that the individual hearing about the suicidal thoughts make the call and alert the person's doctor.
- Have the family or caregiver of the person with HD remove all potential weapons from the home, including guns, bullets, rope and medications. Counsel family members that many chronically ill people may hide medication "in case I need them." It is not only suicide which is problematic, but a decision such as, "I'm not right today – I need to take a few of those pills."
- Instruct the family, caregiver or person with HD to post emergency phone numbers by the phone.
- Request regular contact to periodically evaluate mood changes. Monitoring symptoms of depression helps to identify any suicidal thoughts or feelings as they arise.
- Make a contract with the person to let you know if he or she begins to feel badly.
- Arrange for supervision.

Anxiety

It is common for a person with any chronic progressive illness to experience some excess anxiety, or worries, about the future. However, symptoms of anxiety can become so severe that they interfere with the activities of daily living.

Anxiety can present in a variety of ways including:

- General nervousness
- Excessive worrying
- Repetitive thoughts about troublesome topics
- Fidgeting hands
- Shallow breathing
- Rapid heart rate
- Sweating
- Restlessness
- Fear
- Panic

Anxiety can serve as a common link among several distinct psychiatric disorders. These include social anxiety, panic disorder, and obsessive compulsive disorder. A person with HD may not meet the specific criteria for any one of these disorders, but they may show features of one or more of them.

- **Social anxiety** is worry or fear about how one will be perceived in a social setting. It is not uncommon for persons with HD to worry that other people are looking at their involuntary movements. Additionally, people in the early stages of the disease may be concerned about how co-workers and friends will react to their HD.
- **Panic disorder** is characterized by an acute onset of overwhelming anxiety and feelings of dread, often accompanied by physical symptoms including rapid heart rate, sweating, hyperventilation, light-headedness, or numbness and tingling of fingers and toes. The symptoms typically last about 15 minutes, but residual anxiety often remains.
- **Obsessive-compulsive disorder** is characterized by recurrent intrusive thoughts or impulses (obsessions) that are anxiety provoking, but experienced as senseless. Compulsions are repetitive behaviours that are performed over and over, sometimes in response to an obsession or as part of a stereotyped routine that must be followed. The most common obsessions tend to focus on cleanliness (such as washing hands) or safety (such as checking to make sure the stove is turned off).

Although true panic and obsessive-compulsive disorders are rare in HD, they can occur. Again, it is more common to see components of these anxiety disorders, such as obsessive preoccupation with particular ideas (as discussed in the section on Perseveration).

Typically, anxiety symptoms become worse in new situations or when the person perceives him or herself as having insufficient skills to handle the situation. Many persons with HD find that they worry more than they used to, even about seemingly trivial matters.

Possible Causes

Much like depression, there are two main reasons that anxiety can be seen in HD. First, there is reactive anxiety to the challenging and changing situations in life. Whether someone is waiting for results from genetic testing, experiencing increasing difficulty at work, observing changing roles within the family or experiencing symptom progression, there are many sources for understandable worry. This can become problematic however, as many times changes occur concurrently, leading to increased worry and anxiety.

The second reason for anxiety in HD is due to changes in the brain. Again, as the disease progresses, physical changes in the brain can both lead to a person developing anxiety, as well as experiencing increased difficulty in dealing with anxiety. These two factors are often connected, and this can lead to increased presentation of symptoms of anxiety.

Examples

- A woman in her thirties, who is in the prodrome stage, worries that she is not able to keep up with her workload. She has discussed her status with her supervisor and has explored options for modified duties. However, even with changes made to her responsibilities, she spends most of her days thinking about her performance. She also reports increased difficulties falling asleep as she finds herself thinking about her job before bed.
- Despite having received positive reviews the last three years, a man with HD thinks his co-workers are looking at him differently now. Even though he can still keep up with his work, he fears people are judging his performance. He has also noticed some movements in his fingers and worries that other people can see it too.
- Still living on her own, a woman diagnosed with HD was seen in clinic along with one of her children. Recently, she had become focused on the idea of traveling across the country to visit her father. The more she thought about traveling, the more worried she became that she would not be all right to fly. She would become agitated, experience physical symptoms including shortness of breath and racing heart, and would get easily confused.
- A person in a care facility had experienced several incidents of choking on food during meals. The last time he choked, someone had to give him abdominal thrusts to dislodge the food. Although an assessment showed that, with attention, he could still manage solid foods, he became worried that any meal would lead to choking. He began to refuse foods and became increasingly uncooperative at mealtime.

