

# Cognitive Symptoms

*The Cognitive Changes Associated with Huntington's disease*

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Huntington's Disease (HD), an inherited neurodegenerative disorder, damages specific areas of the brain, resulting in movement difficulties as well as cognitive and [behavioral changes](#). The term "cognitive" refers to tasks of the brain that involve knowing, thinking, remembering, organizing, and judging. Certain changes in cognitive abilities are characteristic of HD and can significantly impact the lives of individuals with the disease. For example, cognitive changes may affect the ability of a person with HD to work, manage a household or properly care for him or herself.

- [What are some of the cognitive abilities that may be impaired by HD?](#)
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# Cognitive Symptoms Part 1

*The Cognitive Changes Associated with Huntington's disease*

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What are some of the cognitive abilities that may be impaired by HD?

- [Communication](#)
- [Memory](#)
  - Ability to learn new information
  - Ability to recall information
- [Awareness](#)
- [Organization](#)
  - Decision making
  - Planning, sequencing & prioritizing
  - Attention/concentration
- [Regulation](#)
  - Repetition
  - Initiation

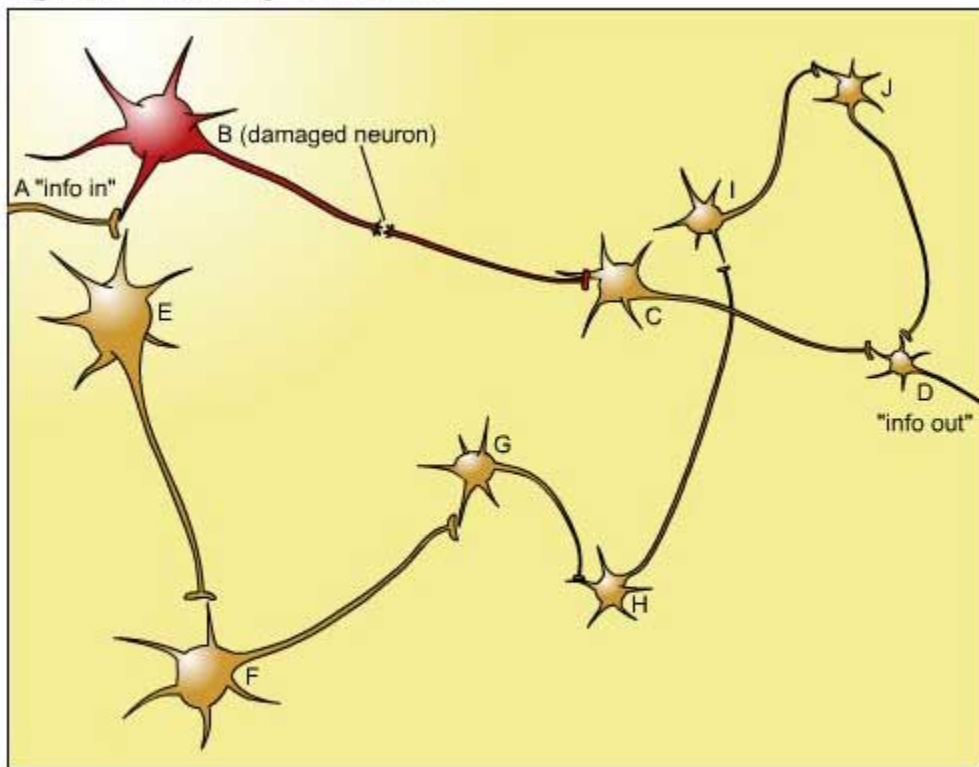
- Temper Control
- [Visual Spatial Ability](#)

## Communication

Communication is a complex process, requiring the cognitive ability to express and understand as well as physical abilities such as muscle control and breathing. Typically, neural degeneration, resulting from HD, begins in the core of the brain at the [caudate nucleus](#) and may spread to areas on the left and right side of the brain, such as the control centers for cognitive function, speech and language. Thus, communication problems tend to become more prominent as the disease progresses.

