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Behavioral Changes in Huntington Disease

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Abstract

To mark the 30th anniversary of Cognitive and Behavioral Neurology (which was titled Neuropsychiatry, Neuropsychology, and Behavioral Neurology from 1988 to 2002), the editors have identified several articles previously published in the journal that might be termed “classics” in the field. These articles will be reprinted in the four issues to appear in 2018, each with a brief commentary by a contemporary working in that subject area. The aim of this project is to highlight the progress made in our subspecialty during the past few decades. Here, Jason Brandt introduces one of two articles reprinted in the current issue: Craufurd D, Thompson JC, Snowden JS. 2001. Behavioral changes in Huntington disease.

Although a psychiatric component of hereditary chorea was described in George Huntington’s original report on the disease in 1872, Huntington disease (HD) was largely overlooked by psychiatrists and psychologists for a century. In the 1980s, publications on the cognitive, emotional, and behavioral manifestations of HD began to emerge in earnest; this was made possible in the United States by the creation of “centers without walls” for HD in Boston and Baltimore (Brandt et al, 1988; Folstein et al, 1987; Schoenfeld et al, 1984; Starkstein et al, 1988). Studies on the behavior, personality, and psychopathology of individuals with manifest HD and those at risk proliferated (Caine and Shoulson, 1983; Folstein et al, 1983; Folstein and Folstein, 1983; Kessler, 1987; Lam et al, 1988; Stewart et al, 1987). With the discoveries of the chromosomal locus for HD in 1983 and the actual DNA mutation in 1993, and the launch of presymptomatic genetic testing programs shortly thereafter, understanding the emotional and behavioral phenotype of the illness took on a whole new dimension.

In Europe as well, interest in the neuropsychiatric aspects of HD blossomed (Jensen et al, 1993; Kuwert et al, 1989; Lundervold and Reinvang, 1991; Naarding et al, 2001; Scourfield et al, 1997; Tyler et al, 1983). In 2001, Craufurd and colleagues described their comprehensive analysis of behavioral changes in one of the largest series of HD outpatients studied until then. Their article was groundbreaking in several respects. First, the authors studied a relatively unselected group of patients attending a regional neurogenetics outpatient clinic. Thus, they avoided the biases inherent in studying inpatients or those seeking psychiatric services. Second, they eschewed the then-available psychiatric rating

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scales, clinical interviews, psychological tests, and *DSM*-based nosology. Rather, they developed, refined, and determined the psychometric properties of a new instrument, the Problem Behaviors Assessment for Huntington Disease (PBA-HD), to measure the specific symptoms prevalent in this population. Third, they interviewed caregivers as well as patients in order to get a fuller picture of patients' behavior. Fourth, they determined—as reasonably as possible within the context of a cross-sectional study—which behavioral abnormalities were associated with having the disease independent of duration (ie, those reflecting irritability and depression) and which were associated with chronicity (ie, those reflecting apathy). The former, it is argued, can be thought of as a manifestation of the genetic vulnerability of the illness and the latter as a marker of the progressive neurodegeneration.

This article by Craufurd and colleagues, published in the predecessor to *Cognitive and Behavioral Neurology*, set the stage for multiple subsequent studies of psychiatric features of people with manifest HD (Thompson et al, 2012; Thompson et al, 2002; van Duijn et al, 2014) and presymptomatic gene carriers (Duff et al, 2007; Julien et al, 2007; Poudel et al, 2015). Its status as a classic is well deserved.

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