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May is Huntington's Disease Month

One of the most difficult things a person can experience is to watch a loved one deteriorate when suffering from a degenerative disease. Huntington's disease (HD) is one example of such a condition that may be more prevalent than most people imagine. Over ¼ million Americans are estimated to have this disorder, or to be "at-risk" of inheriting it. If a parent has Huntington's disease, each child has a 50% chance of inheriting the gene for this condition. If the gene is present, that person is certain to develop the disease.

Huntington's disease is a genetic, degenerative brain disorder that affects both the body and the mind. It is invariably fatal. However, while HD usually appears in middle age (30 - 45 years old), it has been known to manifest itself in children as young as 2 and adults as old as 84. Children who develop this condition do not survive to adulthood. It is likely to occur equally in men and women, in all races, and across all socio-economic classes.

Early symptoms of HD include depression, mood swings, clumsiness, involuntary twitching, and lack of coordination. As the disease progresses, cognitive, emotional, and motor disturbances become more and more evident. Over time, short-term memory becomes affected, the ability to concentrate decreases, and involuntary movements of the head, trunk, and limbs increase. Eventually, walking, speaking, and even eating become extremely difficult, and the person is no longer able to care for himself. According to the Huntington's Disease Society, the affected person continues to deteriorate until death results, usually from complications such as choking, infection, or heart failure.

As the disease worsens, frustration on the part of the patient increases and contributes to the emotional difficulties that may already exist, such as depression and irritability. Speech becomes slurred, making communication difficult. Motor skills diminish and the person becomes less and less able to care for him or herself, undermining the sense of self-worth. Even though one characteristic of HD is the emergence of a voracious appetite, the ability to swallow grows increasingly difficult. Despite the constant feeling of hunger, the amount of food consumed is usually inadequate, resulting in weight loss.

Although the gene for HD was discovered in 1993, no cure has been found to date. Treatment usually consists of a multi-disciplinary approach that targets the individual symptoms of the disease. Drugs can be useful in controlling some of the involuntary movements as well as the emotional conditions, and speech therapy can be helpful in improving speech and swallowing ability. Both the age of onset and the rate of progression of Huntington's disease vary from individual to individual, and the symptoms themselves often change within an individual.

Previously, it had been believed that the age of onset of Huntington's disease in an individual was determined exclusively by the make-up of the gene for this condition. However, very recent research has found that the age of onset is also affected by other conditions, such as environment and other genetic factors. This finding has provided new hope that even if a cure cannot be found, it may be possible to develop drugs that would delay the onset of the disease, perhaps even pushing it beyond the normal life

span. If that can be accomplished, a person carrying the fatal gene would be able to live an entire lifetime before the disease would have the opportunity to develop. Additionally, it may be possible to use gene therapy to "switch off" genes in an effort to slow down or even prevent the development of HD.

At this time, a genetic test is available to determine if a person at-risk has the gene for Huntington's disease. While this knowledge may affect a couple's decision to have children, many people choose not to be tested because of the numerous financial, legal, ethical, and personal ramifications that are involved.

To learn more about HD, contact the Huntington's Disease Society of America at 800-345-4372 (www.hdsa.org), the Huntington's Disease Association at 020-7223-7000 (www.hda.org), or the Hereditary Disease Foundation at 212-928-2121 (www.hdfoundation.org). Additional information is available about this and other public health topics at the Central Connecticut Health District at 860-721-2822 (www.ccthd.org).