

Huntington's Disease: Unique Problems or Similar Impact to Other Degenerative Neurological Disorders?

Steve Smith*

University of East Anglia, School of Health Sciences, UK

Huntington's disease (HD) is strikingly different from other progressive neurological disorders such as Alzheimer's disease (AD) in many ways. For example, HD is inherited as a dominant genetic trait, with children of affected parents being at 50% risk of developing symptoms in their own midlife [1] whereas the etiology of AD is multifactorial [2]. First manifestation of HD symptoms at the age of 60 years is regarded as 'late onset' [3] while AD symptoms occurring prior to 65 years of age identifies 'Early onset AD' [4]. The choreic movement disorder, psychiatric problems and cognitive decline associated with HD are among the distinguishing features referred to as the 'unique constellation of HD signs and symptoms' [5]. Dorey et al. suggest that these HD features cannot be captured when attempting to measure quality of life in people with HD using a generic quality of life instrument, and suggested this supported the need for a HD disease-specific tool.

One family caregiver summed up the impact of inheritance which is one factor that contributes to the uniqueness of HD: 'I met and soon married the love of my life. Her mother was ill and came to live with us. She became more and more dependant. Her behaviour became unpredictable and unmanageable as she lost inhibitions and the ability to think laterally or to appreciate another's point of view. Yet her memory for events, long and short term remained intact and she could hold intelligent conversation, making it hard to make allowances for her seemingly increasing self-centred outlook. She recognised us all by our faces and voices until she died some 8 years later. By then, my wife was starting to exhibit early HD signs and I learned that my children were each at 50% risk of inheriting the same condition in due course. I had no idea what I was committing to when I married, not that I would have decided differently.'

Despite these very specific pathophysiology and physical, psychological, cognitive and social aspects of HD I would argue that the impact of HD on family caregivers has much in common with that associated with AD and probably with other degenerative neurological disorders. Two studies I have carried out with colleagues support this position. The first study compared the psychometric performance of a disease-specific quality of life instrument developed to measure the impact of HD on family caregivers (the Huntington's disease quality of life battery for carers, -HDQoL_C) [6], with that of an instrument designed for use with caregivers for people with Alzheimer's disease (the Alzheimer's Carers' Quality of Life Inventory - ACQLI) [7], both used to measure quality of life in 61 HD carers.

The HDQoL_C incorporates three subscales (Practical aspects of caregiving (PC), Satisfaction with life (SL) and Feelings about living with Huntington's disease (FL), and a total quality of life score is achieved by combining the subscale scores. In contrast the ACQLI has only one scale and so presents a single quality of life score. As reported previously, Hagell and Smith [8] suitability of the HDQoL_C as an HD carer quality of life measure was not supported by the study. Two of the subscales did not exhibit fundamental psychometric properties and the third subscale did not outperform the ACQLI. The findings suggest that impact on quality of life of carers for people with degenerative neurological disorders is not specific to particular diagnoses.

A more recent study explored the views of people with HD, caregivers and health professionals involved in HD care provision, on what helps people live with HD [9]. In a cross-sectional mixed methods concept mapping study, 126 participants (39 people with HD, 48 caregivers and 39 health professionals including nurses, physicians, physiotherapists, occupational therapists, speech and language therapists and social workers) generated statements to answer the question: what helps people with HD live with their condition? They then prioritised the statements and grouped statements into clusters. Data analysis involved multidimensional scaling and hierarchical cluster analysis achieved using concept mapping software known as Ariadne* [10].

Qualitative explanations for the quantitative rankings of statements were provided by participants. The importance of having access to health professionals with HD specialist expertise was emphasised. The highest priority identified in this study was having access to expert assessment and treatment for co-morbid mental health problems. Integrated specialist multi-disciplinary HD expertise was called for and carers and people with HD expressed willingness if necessary to travel long distances to attend a centre that could provide this. Centres that provided an opportunity on one day to be seen by a physiotherapist, speech and language therapist, occupational therapist, social worker and neurologist and/or psychiatrist each with HD expertise, who would meet together and discuss the cases they had seen in that day and coordinate plans were especially appreciated as having the capacity to turn a chaotic experience of trying to cope with HD into a managed way of living with the condition.

An emphasis also was placed on care provided needing to be flexible and timely. Given the progressive nature of HD, delayed provision of equipment or home adaptations that are delivered after the period when it may have been useful was viewed as particularly frustrating as was having someone arrive to assist with washing and dressing the person with HD, who was not prepared to assist with a different task instead and free the family carer to give personal care.

Clearly there is agreement that unique features of Huntington's disease do pose problems that are particular to the condition and it was important to participants that this should be recognised and provided for. Yet seamless integrated interdisciplinary team working, sharing case information, responsive and flexible, timely service provision was demonstrated to reduce the impact for people with wide ranging long term neurological conditions in an evaluation of the impact of the National Service Framework [11].

*Corresponding author: Steve Smith, University of East Anglia, School of Health Sciences, UK, Tel: +44 7743 960129; E-mail: Steve.Smith@uea.ac.uk

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The priority given to the need to address mental health problems even in a condition such as HD that poses very specific problems such as chorea and dominant inheritance, should not be surprising given that more broadly, neuropsychiatric symptoms including anxiety and depression in neurological disorders such as dementia are associated with increased caregiver impact, greater functional impairment, greater likelihood of institutional care and a poorer quality of life [12]. However the high impact of psychiatric co-morbidity is common to many long term conditions in terms of increased service need and negative impact on QOL [13] and this is particularly the case for people with neurological disorders such as Alzheimer's disease and Parkinson's disease [14]. Concerns to address mental health for people with dementia and their carers feature prominently in a list of top ten priorities for dementia research resulting from the James Lind Alliance dementia priority setting partnerships [15]. The third and fourth priorities identified refer to the importance of finding ways to manage mental health and challenging behaviour, respectively [15]. Furthermore a priority setting partnership aimed at establishing the top ten research priorities for the management of Parkinson's disease rated the question 'what approaches are helpful for reducing stress and anxiety in people with Parkinson's?' as the second priority area [16].

In summary, long-term neurological disorders bring with them challenges that are unique to each condition, but much of the impact on quality of life, and the service provision needed to make living with these conditions more manageable from the perspectives of patients, caregivers and health professionals involved in their care is shared across diagnoses.

For example, HD onset is typically earlier in life than AD. HD is inherited as a single gene dominant trait whereas the family implications of developing AD are more complex and pose a comparatively much lower risk for offspring of those affected. Cell death in AD is distributed much more widely in the brain from the onset of symptoms compared to HD, and consequently cognitive decline in early HD is more limited to specific aspects such as flexibility of thought and control of impulses than is the case in AD where effects are more global. Yet caregivers for people with AD and those for people with HD are faced with similar problems to deal with such as impaired sleep, co-morbid mental health problems such as depression and anxiety and increasing dependence on them for assistance with activities of living.

Working together to call for greater flexibility, integrated working and placing a high priority on assessment and treatment of comorbid mental health conditions is in the interests of all.

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