

Exploring supportive care for individuals affected by Huntington disease and their family caregivers in a community setting

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Aim. The aim of this research was to identify the needs and coping strategies of individuals with Huntington disease and their family members/carers and to ascertain the extent to which they were supported by the type of specialised service provided in this unit.

Background. The duration of the illness may be up to 30 years and there are far reaching implications for the affected person, family members and carers. Services for families need to provide expert psychological and practical support, whilst remaining planned and flexible. The aim of this research was to identify needs and coping strategies of individuals with Huntington disease and their carers and ascertain the extent to which they were supported by a specialised community service.

Methods. Five focus groups and three individual interviews were used to gather data from affected individuals, family, carers and staff of the unit ($n = 33$). A semi-structured discussion guide was used in every case. Data were analysed using an inductive coding technique.

Results. Three main themes emerged; (i) Transitions and the journey, (ii) Challenges in finding and providing support and (iii) The role of the unit in providing supportive care. While the service was seen as supportive, the need for flexibility was emphasised to facilitate the transition from unaffected to affected status. Affected individuals felt it was important to be known as the person they had been, despite the changes due to the disease.

Conclusions. Provision of service in a dedicated unit is an appropriate way to support affected patients and their carers. Enhancing understanding of the disease

and allowing the individual to make choices can assist coping with this challenging disease.

Relevance to clinical practice. It is helpful to allow patients control over timing and frequency of contact with professionals. Improving understanding of the condition in the public and health professionals may enable patients to access a greater range of social activities.

Key words: caregivers, chronic illness, community, focus groups, Huntington disease, supportive care

Introduction

Huntington disease (HD), formerly called Huntington's chorea, is a chronic neurodegenerative condition caused by a genetic mutation in the huntingtin gene, situated on chromosome 4 (MacDonald *et al.* 2003). The condition is inherited in an autosomal dominant pattern. Any individual who has inherited the full gene mutation will develop the condition during their lifetime, unless death occurs because of some other cause before symptoms present (Myers 2004). The age of onset varies from childhood to old age, but is typically between 30–50 years (Myers 2004). The incidence is 1–10 000–20 000 in the general population (Anderson & Marshal 2005), although there may be many affected individuals in one family, because of the inherited nature of the condition.

The signs and symptoms of HD may be classified into three main groups. These are: disorders of movement including uncontrollable jerky movements of the face and limbs (chorea), emotional problems and cognitive deficits that increase with the progression of the disease (Wood *et al.* 2002). Over 50% of people with HD will have behavioural changes; these may be treatable (Skirton & Glendinning 1997). Loss of memory and labile mood cause major problems (Bonelli & Hofman 2004), while some individuals demonstrate irritability, aggressiveness, anxiety disorders, apathy or obsessional thoughts and behaviours (Ranen 2002). Medication can play an important part in treatment of these symptoms, as can structuring the day, acceptance of the patient's mood and initiation of activities (Ranen 2002).

Since the gene mutation that causes HD was identified in 1993 (The Huntington's Disease Collaborative Research Group 1993), it has been possible for those at risk of the condition to have predictive testing to detect whether or not they have inherited the mutation. However, because of the serious nature of the disease, the lack of preventive treatment and the potential adverse impact of knowing one's genetic status prior to onset of symptoms, strict protocols including pre- and posttest counselling are used in conjunction with these tests.

Background

Huntington disease is a long illness, possibly lasting up to 30 years (Kent 2004). Individuals often become symptomatic when relatively young, when they may be parenting young children. The disease has a powerful impact on families, not only because of the symptoms present in the affected person, but also because of the implications for other family members and their descendants (Semple 1995). Carers report feeling anxious, depressed, helpless, lonely, fearful, tired and vulnerable (Semple 1995). Services for people with HD (and their family carers) need to provide expert psychological and practical support, yet be carefully planned and flexible (Dawson *et al.* 2004).