Treatment

In dealing with symptoms of anxiety, there are both preventative measures that can help avoid the emergence of anxiety and ways to address symptoms that are already present. A number of medications have been found useful in treating symptoms of anxiety in HD. Specific recommendations for medications are unique to each case, and the clinician should first assess whether the anxiety is a symptom of some other psychiatric condition, such as a major depression. Potential treatments include selective serotonin reuptake inhibitors (SSRIs) and benzodiazepines. Please refer to HSC's *A Physician's Guide to the Management of Huntington Disease* (third edition) for additional information on medications for treating anxiety.

Cognitive-behavioural interventions can help avoid or minimize the likelihood of a person with HD becoming anxious.

The cognitive component looks at a person's anxious thoughts and teaches him or her how to challenge these beliefs. Having the person with HD identify the anxiety-provoking thoughts is the first step (e.g. "I won't be able to finish this project at work" or "everyone thinks I can't be helpful because I have HD"). Having the person develop a challenging statement to the original thought can then lessen the anxious response (e.g. "I don't know what everyone else is thinking, but my family still thinks I am helpful around the house"). Besides challenging

anxious thoughts, using positive self-talk, such as “I’m going to be okay,” can be effective in reducing anxiety.

The behavioural component provides interventions that minimize the physiological aspects of anxiety, including relaxation training. The simplest form of relaxation training focuses on breathing. Then, depending on the abilities of the person with HD, there are more complicated exercises, including biofeedback and hypnosis.

Strategies

Below are some strategies to share with family members and caregivers.

- Establish regular routines and stick to them. As mentioned in previous sections, routines and schedules can help minimize stress and provide a structure in which the person with HD feels secure.
- Keep the home environment simple. An overabundance of stimuli can lead to the person with HD becoming overwhelmed. Turning the television off when leaving the room, setting the phone ringer on low volume and minimizing clutter can help reduce potential triggers for stress.
- Simplify requests and demands on the person with HD. Due to changes in the brain, it can become more difficult for the person with HD to organize thoughts and carry out complex actions.
- In some cases, it can be helpful to refrain from discussing future events until the day before the event is to occur. This is not a blanket recommendation, but for many persons with HD who tend to worry and perseverate, waiting to alert the person until a time close to the event can minimize anxiety.
- In other cases, again depending on the person, gently introducing an event in advance and adding details over time can minimize anxiety. The approach will depend on the individual.
- Plan ahead for changes in routine and the alterations in routine that will be needed. Encourage everyone involved, including the person with HD, to think about upcoming modifications to a routine in order to minimize anxiety or worry at the time of the change.
- Look for any identifiable trigger(s) and recommend that the person with HD discontinue the activity that is contributing to the feelings of anxiety.
- Behavioural interventions will change as the disease progresses. Persons who are in the prodrome and earlier stages of diagnosed HD can benefit more from certain interventions than those in later stages.
- Cognitive-behavioural therapy techniques are well known for treating anxiety, although there has been no research in this area specific to HD. Anecdotally, these interventions can be very helpful, and a referral to a mental health care provider trained in this technique is recommended. There are two components to the cognitive-behavioural approach: learning to recognize and manage unhelpful thoughts or emotions and changing dysfunctional behaviour.

Psychosis: Hallucinations and Delusions

Psychosis is a psychiatric term for a mental state in which a person experiences a loss of contact with reality, usually including the presence of hallucinations or delusions. A hallucination is when a person sees, hears or experiences something that is not real. Delusions are thoughts about unreal situations and relationships. Fortunately, psychosis is rare in HD.

There are many kinds of delusions including: paranoid or persecutory delusions, involving the belief of being followed, watched, or in danger; erotomania, involving the belief that another person is in love or in a relationship with the individual; and grandiose delusions, which are the belief that the person has special talents, powers or abilities. Delusions may be bizarre (“the next door neighbours are really aliens from another planet”) or not bizarre (“I think the police are watching me”).

