

Cognitive impairment and behavioural difficulties in patients with Huntington's disease

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Summary

Huntington's disease (HD) is a degenerative, neurological disorder that has physical, psychological and cognitive effects. A review of the nursing literature on HD suggests that the main focus has been on explaining and managing the physical and psychological symptoms that arise. Cognitive symptoms appear to be only briefly discussed in most articles, perhaps because they are often the most difficult to explain and understand. This article examines the link between cognition and behaviour in patients with HD and suggests strategies to manage some of these behaviours.

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HUNTINGTON'S DISEASE (HD) is a degenerative, neurological condition which affects one person in 100,000 of the population (BUPA Health Information Team 2004). It has physical, psychological and cognitive effects, and because of the degenerative nature of the condition and the complex symptoms that arise, it can be emotionally and physically draining for patients, carers and staff.

The nursing literature has discussed the full range of symptoms associated with this disease. A recent article by Kent (2004), for example, provides an overview of the history of HD, the major characteristics of the disease, treatment and its effects on the patient's family members. Simpson (2004) describes the main clinical features of HD, for example, motor problems, speech difficulties and cognitive problems, and discusses ways of assessing symptoms, addressing financial issues and drug therapy.

Articles such as these are invaluable in raising the awareness of HD among professionals and carers, primarily because they highlight the main symptoms of the illness and examine ways in which they can be managed to provide a better quality of life for the patient and his or her family. This article does not attempt to replicate this work. Instead, the main behavioural problems associated with cognitive impairment are outlined and strategies that formal and informal carers can adopt to make the care and treatment of patients with HD more effective are discussed.

Hamilton *et al* (2003) explore the interaction between motor, cognitive and behavioural impairment and emphasise that behavioural dysfunction in patients with HD can only be explained fully when this complex interplay of symptoms is considered. Because of the rarity of the condition there is a lack of specialist services to cater for the needs of patients with HD, which can result in patients being inappropriately placed, for example, on adult acute psychiatric wards or in non-specialised residential and nursing homes. Staff working in these areas frequently have little or no experience in caring for patients with HD, or their families.

The authors and the other members of the Leicestershire Huntington's Disease Service have found that nursing staff and carers of patients with HD residing at home, in NHS inpatient areas or in residential and nursing homes, have some knowledge and understanding of the physical effects and psychological symptoms of the condition, for example, impaired swallowing, movement disorders, anger and depression. They also have some knowledge and understanding of how these symptoms arise from the disease. Although the physical and psychological symptoms are invariably debilitating for the patient, cause concern for staff and carers and are not always easy to manage, they can usually be explained and understood. However, when faced with cognitive difficulties staff and carers may find it difficult to understand how impaired thinking can lead to behavioural disturbances.

In the author's experience, problems associated with cognition are less well understood, for example, fixed thinking, slowed responses or an inability to sequence activities. Spouses and friends of those with HD often report that the patient has undergone a complete 'personality change', which can be very distressing. Nursing staff who understand how cognition and behaviour are intrinsically related can help family members and carers to gain more insight into the effects of cognitive impairment on patient behaviour, thereby enhancing quality of life and promoting a more therapeutic relationship.

Cognition

A good understanding of cognitive functioning and techniques to manage related behaviour are essential in providing effective care for patients in many different specialties. Cognition is the ability to take in information via the senses, process the details, commit this information to memory and recall it as necessary.

In the author's experience, patients who are low in mood can typically find it more difficult to concentrate and assimilate information and are therefore less likely to successfully commit items to memory. Patients with auditory hallucinations may find it difficult to respond in a meaningful way if they have not heard a question clearly. Those who have sustained a brain injury may find it difficult to retrieve memories. The elements that make up cognition, including attention, concentration, spatial awareness and memory, have a direct impact on mental and physical health and wellbeing, and need to be considered during patient assessment and nursing intervention.

Cognitive disorder in patients with HD is associated with a progressive impairment in attention, executive function and immediate memory (Ho *et al* 2003). There are many assessment tools available for cognitive impairment, such as the Addenbrooke's Cognitive Examination (Mathuranath *et al* 2000). Such tools are useful in assessing the nature and level of cognitive impairment, and provide a benchmark to work from.