Perhaps because HD is a relatively rare condition, there is a paucity of research into non-clinical management of HD and a subsequent need for studies into the functional, psychological and social consequences of the condition (Kaptein *et al.* 2006). Skirton and Glendinning (1997) conducted a study of the needs of patients and carers and found that all 35 patients studied had significant unmet needs in terms of physical and psychological care, mental health support and social support, while carers felt unsupported by the health and social care systems. Few studies investigate specific services for patients affected with HD, but a small number describe the care provided in specialised inpatient units for patients with advanced HD in the United Kingdom (Crouch 2003) and the USA (Rajecki 1991, Hegland 1994). Smith (1998) claims the effects of HD on patients can be alleviated through appropriate nursing care, believing that the quality of life of patients with neurological conditions is improved in specialised units, although this claim is supported by anecdotal evidence only (Smith 2002). Moss (1995) describes a small quantitative study in which patients in a specialised HD unit showed less deterioration over time than a comparable group in a general setting.

A robust study by Dawson *et al.* (2004) in Australia used a qualitative approach to investigate the need for supportive care for persons affected by Huntington disease and their carers, highlighting the unsuitability of using facilities

for the aged for younger people affected by long-term conditions. The main themes to emerge from their study were adjusting to the impact of the illness, surviving the search for information, gathering practical support, bolstering the spirit, choreographing individual care and fearing for the future. While the study identified the need for specialised services for this group of patients and their carers, it did not examine the extent to which services were meeting the needs of those studied.

Authors focussing on the quality of life of carers (Helder *et al.* 2002) or patients (Helder *et al.* 2001) conclude that HD impacts greatly on the person's physical wellbeing, but affects their psychosocial wellbeing even more severely. Recent research suggests that psychological factors, such as social support and patients' strategies for coping with their disease, play a more critical role in the wellbeing of chronically ill patients than medical factors, such as disease severity (Kaptein *et al.* 2006). Government initiatives in the United Kingdom have directed efforts to manage chronic diseases more effectively in the community. Current government policy involves helping patients maintain independence as long as possible and, in partnership with health professionals, enabling patients to take more control over managing their illnesses (Department of Health 2001, 2002, 2005).

To try to address these needs, a multi-disciplinary day service was developed at the specialised community unit involved in this study. Initially a residential care centre, the service at the unit was reconfigured 2.5 years before the study, to provide community-based services for individuals affected with HD. Some patients attend this unit on a day care basis and activities are aimed at helping individuals to help maintain emotional and psychological wellbeing, mobility and cognitive functioning. Others use the community outreach service, which aims to provide support to anyone affected by HD in their own homes. A specialist medical outpatient clinic operates through the centre and speech and language therapy, physiotherapy, occupational therapy and dietetics are all offered to the patients as part of the package of care. As part of the mental health service, patients are managed under the Care Programme Approach (CPA), designed to ensure that complex care needs are met and managed. A multidisciplinary approach is used and partnerships developed across statutory, private and voluntary services. The aim of this research was to identify the needs and coping strategies of individuals with HD and their family members/carers and to ascertain the extent to which they were supported by the type of specialised service provided in this unit.

Objectives

The objectives of the research were:

- To identify the support needs and coping strategies of people with HD and of those who care for a family member with HD.
- To ascertain the extent to which they were supported by the type of specialised service provided in the unit.
- To identify unmet needs of those who use the unit.

Methods

An exploratory research design using qualitative methods was used to obtain in-depth data from a range of stakeholders, including patients, staff and carers. Due to the cognitive deficit in some of the affected individuals, verbal discussion was more likely to be effective than written methods in obtaining their views. Walters (1995) has argued that it is essential that shared understanding of the cultural and linguistic concepts develop through the process of this type of research. In this study, the focus groups were facilitated by a researcher with clinical and research experience of the topic to maximise that understanding. The research team consisted of the manager of the unit and two academics from the regional University. The two academics (who were not involved in operation of the unit) obtained consent from participants and conducted all focus groups and interviews. Ethical approval was obtained for this study from South West Devon Research Ethics Committee.

Setting

This study was conducted in a specialised community-based unit (described above) under the management of a Primary Care Trust in South West England.