Possible Causes

Changes in the brain due to the progression of the disease may create psychotic features. Psychotic symptoms may also emerge from an underlying condition unrelated to HD, such as depression or mania, substance use and abuse or other health concerns. It is imperative to rule out any possible causes before treating the psychotic symptoms.

Examples

- A man in his forties believes that his wife is trying to poison him, so he refuses to take his medications. This increases his delusional and paranoid symptoms to the point where he is threatening his wife and frightening his family.
- A woman with HD who has been married to her husband for nearly twenty years confesses to a series of extramarital affairs, including a former college friend, a local news personality, her medical doctor and a famous celebrity. While there is no evidence for her claims, she feels guilty for cheating, and tells everyone she knows about what she has done and how bad she feels.
- A person with HD reports that his neighbours are devil worshippers. “I have been watching them for the last couple of weeks and I know there is something going on in their garage. In fact, I can hear them chanting when I am in bed at night.”
- A man with HD suffered from hallucinations in which he reported seeing “people from the past” in his room. He was placed on an antipsychotic medication and he reported seeing the people less often. As his symptoms worsened, he was no longer able to communicate and became bed bound, where he would become agitated, thrash in his bed and groan. While he couldn’t confirm that he was hallucinating, increased antipsychotic medication was calming.

Strategies

Because psychotic symptoms do not occur that frequently in HD, the first line of treatment would be to consult with a psychiatrist. Once any underlying medical causes, untreated mood symptoms or substance use is ruled out, psychosis is typically managed well with medications. Neuroleptics or antipsychotic medications are used to treat hallucinations or delusions. They are also useful in treating anger, irritability, perseveration or obsessiveness, difficult depressions and even the movement symptoms of HD. Because of the various side effects of these medications, it is strongly recommended to consult a psychiatrist or medical doctor (i.e. neurologist/HD specialist) familiar with the use of this type of medication in the HD population.

In addition to medication, treatment with a mental health care provider can be beneficial. Behavioural interventions can help provide structure, decrease the distress caused by experiencing hallucinations or delusions, and/or help family members or caregivers deal better with the person with HD.

When working with a person with psychotic symptoms, a primary goal should be safety. Assess if the hallucinations or delusions are making the person with HD a threat to themselves or others and take immediate steps if necessary. If the delusional content becomes problematic or the hallucinations become disturbing, an immediate interventional strategy must be implemented. Under these circumstances, it is more likely the person with HD will act out.

Below are some strategies to share with family members and caregivers.

- If there is a threat of violence, know when to leave or to contact the authorities. Prepare an escape routine. While it is hard to imagine calling law enforcement on a person you care for, making the call is better than harm occurring to either party.
- If there is no threat of violence and the person with HD is comfortable with the delusion or hallucination, try to work around the beliefs, even if it is difficult. For example, a person with HD, who lives in a care facility, may believe that he is one day going to leave the country to marry a foreign diplomat. His wife is frustrated and upset whenever she hears this. Even though he is happy and content with the belief, it is having a negative impact on his spouse. If you are dealing with a similar situation, try to develop a plan for changing the topic or limiting this particular conversation. Going for counselling may be helpful in this challenging situation.

Because psychosis in HD is rare and the symptoms can be so varied, approach each case individually and develop case by case strategies. When possible, it can be quite beneficial to collaborate with a mental health professional in developing interventions.

Changes in Sexuality

Although changes in sexual behaviour are often uncomfortable for people to discuss, they are very common in persons with HD. The types of changes involve sexual interest and functions. Some persons with HD report an increased interest in sex or a more intense sexual drive, whereas others report diminished sexual interest. The most commonly reported changes in sexual behaviour due to HD, in both men and women, are hypoactive sexual desire and inhibited orgasm. Other symptoms include sexual assault, promiscuity and risky or out of character behaviours. Although the reasons for sexual behaviour changes are not fully understood, changes in sexual functioning often need to be addressed.