Throughout the course of the disease, communication problems vary in nature and severity. However, variations in the nature and severity of communication problems also occur from person to person. While one individual may have difficulty initiating conversation, another may have very little difficulty initiating, but severe difficulty word-finding (an aspect of memory recall). Although there are a number of communication problems that may arise for people with HD, the most common communication difficulties are four: speaking clearly, initiating conversation, organizing what is to be said and understanding what is being said.

Figure X-1: Damaged Neurons



Damaged neurons make it more difficult for information to be sent quickly along a neural "circuit." In this case, the information comes in through neuron A and exits through neuron D. However, the normal route of B-C-D is hampered because neuron B is damaged. Therefore, the information must be shuttled through a different, longer route (E-F-G-H-I-J-D).

As HD damages [neurons](#) in the [caudate](#), proper regulation of motor information that tells the body how to move specific muscles at precise times may be impaired. The caudate's inability to regulate motor information can result in slurred speech and stuttering as well as the uncontrolled bodily movements, often referred to as [chorea](#).

The ability to initiate conversation or activities is a very complex brain function. Damage to the caudate affects the brain's ability to regulate the sequence and amount of information being transmitted, which may result in difficulty starting and stopping communication. The inability to initiate conversation may also be the result of word-finding difficulty. As neurons in the caudate die, the intact neurons have more difficulty sending information along the neural "circuit." For example, it may take longer than expected for a person with HD to answer a question because it may be more difficult to find the right word. While word-finding is often impaired, knowledge of vocabulary is retained.

As the caudate and its connections with other areas of the brain deteriorate, some kinds of information may not reach the [frontal lobes](#). Without the frontal lobes to sequence and prioritize outgoing information, the speech of a person with HD may become garbled or seemingly illogical. Damage to the caudate, resulting in impaired access to the frontal lobes may also make it difficult for an HD-affected person to understand what is being said; however, the ability to understand usually remains intact, even in the later stages of the disease. For example, each word of a sentence may be understood, but the frontal lobes and caudate may not be able to organize them properly, possibly resulting in miscommunication. This inability to organize incoming information can also contribute to a slowed response time, even if comprehension remains normal.

If you are interested in reading about strategies and tools that may improve communication for and with a person who has HD, click [here](#).

## Memory

An individual suffering from the cognitive symptoms of HD may have memory difficulties. It is important to note that the memory problems that can occur in people with HD are different from the memory difficulties that can occur in people with Alzheimer's Disease. Whereas people with Alzheimer's Disease may get lost in familiar places or forget the names of familiar people, individuals with HD will know and recognize people as well as places.

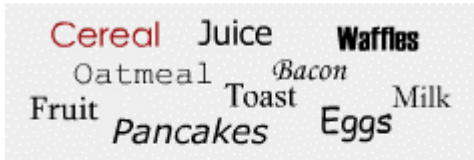
Table X-1: Comparison of Huntington's Disease & Alzheimer Disease		
Ability	Huntington's Disease	Alzheimer Disease
Speed of processing	Slow, mostly accurate	Slow; often inaccurate
Speech output	Slurred & slow; accurate	Normal rate & clarity; often inaccurate
Learning new information	Disorganized & slow; can learn	Rapid forgetting; cannot store information
Free recall	<u>Impaired:</u> <ul style="list-style-type: none"> <li>• cannot find the right words</li> <li>• can recognize with choices</li> <li>• benefits from cues</li> </ul>	<u>Impaired:</u> <ul style="list-style-type: none"> <li>• cannot recall memories</li> <li>• cannot recognize with choices</li> <li>• does not benefit from cues</li> </ul>
Motor memory	<u>Impaired:</u> <ul style="list-style-type: none"> <li>• cannot learn or recall motor memories</li> </ul>	<u>Impaired:</u> <ul style="list-style-type: none"> <li>• can learn &amp; retain motor memories</li> </ul>

(Table adapted from Paulsen *Understanding Behavior in Huntington's Disease*)

Throughout the course of HD, there are two primary memory difficulties that result from cognitive impairment: learning new information and recalling stored information. The impaired ability to learn new information may be the result of damaged [neural connections](#) between the [frontal lobes](#) and the [caudate](#) in the brain. Without efficient use of the frontal lobes, the brain cannot effectively organize and sequence the information to be learned. For example, learning a new phone number may be very difficult for an individual with HD because the brain may not organize or group the numbers together in a way that is easy to remember. For example, the series of numbers “3456978” is much more difficult to remember than “3-4-5-69-78.” When information is not organized in an efficient manner, retaining and recalling the learned information is very difficult.

