Huntington's disease

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Overview

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Huntington's disease is an inherited disease that causes the progressive breakdown (degeneration) of nerve cells in the brain. Huntington's disease has a broad impact on a person's functional abilities and usually results in movement, thinking (cognitive) and psychiatric disorders.

Most people with Huntington's disease develop signs and symptoms in their 30s or 40s. But the disease may emerge earlier or later in life.

When the disease develops before age 20, the condition is called juvenile Huntington's disease. An earlier emergence of the disease often results in a somewhat different set of symptoms and faster disease progression.

Medications are available to help manage the symptoms of Huntington's disease, but treatments can't prevent the physical, mental and behavioral decline associated with the condition.

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Symptoms

Huntington's disease usually causes movement, cognitive and psychiatric disorders with a wide spectrum of signs and symptoms. Which symptoms appear first varies greatly among affected people. During the course of the

disease, some disorders appear to be more dominant or have a greater effect on functional ability.

Movement disorders

The movement disorders associated with Huntington's disease can include both involuntary movement problems and impairments in voluntary movements, such as:

- Involuntary jerking or writhing movements (chorea)
- Muscle problems, such as rigidity or muscle contracture (dystonia)
- Slow or abnormal eye movements
- Impaired gait, posture and balance
- Difficulty with the physical production of speech or swallowing

Impairments in voluntary movements — rather than the involuntary movements — may have a greater impact on a person's ability to work, perform daily activities, communicate and remain independent.

Cognitive disorders

Cognitive impairments often associated with Huntington's disease include:

- Difficulty organizing, prioritizing or focusing on tasks
- Lack of flexibility or the tendency to get stuck on a thought, behavior or action (perseveration)
- Lack of impulse control that can result in outbursts, acting without thinking and sexual promiscuity
- Lack of awareness of one's own behaviors and abilities
- Slowness in processing thoughts or "finding" words
- Difficulty in learning new information

Psychiatric disorders

The most common psychiatric disorder associated with Huntington's disease is depression. This isn't simply a reaction to receiving a diagnosis of Huntington's disease. Instead, depression appears to occur because of injury to the brain and subsequent changes in brain function. Signs and symptoms may include:

- Feelings of irritability, sadness or apathy

- Social withdrawal
- Insomnia
- Fatigue and loss of energy
- Frequent thoughts of death, dying or suicide

Other common psychiatric disorders include:

- **Obsessive-compulsive disorder** — a condition marked by recurrent, intrusive thoughts and repetitive behaviors
- **Mania,** which can cause elevated mood, overactivity, impulsive behavior and inflated self-esteem
- **Bipolar disorder** — a condition with alternating episodes of depression and mania

In addition to the above symptoms, weight loss is common in people with Huntington's disease, especially as the disease progresses.

Symptoms of juvenile Huntington's disease

The start and progression of Huntington's disease in younger people may be slightly different from that in adults. Problems that often present themselves early in the course of the disease include:

Behavioral changes

- Loss of previously learned academic or physical skills
- Rapid, significant drop in overall school performance
- Behavioral problems

Physical changes

- Contracted and rigid muscles that affect gait (especially in young children)
- Changes in fine motor skills that might be noticeable in skills such as handwriting
- Tremors or slight involuntary movements
- Seizures

When to see a doctor

See your doctor if you notice changes in your movements, emotional state or mental ability. The signs and symptoms of Huntington's disease can be caused by a number of different conditions. Therefore, it's important to get a prompt, thorough diagnosis.

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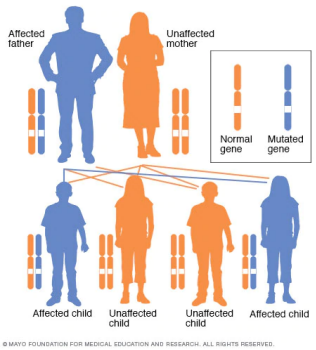
Causes

Huntington's disease is caused by an inherited defect in a single gene. Huntington's disease is an autosomal dominant disorder, which means that a person needs only one copy of the defective gene to develop the disorder.

With the exception of genes on the sex chromosomes, a person inherits two copies of every gene — one copy from each parent. A parent with a defective gene could pass along the defective copy of the gene or the healthy copy. Each child in the family, therefore, has a 50 percent chance of inheriting the gene that causes the genetic disorder.



Autosomal dominant inheritance pattern
In an autosomal dominant disorder, the mutated gene is a dominant gene located on one of the nonsex chromosomes (autosomes). You need only one mutated gene to be affected by this type of disorder. A person with an autosomal dominant disorder — in this case, the father — has a 50% chance of having an affected child with one mutated gene (dominant gene) and a 50% chance of having an unaffected child with two normal genes (recessive genes).

Complications

After the start of Huntington's disease, a person's functional abilities gradually worsen over time. The rate of disease progression and duration varies. The time from disease emergence to death is often about 10 to 30 years. Juvenile Huntington's disease usually results in death within 10 years after symptoms develop.

The clinical depression associated with Huntington's disease may increase the risk of suicide. Some research suggests that the greater risk of suicide occurs before a diagnosis is made and in the middle stages of the disease when a person has begun to lose independence.

Eventually, a person with Huntington's disease requires help with all activities of daily living and care. Late in the disease, he or she will likely be

confined to a bed and unable to speak. However, he or she is generally able to understand language and has an awareness of family and friends.

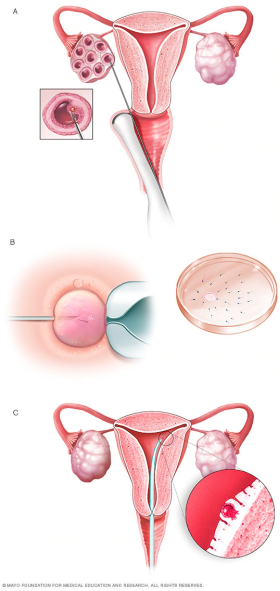
Common causes of death include:

- Pneumonia or other infections
- Injuries related to falls
- Complications related to the inability to swallow

Prevention

People with a known family history of Huntington's disease are understandably concerned about whether they may pass the Huntington gene on to their children. These people may consider genetic testing and family planning options.

If an at-risk parent is considering genetic testing, it can be helpful to meet with a genetic counselor. A genetic counselor will discuss the potential risks of a positive test result, which would indicate the parent will develop the disease. Also, couples will need to make additional choices about whether to have children or to consider alternatives, such as prenatal testing for the gene or in vitro fertilization with donor sperm or eggs.



In vitro fertilization
During in vitro fertilization, eggs are removed from mature follicles within an ovary (A). An egg is fertilized by injecting a single sperm into the egg or mixing the egg with sperm in a petri dish (B). The fertilized egg (embryo) is transferred into the uterus (C).

Another option for couples is in vitro fertilization and preimplantation genetic diagnosis. In this process, eggs are removed from the ovaries and fertilized with the father's sperm in a laboratory. The embryos are tested for presence of the Huntington gene, and only those testing negative for the Huntington gene are implanted in the mother's uterus.

By Mayo Clinic Staff

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