Alzheimer Society

Huntington disease

Huntington disease is a hereditary disease that causes cognitive, emotional, and physical changes. Huntington disease is progressive, meaning that symptoms get worse over time. Although Huntington disease is primarily thought of as a condition that affects movement, the damage to the brain also results in symptoms of dementia, such as a decline in memory and challenges in judgment.

Huntington disease is a genetic disorder.

In your chromosomes, there is a gene responsible for making a protein called **huntingtin**. One of the functions of the huntingtin protein is DNA damage repair. When the protein is not working properly, it can cause certain parts of the brain to die. As the brain cells die, the person experiences physical, cognitive, and emotional symptoms of Huntington disease.

The Huntington disease mutant gene is dominant, which means that each child of a parent with Huntington disease has a 50% chance of inheriting the disease and is said to be at-risk[1].

Symptoms usually begin in people between the ages of 30 and 55.

When a person under 20 years of age is diagnosed with Huntington disease, they have **Juvenile Huntington disease**, while someone diagnosed over the age of 60 has **Late Onset Huntington disease**.

Symptoms usually progress over a long period of time – around 10 to 25 years. Because early symptoms can appear mild at first, Huntington disease may be initially overlooked or mistaken for other conditions.

Huntington disease is diagnosed through genetic testing.

Genetic testing is typically done through a genetic clinic and it includes genetic counselling. A **genetic counsellor** can tell you whether genetic testing is relevant for you based on your family history.

They will also make sure you know what supports are available before, during, and after the testing process. To find a genetic counsellor near you, visit cagc-accg.ca/?page=225

Symptoms of Huntington disease[1]

Early stage

- Decreased attention to detail
- Minor involuntary movements such as twitching of the limbs or excessive restlessness
- Depression and irritability

Intermediate stage

- Personality changes, such as irritability and mood swings
- Weight loss
- Difficulties with memory, concentrating or making decisions
- Depression and anxiety
- Obsessive-compulsive behaviours, such as continually repeating the same activity
- Increased difficulty with swallowing and speech
- More obvious involuntary movements of the head, neck arms and legs called "chorea" which means "dance"

Advanced stage

- Increased difficulty with swallowing and increased risk of choking and aspiration pneumonia
- Decrease in involuntary movements and increase in rigidity and muscle contracture
- Decreased ability to communicate, but understanding what is being said remains possible

Not everyone living with Huntington disease will experience the same presentation and progression of symptoms.

1. This content is from experts from the Huntington Society of Canada "What is Huntington Disease" fact sheet. © Huntington Society of Canada, July 2021. Reproduced with permission. Adapted with permission.

Predictive genetic testing is available for adults who are not showing signs of the disease, but have a family history of the disease. In this case, a result of "mutation positive" or "mutation negative" is given, but a Huntington disease diagnosis is not given until symptoms develop.

Brain imaging tools that let us take a closer look at the brain, such as magnetic resonance imaging (MRI), are not usually needed. However, a doctor may request brain imaging to detect any structural changes in the parts of the brain that are affected by Huntington disease or to rule out other conditions.

There are treatments that can manage the symptoms of Huntington disease.

There are currently no treatments to slow or stop the damage caused by the disease. However, there are strategies to help manage some of the symptoms:

- **Occupational therapy and assistive devices** can help improve functional ability.
- **Physical therapy** can help maintain physical abilities.
- **Speech therapy** can help improve verbal communication and can address eating and swallowing challenges.
- **Certain medications** can help with the psychiatric symptoms such as depression and anxiety.

Since Huntington disease develops differently in each person, it's important that caregivers and professionals work together to help manage the most effective treatment for each individual.

There are many things a person can do to manage the dementia symptoms associated with Huntington disease, including making healthy food choices, being physically active, challenging their brain and participating in social activities.

For brain-healthy choices and tips, please visit alzheimer.ca/brainhealth.

Support is available.

Contact your local Alzheimer Society for more information about dementia.
Visit alzheimer.ca/helpnearyou

Additional resources.

Contact the Huntington Society of Canada for more information about Huntington disease.

Visit huntingtonsociety.ca

Visit brainxchange.ca to search for webinars on Huntington disease.

This resource is informed by research and the experiences of people living with dementia and their caregivers. We thank Dr. Ray Truant, Professor, McMaster University, and Angèle Bénard, RSW, Huntington Society of Canada, for their generous contribution to the development of this resource.

To provide feedback on this factsheet, please email publications@alzheimer.ca

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