Huntington's Disease (HD): Facts Corrections Professionals Need to Know

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HD: Prevalence and Overview[i]

Huntington's disease (HD) is a genetic neurological disorder (Huntington's Disease Society of America (HDSA), 2012). It presents symptoms in three different categories: motor, cognitive, and psychiatric or behavioral (Nance & Myers, 2001; Walker, 2007). The National Human Genome Research Institute (2011) estimated that 30,000 individuals in the United States have HD and 75,000 people carry the gene that will eventually lead to the development of Huntington's disease. Each child of a carrier of the abnormal dominant gene that causes HD has a 50/50 chance of inheriting the disease. The onset of HD usually occurs between 30-50 years of age, with the duration of the disease lasting 15-20 years between onset and death (Family Caregiver Alliance, 2004; Genetics Home Reference, 2015). Despite the lack of a cure or efficacious treatment options for HD, there are medications that can help manage the associated motor, cognitive, and psychiatric symptoms (Aubeeluck, Dorey, Squitieri, Clay, Stupple, De Nicola, & Toumi, 2013). Although a basic understanding of HD among inmate populations by correctional professionals would be beneficial, it has not been studied among prison inmates in great detail.

Individuals with HD may be misdiagnosed with a variety of other conditions that present with similar symptoms. Some of the more common misdiagnoses may include Tourette's syndrome, schizophrenia, bipolar disorder, or Parkinson's disease (Halpin, 2011). Some individuals with HD are irrational, impulsive, and lack sound judgment for their own safety or the safety of others. These symptoms are attributed to deterioration in the brain as a result of HD.

This can lead to misunderstandings about the behavior and the uncontrollable physical manifestations associated with HD (Bourne, Clayton, Murch, & Grant, 2006). The following are the defining characteristics of HD (HDSA, 2012):

- Inherited, genetic disease
- Uncontrollable dance-like/writhing movements/tics (chorea)
- Progressive decline in functioning over a span of 15-20 years
- Neurological disease, which does not impact other organs
- A severe reduction in emotional expressiveness (flat affect)
- Long response times following verbal or physical prompts
- Terminal disease with no known cure

Common Issues Inmates With HD May Face While Incarcerated

Misunderstandings between correctional staff and inmates diagnosed with HD may create a crisis requiring hospitalization for appropriate medical treatment (HDSA, 2012). Common concerns associated with incarcerated individuals who have HD can include: personal safety, risk of suicide, victimization, and other behavioral and physical health issues. Response to these concerns may necessitate 24-hour supervision, specialized long-term care in a Huntington's disease-specific care setting, or involuntary psychiatric hospitalization.

Psychiatric or behavioral symptoms associated with HD may include one or more of the following: irritability, combativeness, sexual disinhibition, severe mood swings, obsessive-compulsive behavior, paranoia, delusions, or hallucinations. These symptoms can cause inmates with HD to be targeted in the general population of a correctional facility. As HD progresses, speech and communication skills decline until the individual is completely incoherent. Incarcerated people with HD may not be able to articulate their needs or their struggles (HDSA, 2012). The diminished level of functioning of inmates with HD places them at an increased risk for victimization. Individuals who have HD incur significant motor skill, cognitive processing, and personality loss throughout the progression of their terminal disease. Those affected by HD experience a reduction in the ability to function in order to work, drive, walk, maintain relationships, verbally communicate, or care for themselves. Eventually these individuals must rely on 24-hour care and supervision. The loss of independence in combination with the brain deterioration often results in depression. In turn, rates of suicide are far higher in this population than in the general public (Walker, 2007). Inmates with HD must receive routine mental health assessments and evaluation for suicidal ideations.

Malnutrition, dehydration, and fatigue are but a few of the health concerns resulting from uncontrollable choreic movements that characterize HD. The energy needed to focus and function is slowly sapped from the nonconsenting individual throughout the course of the disease. The involuntary movements cause individuals with HD to have very high metabolisms, which require many more calories than would be required by healthy individuals who have not been afflicted. As the disease progresses through early, middle, and advanced stages, feeding requirements, including restrictions, become more stringent in an attempt to ameliorate the effects of muscle degeneration. The loss of motor coordination and muscle control increases the risk of choking when eating and drinking. Supervision while eating is strongly encouraged. When coughing or choking episodes occur, accommodations should be made to the individual's diet. As HD enters the advanced stages, hiring an experienced professional caregiver, or using a feeding tube, can help reduce swallowing complications. Some individuals with HD must eat continuously to maintain their weight. Eating becomes increasingly difficult as the disease progresses. Reminders and prompts to drink fluids and to eat regularly are necessary to combat a lack of awareness about thirst and hunger. Managing HD is arduous for the individual, requiring increased amounts of sleep.

Intervention and Treatment Strategies for Correctional Professionals Overseeing Inmates With HD

- Allow 10 seconds between questions. Individuals who have HD may take longer to process and respond to a request.
- Use simple, clear, and unambiguous communication such as Yes/No questions. Avoid being vague or offering too many questions overall.
- Be patient. HD affects focus and concentration.
- Clearly and slowly explain the rules and expectations in a firm but respectful manner. The rapid presentation of information can overwhelm an individual suffering with HD.
- Involve family or friends. Since HD is a hereditary disease, the rest of the family is most likely aware of the best approaches to take in order to work with the individual.
- Do not try to reason with the individual if he or she is being unreasonable.
- Avoid confrontation. Uncontrollable movements often increase in severity when the individual becomes nervous or is anxious (Huntington's Disease Society of America (HDSA), 2012).

Conclusion

Although HD is fairly uncommon, it can pose unusual challenges to professionals within corrections. Fortunately, there is support available through the HDSA and medical professionals who specialize in treating HD. Patience and understanding, however, still go a long way when working with someone who is affected by HD. Working professionals within the correctional system can benefit greatly from further education and understanding about HD, specifically offender populations. Individuals and families are appreciative of any steps taken by correctional personnel to be better educated and more understanding about Huntington's disease. Knowledge about the manifestations of HD symptoms aids law enforcement and correctional professionals in differentiating malicious criminal behavior from involuntary HD symptomatic behavior. Ensuring that working professionals are knowledgeable about HD in correctional settings can improve the safety of both the inmates and facility staff. The Huntington's Disease Society of America is a comprehensive resource that offers webinars and informational packets for caregivers and working professionals alike who seek further education about HD.

Biographies

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Julia Besser, M.A., is a former humanitarian aid worker who is currently earning her Ph.D. in Counseling Psychology with a focus on corrections and policy making. She teaches undergraduate coursework in Psychopathology and Professional Development and serves as the President of the Graduate Student Council at Texas Woman's University. Her current research focuses on how the punitive criminal dilemma is affected by issues of competency and the complexity of detecting malingering. Her doctoral research is guided by her years of experience within the Texas state prison system, a forensic psychiatric hospital, and a substance use treatment center.

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[i] The Huntington's Disease Society of America is a nationwide agency that provides education, support, and research for HD patients and the community at large. Through its website, there are webinars and publications specifically for law enforcement and corrections professionals. There are also local chapters and social workers who are available to lend support, resources, and free in-service trainings for interested groups. Contact the HDSA for publications, information, and resources at 1-888-HDSA-506.