Participant inclusion and exclusion criteria

All adults affected with HD who attended for day care at the unit were invited to participate. While some of these had a cognitive deficit because of the condition, involving people who were using the service was important and care was taken to explain the study to all potential participants. For each affected individual, one person providing care for the person at home was invited to participate. Of the 20 day service users who were invited, 13 accepted, while nine out of 16 carers were involved in the study. In addition, nine individuals using the outreach service (being visited at home) were invited and three accepted. All unit staff who had direct patient contact were asked to participate; eight out of 15 staff

Table 1 Demographic profile of participants

Category	Number of participants in focus groups	Number of participants in individual interviews	Marital status	Age (years)
Women using day services	7		3 married 1 never married 1 widowed 1 divorced	47–64
Men using day services	5	1	4 married 2 divorced	41–60
Persons using community services	2 (both male)	1 (male)		
Carers	8 (3 male, 5 female)	1 (female)		24–65
Unit staff	8 (7 female, 1 male) 1 registered nurse 1 physiotherapist 1 physiotherapy support worker 1 health care assistant 1 chaplain 1 occupational therapy technical instructor 1 occupational therapy assistant 1 occupational therapist			

accepted the invitation. Thirty-three people participated in group or individual interviews; their characteristics are shown in Table 1.

Six carers were spouses or partners of an affected person, while one was the mother of an affected woman and one was paid as a carer. A number of healthcare disciplines were represented in the staff group (Table 1).

Data collection

Research participants were invited to take part in one of five focus groups or individual interviews. Focus groups were used because it was intended that there would be some debate about the discussion topics and the interviewers wanted to explore experiences, concerns, opinions and wishes (Webb & Kevorn 2001). Four discussion groups were already established in the unit for affected women, affected men, carers and staff; these were used as the basis for four focus groups. It was felt that using pre-existing groups would lessen anxiety and enable the participants with cognitive deficits or mental health problems to talk freely. Moreover, this would constitute less change for people with HD, who tend to find change difficult because of the mental inflexibility associated with the disease. The fifth focus group consisted of those people with HD receiving outreach services. As these patients do not usually attend the unit they were invited to meet in a neutral venue. The focus groups were facilitated by an experienced interviewer with an understanding of the difficulties of interviewing people with cognitive impairment and an

Table 2 Focus group topic guide – general themes

History of having/caring for someone with HD
Response to diagnosis
Factors that inhibit or support coping
Specific issues arising from living with HD or caring for someone with HD
Impact of day service or community service on coping
Potential improvements to the day or community service
Issues that have an impact on feeling in control of own life
Perceived needs in future

observer took field notes. A topic guide was used (Table 2), but adjustments were made to questions to make them relevant to the group. Individual interviews were offered to those who did not wish to or were unable to attend a group. Both individual interviews and focus groups were facilitated by the same interviewers and these were audio-taped and transcribed in full.

Data analysis

Data analysis was undertaken using an inductive coding technique (Miles and Huberman (1994). Initially, transcripts were read several times to ensure familiarity with the content. Each section of the transcript was then annotated by two researchers (one academic and one service manager) using basic codes. As the interaction between focus group members is important to the analysis, areas of agreement and disagreement were noted (Stevens 1996). The codes were

organised into categories and then several broad themes. To ensure trustworthiness and accuracy of the analysis and to reduce bias, codes and themes were discussed by all three researchers until consensus was reached to ensure consistency.

Ethical issues

Interviewers were aware that affected participants and their carers formed a vulnerable group and the potential for distress was recognised. However, it was crucial to give individuals an opportunity to express their views as the service exists for them (Allan 2001). Researchers understood that those groups consisting of patients might be challenging to facilitate because people with HD have cognitive problems which make it difficult for them to concentrate or follow the discussion and make them potentially slow to respond. In fact, a level of inflexibility in thinking was noted which made it difficult for some individuals to follow the change in direction of questions during the interview.

All participants were deemed to have capacity to make an informed choice about whether to participate and each was able to sign his/her name. In the information sheet and discussion prior to the interviews it was explained that comments would not be attributable to individual participants and staff of the unit would not be aware of who made the comments. However, participants were informed that limited disclosure might occur if a member of staff or patients were thought to be at risk in some way. Nursing staff known to the group members were available to offer support should any individual become upset by the discussion, although this was not required.

Findings

Themes

Three main themes emerged from the data; (i) Transitions and the journey, (ii) Challenges in finding and providing support and (iii) The role of the unit in providing supportive care. Quotations are labelled according to the category of respondent: OG = outreach group, MA = men's group, WA = women's group, CA = carer's group, ST = staff group.

