



## Huntington's Disease (HD): An Introduction for Professionals

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Huntington's disease (HD) is a terminal neurodegenerative disorder that affects cognitive, motor, and behavioral functioning (Abel & Zukin, 2008; Lin & Beal, 2006). The disorder has long been suspected to be heritable and in 1993, after several decades of research, scientists identified the causal gene. The genetic mutation is transferred to offspring by a single gene from one parent. Carriers of the Huntington gene mutation have a 50% chance of passing on the mutation to every child (Andersson, Juth, Petersén, Graff, & Edberg, 2013). A laboratory blood test can be used to positively verify a diagnosis. Research has also shown that the onset and severity of Huntington's disease are indicated by the number of repeated CAG counts on chromosome 4 (Vassos, Panas, Kladi, & Vassilopoulos, 2008). Higher numbers of repeats indicate the possibility of earlier onset and more severe declines in functioning. The aims of the current article are to provide brief educational material regarding this disorder, to present accessible information for professionals working with individuals who are affected by Huntington's, and to encourage additional consultation regarding this topic.

Any person who has the faulty gene that causes HD will eventually exhibit symptoms of the disorder because the gene is dominant (Brouwer-DudokdeWit, Savenije, Zoetewij, Maat-Kievit, & Tibben, 2002). Most individuals with Huntington's disease are diagnosed in their 30s-50s, although rapidly progressing juvenile forms of the disease can be diagnosed during the early adolescent years (Scerri & Cassar, 2013). Regardless of the time of onset, cognitive changes may begin up to 15 years earlier (Nance, Paulsen, Rosenblatt, & Wheelock, 2011). Those suffering from juvenile onset HD tend to have a larger range of clinical symptoms than those suffering from adult onset HD. These clinical symptoms can include an increased likelihood of seizures, oral motor dysfunction, and increased behavioral disturbance

(Gonzalez-Alegre & Afifi, 2006; Nance & Myers, 2001). The expected lifespan for individuals suffering from HD is generally 15-20 years post-diagnosis (Krobitsch & Kazantsev, 2010).

Huntington's disease is typically first indicated by the declines in the ability to emotionally regulate, organize thoughts or spaces, and navigate complex decisions. These symptoms make early diagnosis difficult. The first visual symptom characteristic to HD is chorea. Chorea is an uncontrollable, jerky "dance like" movement (Nance & Myers, 2011). Individuals displaying choreic movement may appear to have a tic or twitch or appear intoxicated due to progressive loss of voluntary movements. Eventually, chorea becomes constant and has a significant effect on an individual's metabolic rate such that weight loss becomes common during the later stages (Krobitsch & Kazantsev, 2010).

There is presently no cure for Huntington's disease. As such, any treatments for HD focus on the reduction of symptoms, proactive awareness and management of the disease as it progresses, and palliative care (Murphy et al., 2015). Many of the associated psychiatric symptoms such as depression, anxiety, mania, obsessions, compulsions, and psychosis are manageable with medications, psychotherapy, or both (Nance & Myers, 2011). Individuals suffering from HD grow increasingly less able to verbalize their thoughts and process information. One of the greatest challenges identified by patients, family members, and caregivers is the adjustment to the changes in cognition and the continuously diminishing ability to communicate (Hartelius et al., 2010).

The latter stages of Huntington's are marked by severe declines in muscle coordination and pneumonia, which are the leading causes of death for those with HD (Heemskerk & Roos, 2012). Individuals may easily aspirate on food or water as they lose the ability to clear their air pathways. In addition, they are also particularly susceptible to similar airway blockages as a complication of pneumonia.

Researchers searching for treatments, or a complete cure, are working diligently to find some form of relief for patients with Huntington's. As many of the symptoms are shared with other neurodegenerative cognitive disorders, research focusing on other diseases (e.g., Alzheimer's, Parkinson's, ALS, MS) benefits the HD community. Similarly, HD research benefits the results of focused research on those disorders as well. The following four informational pieces are specifically designed to be of assistance to psychologists and therapists working with this population.

**1. HD is a complex genetic disease. The reasons for seeking therapy are equally complex and varied.** Affected individuals may deal with a wide array of concerns including grief, reproductive issues, substance abuse, irritability, apathy, paranoia, and end-of-life anxiety (Goh, 2011). Individuals within the family who have not been tested or diagnosed wrestle with anxiety about their own possible diagnosis. If those individuals have children, they also must consider the possibility of the children testing positive as well. By virtue of supporting a diagnosed family member, their own apprehension about genetic testing and fears that their own future is being foreshadowed can weigh heavily upon the at-risk individuals in various stages of family planning; they may struggle with personal and ethical dilemmas regarding prenatal and predictive genetic testing options. Individuals with HD may struggle with feelings of envy for those without HD and their own finite time to live. Testing negative also brings its own struggles. Individuals who have tested negative can wrestle with survivor's guilt (Meiser & Dunn, 2001).

**2. Psychiatric symptoms often occur 10 or more years earlier than HD onset (Paulsen et al., 2008).** Patients who have HD are often diagnosed with psychiatric disorders related to symptoms that display prior to the presentation of the symptoms characteristic of definitive Huntington's disease. This time period, known as the prodromal period, occurs before recognizable motor symptoms appear leading to the diagnosis of HD. It is marked by a number of cognitive and psychiatric symptoms that may occur (Van Liew, Gluhm, Goldstein, Cronan, & Corey-Bloom, 2013). Psychiatric symptoms that mimic depression and anxiety are often noticed first and can be treated through pharmacotherapy,

psychotherapy, behavioral modification strategies, family education, or some combination of those (Nance et al., 2011).

**3. Patients who have HD need a longer response time** when asked a question or responding during casual conversation. The wait time may be uncomfortable for some, but it is important to allow people with HD time to understand the question, search for an answer, and then articulate a response at their own pace (Nance et al., 2011).

**4. Suicide is more common among HD affected individuals** than the general population. It is a leading cause of death for individuals who are diagnosed with Huntington's disease (Booij, Engberts, Tibben, & Roos, 2013; Nance et al., 2011). Symptoms of depression are intensified by the loss of work, relationships, and the ability to care for oneself. Those at-risk for or diagnosed with HD should be asked about suicidal ideations on a routine basis (Robins Wahlin, Bäckman, Lundin, Haegermark, Winblad, & Anvret, 2000).

## Biographies

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