Figure X-2: Recognition vs. Recall

1) What did you have for breakfast this morning?



2) Did you have "toast" or "cereal" for breakfast this morning?



Although the stored memories remain intact, it is often difficult for a person with HD to recall the correct one, especially if there are a lot of choices. However, if the person is prompted and the choices named, they may have more success recognizing the correct response instead of recalling it.

Recalling stored information is the other primary memory problem for people with HD. For example, a hypothetical person, Silvia, knows what she had for dinner last night, but may not respond very quickly when asked. However, if you ask her whether she had pizza or chicken, she'll be able to correctly identify which of the two choices she had for dinner. The [neurodegenerative](#) nature of HD disrupts the brain's search mechanism, which makes recalling stored information more difficult, although the memory likely remains intact and can often be recalled through cues or recognition. Also, the person suffering from memory difficulties usually maintains the ability to understand and comprehend information.

Although most memories remain intact, motor memories are often impaired. Motor memories, such as driving a car or tying shoes, are considered **implicit** or "unconscious" memory. The impairment of these motor memories means that a person has to rely on "conscious memory" to perform these tasks, which requires more concentration. Since these simple, once automatic, tasks may require more concentration, people with HD often have difficulty multi-tasking or dividing their [attention](#). For example, an individual suffering from memory problems due to HD may have difficulty making dinner while listening to the radio.

Recognition memory: stored information can be recalled through a cue. For example, an individual may not remember what time his haircut appointment is scheduled for, but when asked, “are you getting your haircut at 1:00 or 2:00?” he remembers that the appointment is at 2:00

Long-term memory: stores an unlimited amount of rehearsed information; each memory can be stored for a long period of time

Language comprehension: ability to understand the meaning of words as well as how they are organized in order to understand what is being said

Memory retrieval: recalling stored information

Verbal fluency: ability to use and organize words in order to clearly express thoughts, feelings and ideas

Word finding: recalling and using the proper word to communicate

## Executive Functions

Critical to our ability to function effectively at home or work, the “executive functions” include prioritizing, problem solving, judgment, abstract thinking, controlling emotions and awareness of self and others. The [frontal lobes](#), often referred to as the “boss” of the brain, are in charge of the executive functions. The part of the brain responsible for regulation information being sent to the frontal lobes is the [caudate nucleus](#). When HD destroys neurons in the caudate nucleus, a person with HD may have difficulty efficiently performing tasks that were previously simple, such as running errands.

Many of the executive functions that may be impaired in individuals with HD fall into one of three categories: awareness, organization and regulation.

### Awareness

Commonly, denial is used to describe the inability to accept the reality of a distressing circumstance. HD sufferers may deny having HD or be unable to recognize their disabilities. However, this denial is not under the individual’s control, so a lack of awareness or “unawareness” may be a more accurate word for people with HD.

Due to HD, circuits connecting the caudate nucleus, frontal, and [parietal lobes](#) may incur damage, resulting in a lack of self-awareness. People with HD may be unable to recognize disabilities or evaluate their own behavior. The inability to evaluate one's own performance may cause sufferers to be unaware of mistakes that are evident to others. Damage to these [neural connections](#) may also impair the ability to experience a range of subtle emotions and see another's point of view, possibly making social and personal relationships more difficult.