Transitions and the journey

The affected individuals and carers who participated in the study described a series of decisional tasks or adjustments that were part of the journey associated with the condition. Whilst there was some variation between individuals, there

was also much common ground. The steps in the disease journey included:

- Finding out about the disease.
- Making decisions about whether to have the genetic test.
- Making decisions regarding relationships and starting a family.
- Deciding how and when to tell children about the genetic implications.
- Watching a family member or members change and eventually die with the disease.

For many in this study who were affected with Huntington disease or were family carers, being given the diagnosis and realising the implications was described as devastating:

My Mum has Huntington's...the first time I went to [the unit] and saw some of the worst patients I was upset. It was very frightening, very frightening indeed. (OG)

Some individuals however, had found that living with uncertainty about their future had been very difficult and whilst upsetting, the news of the diagnosis provided relief from this:

I think part of it was the relief of knowing – even though it was bad news, just to actually know and not have that uncertainty. (CA)

Following the diagnosis of one member, other individuals in a family with HD may then live with the knowledge that they might have the genetic mutation. While some people had chosen to ignore the possibility of developing the disease, others had wanted to obtain information about the future by opting to have a genetic test. However those who obtained certainty about their genetic status through testing were faced with new uncertainties. Those in the outreach group (who had positive mutation test results) spoke of their vigilance in self monitoring to detect whether the symptoms had started to appear:

It's difficult (to know) whether it's Huntington's like, or a nervousness or a slight jerk....I don't know. (OG)

Affected individuals voiced the need for information about the condition to prepare themselves, but they felt frustrated at the lack of knowledge about HD held by health professionals generally. One talked about the lack of information in local surgeries:

Not even a basic leaflet to understand the basics. (MA)

It is not surprising that having a risk or history of HD in the family has a significant influence on the lives of family members. Some related traumatic experiences, such as seeing a family member become aggressive and unmanageable or cognitively impaired. In one case, the lack of ability

by family members to care for the affected person was troubling:

He got to the stage where he was uncontrollable at home. Nan couldn't look after him, he had to go into a home. (OG)

Understanding and empathy by family members was seen as helpful for the person experiencing symptoms:

Understand and have empathy for the person who's struggling through something that's no fault of their own. (OG)

Staff and carers described how supporting an affected person to undertake activities outside the home could be difficult, because of depression or lack of motivation in that individual. They spoke of the encouragement they had to give the affected person to ensure that he or she continued to engage in such activities:

It's a struggle for him to leave the four walls of his home and the television...but once...he's out there, he's thoroughly enjoying it and he comes back and I know that the depression has lifted, you know. (CA)

However, reluctance on the part of the affected person to engage in social activities was often compounded by the response from members of the public that stigmatised the person with HD:

They want to go into a shop...and the garden centre where there's lots of expensive ornaments and they [patients] just go from one side to another, that takes a lot of energy...to be with them...as we know they're often looked at as drunks or drug takers. (ST)

Challenges in finding and providing support

Every patient had some mobility issues and a general loss of ability to do things he/she could previously do. Both staff and some patients identified a conflict around ensuring that the service provides individuals with freedom of choice, whilst remaining proactive enough to help patients to overcome the symptoms of HD. These symptoms (such as depression or paranoia) may in themselves prevent people from accessing the services offered:

The approach...has got to be very, very gradual. You can't say, 'Come to the Day Unit'. They've got to do it bit by bit...eventually they do but it's a long time before they decide to come in. (ST)

One man described his extreme nervousness when first attending the unit for day care, but could not articulate what it was about attending that had made him nervous. He did state, however, that just experiencing being in the unit enabled him to eventually overcome those feelings and enjoy attending.