Possible Causes

One possible cause for changes in sexual behaviour is due to the effect of HD on the brain. As the disease progresses, the brain is no longer able to regulate the level of sexual drive, resulting in too much or too little. In addition, the delicate balance of hormones in the brain is disrupted by the presence and progression of HD, resulting in variations in behaviours typically regulated by hormone levels.

Besides the physical changes in the brain, changes in sexual behaviour can be due to other behavioural symptoms of HD. Increased promiscuity can be seen secondary to disinhibition, poor judgment or impulsivity. Decreased sex drive can be due to depression, apathy or an inability to initiate activity. Many of the medications used to treat the symptoms of HD can have sexual side effects. While treating the mood symptoms of a person with HD, antidepressant medications can lead to decreased libido and anorgasmia (absence of sexual climax).

Changes in sexual behaviour can also be due to the psychosocial factors related to having a progressive, neurodegenerative condition. When a person experiences changes in roles and relationship dynamics, this can affect sexual behaviour. Other factors are the mood and motor symptoms associated with the disease, as well as awareness of symptom progression. This situation becomes increasingly complex, as almost all of these factors are occurring simultaneously and in an ongoing manner.

Examples

- After receiving his genetic test results and finding out that he was gene positive, a young man reported losing interest in sex with his wife. At his last appointment, he stated, “Why bother? It’s not like we can have kids anyway.”
- A woman who is gene positive and is in her early thirties but does not exhibit any motor symptoms, reports that her interest in sex has decreased over the last few years. Reviewing her history indicates some evidence of depression and anxiety, but she has never been prescribed any medications for those symptoms.
- A man in his mid-thirties has been on an SSRI for approximately two years. Despite stating a desire to continue having sex with his girlfriend, he reports being unable to sustain an erection.
- After reporting psychiatric symptoms associated with HD for several years, a woman seen in clinic acknowledges that she is drinking and having one night stands. She doesn’t think it is a problem, but her family is concerned for her safety.
- After getting divorced last year, a woman with diagnosed HD wants to start dating again. She expresses frustration in meeting a new man and worries about what potential partners might think about her chorea.
- A married couple reports that sex is becoming more complicated, not just because the husband’s chorea is worsening. The medications he is taking make him impotent, which in turn makes him depressed. In fact, it has gotten to the point where he has started to completely avoid physical contact with his wife.
- The personal support worker at a care facility reported that a woman with HD has started refusing to wear her bra and underwear and is lifting her shirt up while in the dining room. This is upsetting other residents. On other occasions, she appears to be masturbating when staff is in her room. When asked not to do so, she becomes agitated and gets upset.
- A man with HD, who lived in a nursing home, pulled a healthcare aide to the floor and lay on top of her. As he tried to remove her shirt, other staff members rushed in and were able to separate them. When asked what he was doing, he was able to indicate that he wanted to have sex because it was his birthday.

Strategies

Each person has the right to achieve his or her highest reasonable potential on the continuum of human sexual development. It is a misconception that inheriting a degenerative disease will cause an end to one’s sexuality. There are several ways to better adjust to the changes in sexuality that HD can bring.

- It is important that open communication exists in both the clinical relationship as well as the personal relationships of the person with HD. Many providers might not feel comfortable discussing sexuality with a person with HD. Others may feel there isn’t enough time in a given appointment to address this issue. In many cases, there are more pressing issues that need addressing, so the topic of sexuality is repeatedly not brought up at all. Talking about sex with a person with HD and his or her family may help lessen the taboo nature of this topic and encourage them to have the conversation within their personal relationships.
- It is rarely just one party who is affected by changes in sexuality. Encourage spouses and significant others to engage in active communication with the person with HD. Advise those involved to consider counselling resources in dealing with the changes in the sexual aspects of their relationships.

- For persons with HD who do not have family members or caregivers, assess for sexual practices and safety. The cognitive and psychiatric changes associated with the disease can lead persons to engage in risky and dangerous behaviours. If a person with HD is alone and incapable of making appropriate decisions, consider enlisting the assistance of social services.

Sexuality is a lifelong process of learning about oneself and growing as a social and sexual being. All persons have a right and a need to the unique pleasure, joy and pain this aspect of identity can bring. For persons living with HD, this includes incorporating changes brought on by the disease. The three areas of symptoms cognitive, psychiatric and motor - all impact a person's sexuality and sexual functioning. As there are many ways that persons with HD experience changes in sexuality, each case should be approached individually.