Unawareness often plays a role in seemingly irrational behaviors. For example, a person may become upset if he or she is not allowed to go back to work or live independently, because of the unawareness of failing capabilities. However, a person may be willing to talk about his or her capabilities, but still be unable to acknowledge that failing capabilities are the result of HD. Unawareness, a behavioral as well as a cognitive symptom, is generally accepted as an untreatable component of HD. To learn about the behavioral symptoms of HD, click [here](#).

## Organization

Since HD damages the [caudate nucleus](#), many aspects of behavioral and intellectual functioning can be affected. The task of the caudate is to organize, regulate and prioritize information transmitted from many areas of the brain to the [frontal lobes](#). If the information reaching the frontal lobes is not organized as a result of HD, the individual with HD may experience difficulty organizing his/her thoughts and activities as well.

In order to plan and prioritize efficiently, our brain must be able to organize activities in a logical order, evaluate all of the steps involved in accomplishing a task, and even think about one particular task while performing another. As a result of the damaged caudate, the brain of an individual with HD may not be capable of performing in such a manner. For example, a person without HD may spend one hour on a trip to the grocery store and the bank. However, it may take a person with HD two or three hours to accomplish this same task. While at the grocery store he or she may have to look for each item in the order of the list, possibly failing to get two items from the same aisle because they appeared in different places on the list.

A diminished ability to make decisions may also become a problem as a result of the brain's failing organizational capabilities. If asked the question, "What would you like to have for dinner?" it may take a while for the brain of a person with HD to organize the words into an understandable question, retrieve the memories of past dinner items, process the feelings regarding each dinner item, and organize the words into a logical response. This difficulty may also be due to memory impairments resulting from HD. Thus, the process of decision making is drastically simplified if a person with HD is given choices, which allows the brain to recognize memories rather than retrieve them. Shorter sentences may also aid in the decision making process, as they contain fewer words for the brain to organize.

Another function affected by the impairment of the brain's organizational capacity is *attention*. While simple attention, the ability to focus on one activity, often remains intact, sustained attention as well as divided attention may become impaired. As a result of memory impairments, "unconscious" tasks that were once automatic may require intense concentration. This makes dividing one's attention very difficult. For example, it may be difficult for a person with HD to walk while carrying on a conversation. With the loss of motor memories, he or she may have to consciously think about each step forward, making conversation difficult. To read more about memory impairment as a cognitive symptom of HD, click [here](#).

## Regulation

The [caudate nucleus](#) serves primarily as a regulator and organizer. It controls the order and amount of information traveling from particular areas of the brain to the [frontal lobes](#). As HD progressively destroys the caudate, it may become difficult for individuals with HD to initiate, maintain, and/or stop behaviors or thoughts.

As mentioned above, the ability to initiate activities or conversation is a complex brain function. Damage to the caudate disrupts the brain's ability to regulate the sequence and amount of information being transmitted, which may result in difficulty starting and stopping communication or activities. The diminished regulatory abilities of the caudate may also result in the inability to maintain an activity or conversation. However, this may be due to the impairment of sustained attention as well. For example, an individual with HD may be able to begin folding laundry but quickly become unable to focus on the task at hand due to distractions. If the radio is playing in the room, the individual may focus his or her attention on the music and be unable to re-initiate and complete the task of folding laundry.

Another possible result of the caudate's inability to regulate the amount of information traveling to the frontal lobes is a lack of emotional control. A person with HD may over-express a feeling of slight frustration or irritation in the form of a temper tantrum or aggressive behavior. Although the emotion itself is often a legitimate response to something in the individual's environment, the caudate cannot regulate the proper amount to be expressed. To read about the behavioral symptoms of HD, including frustration, apathy and others, click [here](#).

## Visual Spatial Ability

Visual spatial ability is the ability to perceive one's body position in the environment. An individual's perception of his or her body position is useful for judgment of where he or she is in relation to walls or how close his or her hand is to a burner on the stove. Impaired visual spatial ability is often evident even in the early stages of HD. Most commonly, the individual suffering from cognitive symptoms of HD is aware of his or her visual spatial impairment.