There are two main reasons why cognitive disorder leads to behavioural problems in patients with HD. First, executive functions can be affected and second, cognitive flexibility – the ability to concentrate on more than one task and to adapt to changing circumstances – can become impaired. This helps to explain why patients with HD lose their initiative and appear apathetic. Patients with HD may easily become overloaded with tasks or experience difficulty in adapting to change and respond in an outburst of temper or in a way that might be considered unreasonable (Quarrell 2002).

In the author's experience family members seem to have more difficulty in coping with cognitive and behavioural deterioration than motor problems. This is often the main reason carers can no longer manage to meet the patient's needs. This, in turn, may mean that an inpatient admission or a nursing home placement is necessary. Cognitive and behavioural difficulties may manifest as aggression (verbal and physical), apathy, a lack of spontaneity, doing 'bizarre' things – such as going out for long walks for hours at a time – and altered sex drive, which may become heightened or lessened (Quarrell 2002).

Hahn-Barma *et al* (1998) state that the rate of cognitive decline in HD patients can often be anticipated. Using different cognitive tests with 99 candidates at a French hospital, they studied early cognitive deficits in asymptomatic gene carriers. They discovered that the presence of a significantly higher number of CAG repeats (code for one of the building blocks of the Huntington protein) in the cognitively impaired gene carriers could be associated with an earlier mean age at onset in this subgroup. It would appear that the lower the age of onset the more rapid the cognitive decline.

One of the major problems with cognitive impairment in HD is the inability of patients to understand how their symptoms are affecting their lives and the lives of those around them. Patients with HD often have a tendency to overestimate their abilities and be unable to predict the consequences of their actions. Anosognosia – the inability to accept or recognise a physical defect or a lack of self-awareness – is a common problem and patients with HD often answer 'I'm fine' to any questions

they are asked (Simpson 2004). Therefore, caring for patients with HD presents a number of challenges because the patient's thinking can be slowed, initiating action is more difficult, learning new things is not as easy as it was and judgement is impaired (Pollard *et al* 2000). Patients with HD often do not realise this and are unaware of the impact that these deficits have on them and those around them.

Cognitive impairment and behavioural difficulties

In patients with HD, cognitive impairment typically gives rise to behavioural problems in the following ways:

Fixed thinking Patients with HD can often become fixed on one idea and behaviourally, this can appear as set patterns and routines, and an apparent unwillingness to adapt to new situations or altered circumstances. The cognitive changes that occur in HD can impair the ability to think and adapt easily to new situations and patients generally feel most comfortable and confident in familiar situations involving fixed routines (Huntington's Disease Association 2005).

For example, a patient may become fixed on the idea that he or she is overweight and will want to exercise vigorously every day to lose weight. This can result in some patients walking miles each day, for hours at a time. Because the patient's thinking has been so rigid he or she may not have considered the damage he or she may be causing, for example, sore feet, physical exhaustion and hypothermia.

Management When caring for patients with fixed thinking, it is important to try to channel the fixed thought into a behaviour that is manageable and appropriate for the individual patient. For example, encourage the patient who is determined to exercise to use a local gym or an exercise bike at home. If a patient with HD is insistent on walking, then try to help them to learn a route that is safe and to which they can become accustomed.

Slowed thinking and responses Patients with HD typically have a problem in accessing memory stores in the brain. Thought processes are slowed, and additional time is required to communicate and achieve routine activities (Paulsen and Stoll-Fernandes 1995). It is for this reason that the patient may often take a long time to answer the telephone, for example. The person realises that something is making a noise, but can take some time to recognise that the noise being made is the telephone ringing and that it needs to be answered.

Slowed thinking and responding is often evident when talking directly to a person with HD. Sometimes the person can take a few minutes to respond to a simple question such as 'how are you?' This may be interpreted as the person being

rude and choosing not to respond. It is only when the carer understands that this is a cognitive problem of slowed response that this behaviour becomes more acceptable.