Sleep Disturbances

Many persons with HD complain of disturbed sleep. Disturbances such as insomnia, restlessness and frequent waking are often just a part of the progressing disease. Persons with HD may also show changes in circadian rhythms, or the biological process that governs a person's roughly 24-hour cycle. Besides the sleep disturbances themselves, not getting enough quality sleep impacts the other symptoms of HD. Although the reasons for sleep disturbances in HD are not fully understood, there are some aspects that can respond to medical treatment.

Possible Causes

As mentioned above, the reasons for sleep disturbances in HD are not entirely clear. Sleep disturbances are seen in other neurodegenerative conditions, like Alzheimer's disease and Parkinson's disease, so changes in the brain due to the disease are most likely a contributing factor. The neurophysiological changes in HD affecting sleep are not clear, and additional research is necessary.

Besides changes in the brain, there are many other factors that contribute to sleep disturbances. Normal aging itself brings about changes in sleep patterns. As people get older, they tend to wake up more often, have lighter sleep, and require a longer time in bed to get adequate rest. In addition, a shift in the body's daily rhythms occurs which makes adults prone to waking in the early morning and fatigued in the early evening.

Although chorea tends to lessen when a person with HD sleeps, it can be a factor in sleep disturbances. The movements may make it harder for a person to fall asleep when they go to bed or to return to sleep after waking in the night.

Mood symptoms also impact sleep. Depression can lead an individual to sleep too much or not enough, and anxiety is often associated with decreased sleep and increased difficulty falling back to sleep. When the person is not getting enough sleep, these symptoms also tend to worsen - creating a vicious cycle.

Finally, common behaviours, seen in both the HD and non-HD populations, can impact sleep. Consuming stimulants like caffeine or nicotine, napping during the day, watching television late into the night and going to sleep later and getting up later all contribute to sleep disturbances.

Examples

- A young man in the prodrome stage of HD is worried that he is starting to experience symptoms. This leads him to become more and more anxious, in turn causing him to experience insomnia.

- As her symptoms began to progress, a woman with HD started to feel more and more exhausted at the end of the day. Since she was still working and had chores to do when she got home, she wasn't able to get enough sleep each night to feel rested. As the week went on, she felt like her level of functioning became more impaired, until she finally got to catch up on sleep during the weekend.
- A man reports at his clinic visit that he cannot fall asleep at night because he is experiencing twitches. The twitches are likely early manifestations of chorea that he does not exhibit upon exam.
- After going on disability, a man with HD notes that he stays up later watching television and playing video games. This leads to him sleeping in later and later each day. This is becoming problematic as his wife reports it is impacting her sleep, and she has to go to work each morning.
- Even though she goes to bed at the same time every night, a woman with HD reports that she will lie in bed for hours without falling asleep.
- A young man with Juvenile HD tells the doctor that he doesn't sleep well at all. However, his mother reports that when she checks on him at night, his eyes are shut and he appears to be asleep.
- A man with HD in a nursing home sleeps throughout the day and then is up during the night. He often gets out of his bed and at times even leaves his room, trying to get out of the building.

Strategies

There are a number of ways to approach sleep disturbances in HD. Pharmacologically, hypnotic medications may be useful in earlier stages, but these are problematic because of abuse potential as well as tolerance issues. Antidepressants (that tend to sedate) and neuroleptics can be useful both for addressing sleep and treating mood and motor symptoms of HD. Melatonin, an over the counter product, can be helpful for those who have difficulty falling asleep. Additional information can be found in HSC's *A Physician's Guide to the Management of Huntington Disease* (third edition). Non-physicians are encouraged to consult with a medical doctor regarding any pharmacological interventions.

Besides medication, there are behavioural interventions recommended for addressing the sleep disturbances associated with HD. Many of these interventions focus on improving both the quality and quantity of sleep a person gets each night. These strategies may be shared with the person with HD and/or caregivers.