For example, due to a diminished visual spatial ability, it may be more difficult for a person with HD to read a map or follow directions, since most directions are given using spatial cues, such as “east” and “west” or distances measured in miles. However, a person suffering from this cognitive symptom of HD may be able to follow directions if they are given using geographic markers, such as: “Go straight on Campus Drive until you reach a stoplight. Turn left and go past the Pet Store. The Post Office will be on the left side of the street with a flag pole in front.” For an individual suffering from visual spatial impairment, directions using “left” and “right” or geographic markers are easier to follow because they do not require the individual to orient his or her body in a particular direction. Regardless of which direction a person is facing, “left” is one way and “right” is the other. However, depending on the orientation of one’s body, “east” may be behind, to the right, to the left or in front of him.

Reading difficulties may also be the result of visual spatial impairment; however, the inability to maintain attention may be a contributing factor as well. For information about attention impairments as a cognitive symptom of HD, click [here](#).

## **Cognitive Symptoms**

### **Part 2**

*The Cognitive Changes Associated with Huntington's disease*

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What causes the cognitive symptoms?

As a [neurodegenerative](#) disease, HD damages many [neurons](#) and [neural connections](#) within the brain, potentially causing cognitive impairment. Most of the damage occurs in the [caudate nucleus](#) and [putamen](#), which are structures of the [basal ganglia](#). To learn more about these brain structures, click [here](#). The primary function of the caudate is the regulation and organization of information being transmitted to the [frontal lobes](#) from other areas of the brain. The frontal lobes are responsible for many important tasks, some of which are:

- Organizing
- Prioritizing
- Controlling impulses
- Self-awareness
- Initiating and ending activities

Thus, damage to the many connections between the caudate and frontal lobes can significantly impair cognitive abilities, such as reasoning, planning, attention, memory, and learning. To read about neurons and neural connections, click [here](#).

The neurodegenerative changes that occur within the brain of a person who has HD are generally the primary cause of the cognitive symptoms of HD, as well as behavioral changes and movement difficulties. An individual suffering from the cognitive symptoms of HD may have difficulty effectively prioritizing his or her daily activities, initiating conversation or activities, recalling memories or making decisions. However, it is important to remember that the cognitive as well as behavioral and physical symptoms of HD vary from person to person. To learn about the behavioral symptoms of HD, click [here](#).

## Cognitive Symptoms Part 3

*The Cognitive Changes Associated with Huntington's disease*

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How do the cognitive symptoms change as HD progresses?

As a general rule cognitive impairments tend to increase in severity as HD runs its course. However, only a few longitudinal studies have been done on the cognitive symptoms of HD, and thus, research has not determined whether the severity of a cognitive symptom can be used as a marker for the underlying progression of the disease.

Although the symptoms of HD vary significantly from person to person, there are some general trends among individuals. Speed of mental processing, organization, and initiation are commonly impaired early in HD and may worsen during the intermediate stages. While individuals with HD are often unable to speak or express their views in the later stages of HD, some cognitive abilities, such as the ability to understand incoming information, may remain relatively intact.

Early Stage	Intermediate Stage	Late Stage
<ul style="list-style-type: none"> <li>• Difficulty organizing routine tasks</li> <li>• Difficulty coping with changes in routine or new situations</li> <li>• Impaired decision-making and attention</li> </ul>	<ul style="list-style-type: none"> <li>• Increased difficulty working</li> <li>• Increased difficulty managing a household</li> <li>• Usually capable of carrying out daily living activities such as self-care</li> </ul>	<ul style="list-style-type: none"> <li>• Can no longer manage daily living activities</li> <li>• Often require professional nursing</li> <li>• Swallowing and communication difficulties are common</li> </ul>

## **Cognitive Symptoms Part 4**

*The Cognitive Changes Associated with Huntington's disease*

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Do the cognitive symptoms of HD vary from person to person?

The expression of HD varies significantly from person to person. Although HD is a progressive disease for affected individuals, there is considerable variation in the type and severity of symptoms a person with HD may experience. Some individuals may experience a number of cognitive and behavioral symptoms and fewer physical symptoms, whereas others may suffer more from physical symptoms, such as [chorea](#). The variation in severity means that while some of the cognitive symptoms may be quite pronounced for one person, those particular symptoms may be much less evident in another.