For the affected individuals who wished to use health and social care services, continuity and access to care were important. Similarly, having a flexible service where patients could maintain some control over their own access was identified as essential:

Just to maintain some sort of general continuity...to maintain some connection there, not that every week I need help, but if I was then I would speak to somebody [at the unit] about that. (OG)

However, there seemed to be a potential for coercion, with some affected individuals reporting that they only used a day service because other people wanted them to:

I've been told I've got to. The consultant said to me I've got to come to [the day unit] a couple of times a week. (MG)

An extremely important concept to emerge was the wish by individuals with HD to be known for their personal qualities. They strongly expressed how important it was to them to be in contact with people who had known them before they became ill, so that these friends, relatives or professionals knew the real person and were more likely to be understanding when behaviour changes occurred:

People who look after you in later life gonna know you in earlier life as young and healthy and whatever, you know, confident and wicked; all those things. (OG)

In turn, carers did seem to respond to the real person and demonstrated a true commitment to the person with HD, carrying on in spite of the difficult behaviour exhibited and trying to maintain their relative's quality of life. There was an acceptance by carers that the affected person was not really at fault:

There's no cure for Huntington's. I want to make it as good as possible for her, simple as that...she's my wife. She'd do for me what I'm doing for her. (CA)

Despite their willingness to care, many caregivers felt unsupported by the health and social care systems. They often talked of their experiences in terms of a struggle, as they fought to access help, for example in the realm of respite care to allow them to have a break:

It's a huge battle and...the whole scenario takes it out of you physically and mentally. (CA)

The role of the unit in providing supportive care

Both day services and community outreach services were seen to provide very useful sources of support, enabling the affected persons to engage with others in a safe environment.

However, some affected people were reported to be uncomfortable with the idea of contact with other affected people:

My main issues are getting [husband] to come to the Unit to join in socially because he won't. He doesn't want to be associated with other HD people. (CA)

For other service users, the opportunity to mix with others who were similarly affected provided an opportunity for positive social interaction that was appreciated. The day service was described as having a happy environment:

I'm very pleased to be able to come up and meet these fellow patients,...it's nice, they're a friendly lot, so I've got no qualms in talking to them. (MA)

Service users who were recipients of the community outreach service appreciated having someone visit them at home. In addition, believing that the staff understood the individual needs of the person reduced their feelings of vulnerability.

They spoke positively of the control they exercised over when and where they were seen by the health professional:

Well, at the moment I'm fairly all right...[community outreach nurse] comes to see me if I need him to or I can come here to [the unit] you know. (MA)

A proactive service that included outreach to them personally provided a '*personal doorway*' to support (OG) and continuity of staff enabled a trust relationship to be built. The individualised nature of the service was emphasised:

Some people want information and some people don't...some people want to talk about Huntington's and other people don't. You know, it's a very intuitive, personal thing. (OG)

Both carers and affected individuals felt that the unit provided a service which was supportive to carers, promoted the wellbeing of the carer and also provided respite. Although all participants were invited to suggest any potential improvements to the service, most felt that their needs were being met, although three carers felt there was still a need for overnight respite care. Feeling reassured about the standard of care was mentioned as important in enabling the carer to entrust the affected person into the care of the unit staff. This was expressed as knowing that the staff were '*deeply concerned*' (WA) about the service users:

The wife relaxes when she knows I'm coming here because she knows of the standard. (MA)

We feel that when our loved ones are here [the unit] that gives us respite. They can call it what they want, the staff all say it's not respite, but it is. (CA)

Discussion

The data derived from this study confirm that both affected persons and carers are living with change and loss over many years, as reported by Skirton (1998). Some participants felt that the positive diagnosis of HD provided a relief from uncertainty. The need to obtain certainty is a psychological trait (Webster & Kruglanski 1994) that motivates the individual to seek information, but this trait varies from person to person and may explain why in this study some people at risk had sought testing and information at an early stage, while others had not.

While a diagnosis may bring certainty, participants expressed the view that adjusting to a diagnosis of HD was not immediate and that facing patients in the advanced stages of the illness when visiting the unit was difficult during this period of adjustment. This suggests that the community service which offers support in a setting external to the unit is helpful, allowing patients more control over management of their condition, consistent with current health care policy (Department of Health 2001, 2002, 2005). Dawson *et al.* (2004) also claim that flexibility is a key feature of a quality service for patients with this disease, so offering patients access to services in locations and at times that they feel are appropriate, particularly as they adjust to the diagnosis, may be useful.

Decisions around genetic testing were discussed in most interviews. Studies indicate that only a minority of those at risk of HD seek testing (Meiser & Dunn 2000, Taylor 2004). Furthermore, Taylor's study reveals the depth of thought given to testing by individuals at risk, with consideration given to the impact of the result on self and others and awareness of the moral right 'not to know'. Better understanding of these decision-making processes could enable staff to develop greater understanding of the ways in which some people manage their risk status.