- Recommend maintaining a regular schedule: getting up and going to bed at the same time every day.
- Encourage establishing a night time routine to help improve sleep. Actions such as taking a warm shower, having a light snack or reading a book are all ways to relax and can help ease the transition to sleep. Engaging in an arousing activity, such as watching an action movie or searching the Internet, can stimulate the brain and make sleep less likely.
- Regular exercise is known to help with sleep, but not within two to three hours of bedtime because it can cause overstimulation. Exercise completed earlier in the day is shown to help with sleep, as well as benefiting mood and balance.
- Reinforce dietary choices that are conducive to good sleep. This includes avoiding heavy, hard-to-digest food, alcohol, salty food and stimulants (e.g. coffee, chocolate) particularly in the evening or before bedtime.
- Encourage the person to refrain from smoking within several hours of going to bed, as nicotine is a stimulant.

- Recommend avoiding excessive napping during the day and especially in the early evening.
- Urge caution to persons with HD using over-the-counter sleep medications. Issues with tolerance and abuse are common with this type of medication. Educate individuals to avoid using over-the-counter options for more than four days unless their use is reviewed by a medical professional.
- Environmental changes can help improve sleep by making the bedroom more conducive to sleep. These include maintaining a cool temperature and keeping the room dark and quiet at night.
- Many people report sleep disruption because they have to urinate. To reduce night waking due to a full bladder, recommend limiting the amount of liquids consumed after dinner and using the toilet before bed.
- Coach the person with HD on how to fall back to sleep by engaging in less active behaviours, such as watching calming television programs at a low volume, listening to relaxing music, or having a small drink of water.
- People with HD who do not sleep through the night may need added safety measures such as a nightlight to minimize fall risk from navigating in the dark. Rearranging bedroom furniture to leave clear pathways for walking is also recommended.
- If a person with HD is prone to night-wandering, caregivers may wish to make it more difficult for the person to get out of the bedroom. Ideas include: hanging bells on doors, installing hard-to-open doorknobs, deadbolt locks or oddly placed latches. Bed restraints are not recommended.

Chapter 4

Other Factors That May Affect Behaviour



IV. Other Factors That May Affect Behaviour

All persons have a history and a life course. They have personal experiences and objectives, and it is important to try to understand how the disease has interfered with the person's life. Whatever the stage of the disease, it is important to remember that there is still a person, an individual, behind the HD. Often the interpreter pays sole attention to the diagnosis and the personality of the individual is neglected. There are many other factors to be considered when looking for the cause or trigger of troubling behaviour.

Underlying Medical Conditions

HD may not be the only health problem of the person. Dental problems, pneumonia, fever, urinary infection, hemorrhoids, a cold or flu, gynecological problems, chronic illness such as arthritis and diabetes, and the natural aging process, including menopause, can lead to changes in behaviour. It may not be easy to identify health problems in a person with HD if there are speech problems, word-finding problems or lack of self-awareness. The observations of a family member or caregiver may be helpful.

The Effects of Medications

Medications prescribed for certain symptoms of HD, such as chorea, depression, aggression and temper outbursts, may have side effects that alter behaviour. Drowsiness, decreased motivation, nausea, dizziness and depression should be reported to the doctor.

Hunger

Hunger can be a significant cause of behaviour problems in HD. Persons with HD need additional calories to perform the ordinary activities of life. They are often less able to identify hunger, need more time to eat, may be easily distracted while eating and may have swallowing problems that interfere with getting enough nutrition. If possible, determine how much nutrition the person with HD is actually getting. The help of a speech-language pathologist and dietitian may be necessary to ensure that the person with HD is getting enough calories.

Fatigue

Fatigue is another significant cause of behaviour problems. Persons with HD use greater amounts of energy to perform ordinary activities of daily life. Fatigue and the inability to recognize tiredness can increase irritability. It may be helpful to suggest additional sleep or "down time" between activities to avoid exhaustion.

Dehydration

Persons with HD often cannot recognize the sensation of thirst or are unable to initiate getting a drink. As a result, they may become dehydrated. The symptoms of dehydration may include dizziness, confusion, refusal to drink, dry skin, fever, flushed appearance and a rapid pulse.