Due to the variation in the type and severity of cognitive symptoms, it may not be useful to use them as an indicator for the onset of HD in an at-risk individual or to diagnose the individual with the disease. Many of the early cognitive symptoms of HD, such as forgetfulness, lack of initiation or fumbling are also fairly common among individuals who are not at risk for HD. “Symptom watching” by individuals at risk for HD may result in a misinterpretation of these thoughts, actions or behaviors as HD. Genetic counselors may be contacted if symptom-watching or anxiety due to being at-risk for HD begins to interfere with one’s ability to function effectively.

## **Cognitive Symptoms Part 5**

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Are there treatments available for the cognitive symptoms of HD?

At the time of this writing (April 2003), there is no cure for the cognitive symptoms of HD or the disease itself. The cognitive symptoms of HD are due to the damage of [neurons](#) and [neural connections](#) in the brain, which at this time are considered irreversible. However, scientists and researchers continue to investigate the brain’s ability

to produce new neurons as well as its ability to form new connections between neurons. For more information about the brain's natural reparatory ability, click [here](#).

Fortunately, there are a number of strategies for coping with and enhancing cognitive abilities impaired by HD. For example, maintaining a calm, predictable environment and establishing routines can improve organization and planning as well as minimize the occurrence of emotional outbursts. A predictable, routine environment enables a person suffering from the cognitive symptoms of HD to organize daily tasks and adhere to that schedule, resulting in fewer organizational or planning problems. There are a number of resources that provide strategies for improving the cognitive symptoms of HD. If you are interested in learning more about these strategies, click [here](#).

Although there are strategies and treatments that can improve the physical, behavioral and cognitive symptoms of HD, there are currently no treatments available that slow down the progression of HD. However, research continues with the growing hope of discovering effective treatments as well as a cure for HD. For more information on potential treatments for HD, click [here](#).

-K.Hammond 3-29-03

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For further reading:

1. American Speech-Language-Hearing Association. Huntington's Disease. <http://www.asha.org/speech/disabilities/Huntington-Disease.cfm>  
*Much like a brochure, this resource explains many of the communication and swallowing problems that may occur as a result of HD. It also lists ways in which a speech-language pathologist can help minimize difficulties.*
2. Australian Huntington's Disease Association. Information – Huntington's Disease <http://www.ahda.asn.au/content/pages/page.asp?iPage=5>  
*This website provides basic information about HD as well as support for individuals with HD and their families.*
3. Huntington's Disease Association. Fact Sheet 10. Behavioral Problems in Huntington's Disease. <http://www.hda.org.uk/charity/download.html>  
*This clearly-written Fact Sheet is just one of the many informative and helpful Fact Sheets available at this site.*
4. Paulsen, Jane S. "Understanding Behavioral Changes in HD: The Neuropsychology of HD and Strategies for Intervention." International Huntington Association, 2002. <http://www.huntington-assoc.com/jane.html>
5. Paulsen, J. "Understanding Behavior in Huntington's Disease" (2nd ed.). Huntington's Disease Society of America, Inc, 1999.  
*This is an easy to read and extremely informative resource with the purpose of providing practical and helpful information about the behavioral as well as cognitive symptoms of HD.*

6. Rosenblatt, A., Ranen, G., Nance, M. & Paulsen, J. "A Physician's Guide to the Management of Huntington's Disease." (2nd ed.) Huntington's Disease Society of America, Inc., 1999.

*This is a guide specifically for physicians. It explains the symptoms of Huntington's Disease and methods for providing optimal management of the symptoms.*

7. South Shore Neurologic Associates (SSNA). HD: Cognitive Disorders.

<http://www.southshoreneurologic.com/move-dis/hd/hd-cognitivedisdr.html>

*The aim of this site is to provide information and support for individuals with neurological disorders. While there is a substantial amount of information on HD, the information itself isn't well-organized. However, this site is still a valuable resource for those affected by HD.*