Huntington's Disease

National Institute of Neurological Disorders and Stroke (NINDS)

Source

National Institute of Neurological Disorders and Stroke (NINDS). Huntington's Disease Information Page.

Huntington's disease (HD) is an inherited disorder that causes brain cells, called neurons, to die in various areas of the brain, including those that help to control voluntary (intentional) movement. Symptoms of the disease, which gets progressively worse, include uncontrolled movements (called chorea), abnormal body postures, and changes in behavior, emotion, judgment, and cognition. People with HD also develop impaired coordination, slurred speech, and difficulty feeding and swallowing. HD typically begins between ages 30 and 50. An earlier onset form called juvenile HD occurs under age 20. Its symptoms differ somewhat from adult onset HD and include rigidity, slowness, difficulty at school, rapid involuntary muscle jerks called myoclonus, and seizures. More than 30,000 Americans have HD.

Huntington's disease is caused by a mutation in the gene for a protein called huntingtin. The defect causes the cytosine, adenine, and guanine (CAG) building blocks of DNA to repeat many more times than is normal. Each child of a parent with HD has a 50-50 chance of inheriting the HD gene. A child who does not inherit the HD gene will not develop the disease and generally cannot pass it to subsequent generations. A person who inherits the HD gene will eventually develop the disease. HD is generally diagnosed based on a genetic test, medical history, brain imaging, and neurological and laboratory tests.