Caffeine Intake

Some persons with HD become compulsive about consuming things such as pop or cigarettes. Caffeinated drinks can become a problem, both in the stimulating effect of the drug and in dehydration. Ask the person with HD (or a family member or caregiver) about the amount of caffeine the person is ingesting daily. Suggest switching to decaffeinated beverages.

Food Reactions

Some persons with HD try herbs and special diets as possible ways to slow the progression of the disease. Unless obviously dangerous, these should not be forbidden but discussed during a medical appointment. Some herbs and foods, such as grapefruit juice, have major interactions with some medications.

Undetected Visual or Hearing Impairments

Persons with HD may have undetected vision and hearing impairments, either because of communication problems or unawareness. Uncorrected, these can lead to frustration and behaviour problems. Vision and hearing should be checked and problems corrected if possible.

Feelings of Loss and Grief

The diagnosis and progression of HD entails loss, and a natural reaction to loss is grief. Research suggests that individuals progress through many stages of loss: denial, anger, bargaining, depression and ultimately acceptance (Kübler Ross, 1997). Coping with loss and grief is one of the greatest human challenges, and emotional outbursts may be a symptom of grief as well as a symptom of HD. Recommend a consultation with a spiritual or grief counsellor who may be able to help the person with HD address their feelings of grief.

Stage of the Disease

The type, frequency and severity of responsive behaviours that occur often vary with the stage of disease. One survey suggested that persons with early HD are more concerned about depression, anxiety and apathy, whereas persons with later-stage HD report agitation, irritability and disinhibition as the most prominent behavioural concerns. The responses to the various stages of HD, however, can vary greatly from person to person. Consider the person's stage of disease progression to better understand the behaviours that are occurring (see Section II, The Stages of HD, page 9).

Special Topic - A General Approach to Responsive Behaviours in HD

Outbursts and other troubling behaviours can be the most disruptive symptoms of HD. If the cause or trigger can be determined, it becomes easier to cope with the behaviour and sometimes easier to reduce the frequency of the responsive behaviour.

Below are some general guidelines that caregivers can use to help identify the causes or triggers for responsive behaviours. If causes are identified, they can often be avoided or managed more successfully.

Step 1: Identify the main problem. Is the problem observable? Is it measurable? Can others see it? Irritability may be occurring because the person's immediate physical needs are not being met.

Step 2: Gather information about the problem and try to break the difficulty down into separate and distinct components. If there is no immediate physical reason for the behaviour, there may still be an identifiable trigger.

- When does the responsive behaviour occur?
- Where does the responsive behaviour occur?
- What precedes the behaviour?
- Who was involved?

- What follows the responsive behaviour?
- What emotion (e.g. fear, anger or frustration) was expressed?

Suggest that the caregiver keep a private diary or log of the behaviour, when and where it happened and who was there. You can review the log with the caregiver at a future appointment.

Step 3: Review possible causes of responsive behaviours with the caregiver, including the stage of the disease, environmental causes, individual contributions and other health-related considerations. A pattern may appear that reveals the trigger for the responsive behaviour.

Step 4: Develop a list of possible strategies to address the responsive behaviour with the caregiver. Suggest environmental changes if needed.

Step 5: Encourage the caregiver to be flexible. Remind the caregiver that it may take several strategies to address the behaviour.

Step 6: Reassure the caregiver after a stressful event that you empathize with the challenges and are trying to understand how to make things better. Encourage the caregiver to ask for advice, assistance and support.

Step 7: Call in other medical professionals if needed, including mental health professionals, speech language pathologists, nutritionists, occupational therapists or others who may be able to alleviate an underlying problem or manage the responsive behaviour medically.

Conclusion & References



Conclusion

Many different behaviours have been discussed in this handbook. No single person with HD will display all, or even most, of the behaviours that have been mentioned. This book was created to present a broad picture of behaviour in HD. Understanding and managing these behaviours is a dynamic process, as symptoms will change over the course of the disease.

The Huntington Society of Canada (HSC) is here to support persons with HD, and the families and health care professionals who care for persons affected by HD. The Family Services team at HSC is available to consult with you about the challenges of understanding and managing behaviour in HD. There are many resources available, and a strong HD community can provide information and support.

For more information about available resources, visit www.huntingtonsociety.ca or call the Huntington Society of Canada at **1-800-998-7398**.

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