Management Use clear, concise sentences when speaking to a person with HD and try to use closed questions that require a specific response. Allow the person the necessary time to respond to any questions asked. Try to ask one question or use one sentence at a time – this can help the patient to assimilate the information being communicated and enable him or her to give a more appropriate response.

Memory problems Memory involves three stages: taking in information, storing it and retrieving it. Ring and Serra-Mestres (2002) state it is the third stage – retrieving information – that usually becomes a problem for patients with HD. They also explain that difficulty in retrieving memory applies equally to information stored a long time ago as that stored more recently. Verbal memory, however, can often be retained and patients find it easier to recognise words.

Management The use of prompts and cues, for example, lists, diaries, calendars, signs around the house or near the patient's bed if he or she is in a nursing or residential home, can all assist in helping a person to retrieve information from memory. Pollard *et al* (2000) explain that, in HD, the person's ability to actively think about and to link information is significantly impaired. However, the provision of memory aids can help to do the linking for them. When learning new information, it can often help to break tasks down into component parts and apply a step-by-step approach to the task. This is called 'process training' and involves working through the process of doing a task, for example, shopping, before actually doing it.

Retrieving memory is directly linked to attention and concentration. The patient with HD will find it easier to respond to what is being said or to concentrate on a task if distractions are minimised, for example, do not have the television or radio on when the patient is trying to read a newspaper.

Difficulty sequencing activities Patients with HD often find it difficult to work through a process in a logical sequence and have difficulties starting and completing activities. For example, a person may get up in the morning, decide an hour later to have a shower and then spend more than an hour in it, because they cannot think to finish that activity and start another one. This can be frustrating for the patient and also for carers who may interpret these actions as apathy and awkwardness.

A study by Hamilton *et al* (2003) found that apathy was the most significant behavioural abnormality that patients with HD develop. The authors state that frontostriatal neuropathology

(damage in the striatum of the basal ganglia – the part of the brain most directly affected in HD) appears to lead to a primary reduction in drive and motivation which presents as behavioural apathy. Therefore, when carers understand that patients with HD can find it difficult to initiate and to finish activities, they realise that apathetic behaviour is not purposeful and has a biological cause.

Management It is important to maintain a regular routine in assisting patients with HD to meet their activities of daily living. Nurses should write specific individualised information in the care plans, ensuring that such routines are achievable and adhered to. The more that behaviour can be routinised and systematic, the easier the person with HD will find it to initiate activities and to work through them sequentially. It is helpful to use prompts and supervision as the symptoms of HD progress and the nurse or carer often needs to prompt the person when to begin and end an activity. Patients may also need to be supervised to help them carry out the activity in a logical order, for example, making sure that a T-shirt is put on before a sweater when getting dressed.

Loss of visuospatial awareness Patients with HD often experience perceptual impairment where they can see objects or people as being closer to them or further away than they actually are. This can be interpreted as aggressive behaviour, for example, when a patient with HD is communicating with another person he or she

may get too close to the other person, or the patient may appear to 'slam' a cup on the table. In both cases, the lack of visuospatial awareness has probably resulted in patients not knowing how close they are when speaking to another, or where the surface of the table actually starts. This is not to say that aggression is not a problem in patients with HD, but when the carer realises that visuospatial difficulties can exist, true aggression can then be differentiated from behaviour that merely appears as such.

Management When speaking to a person with HD, staff and carers may need to take a step back if the person is getting too close in proximity and invading their personal space, because the patient will not be aware of this. Floor surfaces need to be flat and even and preferably made of the same material throughout the house or home to assist safe mobilisation, that is, so that the patient recognises where to take the next step.

Conclusion

Understanding the link between cognitive difficulties and behavioural problems that occur in patients with HD is important because this may assist staff and carers to manage symptoms more effectively. Nurses need to be aware of the adverse effects of cognitive impairment and how this affects the patient's ability to understand what is being communicated to him or her. Confusion may result in maladaptive behavioural responses that can make care planning ineffective. Conversely, if cognitive impairment is understood and appreciated, practical strategies can be established which may result in more positive outcomes for patients **NS**

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