A strong theme that emerged from groups that comprised affected persons and the staff was the need to regard the affected person as he or she had been before the illness. Brock (1995) believes that facilitating a person with a family history of a genetic condition to tell his/her story as a narrative has therapeutic value. For this reason, it may be helpful to spend time in encouraging the affected person to relate aspects of his or her life history.

While activities to stimulate cognitive function have been shown to be useful to patients affected with HD (Kaptein *et al.* 2006), in practice access to activities outside of the home or unit may be limited. Staff and carers were particularly aware of the way in which the affected person was

viewed by others and commented that unsteady gait, slurred speech and swallowing difficulties invite comparisons with drunkenness or drug abuse, making it difficult for people to use external services. Carers found the behaviour of the affected person difficult to manage; this is entirely consistent with the findings of the study by Skirton and Glendinning (1997).

The need by carers for respite from the constant work of caring physically, mentally and emotionally for the person with HD is cited as a major theme by Dawson *et al.* (2004). While the service attended by the affected participants in this study was not designed as a respite service, both patients and carers saw this aspect of providing a specialised service for individuals with HD as valuable because it provided carers with time that could be devoted to their own needs. Whilst there are other similarities with the findings of Dawson *et al.* (2004), including the need to adjust to the illness and a period of gathering practical support, the theme of 'fearing for the future' was not apparent in the present study. This may have been because the data were collected mainly in focus group settings, or it may be that those in this study viewed the future more positively. Further work would be required to explore this issue.

Strengths and limitations

One strength of this study was the inclusive approach which involved a range of stakeholders. However, our experience of focus groups for the affected persons was that they were more like group interviews, as interaction between members was limited and much prompting from the facilitator was necessary to encourage members to contribute. Given the cognitive deficits and resulting difficulties with social skills that often accompany the disease, this is not surprising. In any circumstances group members can feel reticent about talking about themselves (Allan 2001). One affected person was interviewed individually because he was unwell on the day the focus group was held. However, the data obtained through that interview was limited and the researchers believe it is unlikely that other affected persons would have been more forthcoming in individual interviews, where focus on them alone may have been very uncomfortable. The researchers therefore feel that the focus group approach was probably the most acceptable in enabling the affected persons to make a contribution to this research.

Implications for nursing practice and education

It is clear that the support provided by the dedicated unit for those affected with HD is valued, but clients also wish

to maintain some independence, particularly in the area of access to the service in the initial years of the illness. For this reason, flexibility is required in the way the service is offered. Allowing patients control over the timing and frequency of contact, especially in the early stages of the disease, is essential. However, patients may also require encouragement to participate because of loss of executive function and confidence and the subsequent difficulty in adjusting to new situations that may accompany the disease.

Affected individuals were clear that it is important to them to be known as the person they had been before the changes wrought in them by the disease process. This suggests either a need for early contact with staff, or facilitation of later work that focuses on the patient's interests, achievements, goals and personal history.

It was emphasised that better understanding of HD by those in the community would benefit patients and carers and make activities outside the home or unit more feasible. Training sessions for professionals and more interaction between unit staff and general practitioners would help to ensure healthcare professionals have a better understanding of the impact of the disease. However, HD is a rare disease and therefore focusing these efforts on those professionals who actually have current contact with a patient may be most effective.

Further research

This study identified that the patient undergoes a period of transition from unaffected to affected status. Further work is needed to investigate the ways in which patients and their families could be best supported during this important transitional process and how meaningful activity benefits individuals.

Conclusion

Provision of services in this dedicated unit is highly valued by patients, families and carers. People with HD have to face a number of challenges at various points in time and this unit does help patients and their families to address these challenges. However, more work needs to be done to enhance understanding of the disease in the community to enable patients to undertake a broader range of activities. Making an effort to get to know the affected individual personally and allowing people to make choices in the way their HD is managed can assist those who are affected and their carers to cope more effectively with the disease.

Contributions

Study design: BS, PG, HS; data collection: PG, HS; analysis: BS, PG, HS; manuscript preparation: HS, BS, PG.

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