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Huntington disease: families' experiences of healthcare services

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Abstract

Aim—This paper is a report of a study of the perceptions of family caregivers regarding the availability and adequacy of health and social care services for their family member with Huntington disease, and to compare findings from these reports in United Kingdom and United States of America samples.

Background—Huntington disease is an inherited neurodegenerative condition. Family members often take responsibility for care of relatives with long-term conditions. Studies have demonstrated there are both positive and negative outcomes for carers.

Methods—During 2006 and 2007, respondents from the United Kingdom ($n = 108$) and the United States ($n = 119$) who were caring for a relative affected with Huntington disease completed the Community Health Care Services Scale to identify areas of concern and the extent to which specific issues bothered carers. Data were analysed using statistical tests including chi-square, t -tests and factor analysis. Results were compared between carers in the two cohorts.

Results—Three main factors were derived: 'community resources', 'individualized care' and 'knowledge of Huntington disease'. Carers had concerns about the knowledge of healthcare professionals providing care and thought that there were insufficient services to support them and the affected person. There were different challenges for carers when the affected person had a long-term neurodegenerative condition because these carers were also likely to have responsibilities for earning and caring for children.

Conclusion—Comprehensive facilities and resources are needed to support families affected by long-term complex conditions. Healthcare professionals need to be aware of the health needs of carers as well as those of the affected person.

Keywords

carers; families; healthcare services; Huntington disease; long-term condition; nursing

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CONFLICT OF INTEREST

No conflict of interest has been declared by the authors.

AUTHOR CONTRIBUTIONS

HS, JW, JB and JP were responsible for the study conception and design; made critical revisions to the paper for important intellectual content. HS and JW performed the data collection. HS, JW and JB performed the data analysis and were responsible for the drafting of the manuscript. JB provided statistical expertise, administrative, technical or material support. JW and JP obtained funding.

INTRODUCTION

Huntington disease (HD) is an inherited, long-term neurodegenerative condition. An individual affected with HD experiences motor symptoms, such as ataxic gait and involuntary choreic movements, loss of cognitive function, depression and dementia (Quarrell 2008). The onset of symptoms can occur at any age, but most commonly individuals who have inherited the gene mutation that causes the condition will start to have symptoms in the fourth or fifth decade of life (Bates *et al.* 2002). The course of the disease runs for around 15–20 years. The affected person therefore requires health and social care for an extended period of time, with much of this care being coordinated by the family, whether it is provided in the home or in a care facility in the community.

BACKGROUND

Relatives of people with HD may not be direct caregivers, but they have concerns about many aspects of HD, including their role as support persons during predictive testing (Williams *et al.* 2000), the implications of that testing on their own risk for the disease (Tibben 2007), and dealing with the day-to-day issues they encounter (Aubeeluck & Buchanan 2007, Williams *et al.* 2007, 2009a, 2009b). As early as 1993, Kessler was writing about spousal carers as ‘the forgotten person in the Huntington’s disease family’ (Kessler 1993, p. 145), asserting that carers’ needs were often overlooked by healthcare professionals, and that there was a strong need for further research into the experiences and needs of carers. Despite this suggestion, little research focussed on the carers of people with HD has been undertaken. However, Skirton and Glendinning (1997) included carers in their study of the needs of people with HD and found that carers were dissatisfied with the amount of respite care and the support they received from healthcare professionals, which resulted in a series of crises of care. This inevitably placed additional pressures on the family system and relationships.

In the United Kingdom (UK), there have been several initiatives to promote the co-ordination and quality of care for people with long-term neurological conditions. In 2005, the UK Department of Health issued a National Service Framework for people with long-term neurological conditions (Department of Health 2005). Quality Requirement 10 of the National Service Framework is focussed on supporting the family and carers. This states that carers have a right to obtain the support and services they require for the person they care for and for themselves as carers. The potential for carers to suffer from stress and diminished quality of life is acknowledged in this document. Furthermore, some of the strategies recommended in the guidance are that carers are: (i) offered assessments of their own health, (ii) involved in planning care for the affected person, (iii) given appropriate information about the condition, the manifestations of the condition and potential treatment/management strategies and (iv) offered the opportunity to take regular breaks from caring. A series of outcomes published in 2008 in connection with the management of long-term conditions (Department of Health 2008) includes the opportunity for people to have choices about their care (outcome 3) and that the services provided are flexible and responsive to the needs of the individual (outcome 4).

In the United States of America (USA), healthcare providers can access HD clinical information through the Huntington Disease Society of America’s (HDSA) toll-free information hotline and through their local healthcare providers. The HDSA produces and distributes publications and informational material on HD to assist physicians, patients and family members. This organization also supports 21 Centres of Excellence in which staff conduct research, clinical, and education activities. These centres provide regional care, but their services may not be accessible to all people with HD and their families. Healthcare

services for people with HD also reflect local community resources, and a recent survey of HD programmes showed that there are few large cohorts of people with HD in residential care (Lechich *et al.* 2008). Payment for healthcare comes from a variety of sources. For the entire population, employer insurance covers approximately 50%, government programmes cover approximately 30% and individually-purchased insurance another 5%, leaving approximately 15% without healthcare coverage (Nuwer *et al.* 2008). People with HD or at risk for HD cite fear of genetic discrimination from employers or insurers (Oster *et al.* 2008, Penziner *et al.* 2008). The recent signing of the Genetic Information Nondiscrimination Act creates expectations that those providing health insurance will not discriminate based on genetic information (Hudson *et al.* 2008). However, this law will not prohibit insurance providers from rejecting applications or charging higher rates for people who have a medical diagnosis (Erwin 2008).

Huntington disease is a condition for which much of the day-to-day care is provided by family until this no longer becomes feasible (Skirton & Glendinning 1997). Although HD symptoms and symptomatic treatment may not vary across geographical regions, the healthcare context may influence concerns and strategies available to families in managing day to day care of their relatives with HD.

THE STUDY

Aim

The aim of the study was to examine the perceptions of family caregivers regarding the availability and adequacy of health and social care services for their family member with HD, and to compare these reports in UK and US samples.

Design

The data reported here are derived from a larger study, the purpose of which was to identify concerns of family members of people with HD and strategies used to manage these concerns (Williams *et al.* 2001). A mixed method design was used in the larger study. A questionnaire was constructed from themes derived from focus groups in the USA and in Canada with adult relatives of people with HD (Williams *et al.* 2007, 2009b). In this paper we report a further phase of the study, in which the questionnaire was administered to family caregivers from the UK and USA.

Participants

Participants in the UK were eligible for inclusion in the study if they were currently caring for one or more people with HD, were aged 18 years, lived in the UK, were able to give consent to be involved in the study, and were able to complete the questionnaire in English. There were no restrictions on the number of hours per week in which the participant was involved in caring, and they were able to take part in the survey regardless of whether the affected person lived with the carer, in another private home or in a residential or nursing home. Participants were recruited via advertisements in the website of the Huntington's Disease Association (HDA) of the UK. In addition, information about the study was circulated by Regional Care Advisors working across England and Scotland with families affected by HD and by members of the local branches of the HDA support groups.

In the USA, inclusion criteria were broader than those for the UK survey; specifically people were eligible if they (i) were aged 18, (ii) identified themselves as a family member or 'significant other' of a person who had inherited the gene mutation causing HD, (iii) were able to speak English, (iv) were able to respond to a mailed survey. US adult families of people with HD were recruited from the Iowa HD Registry, the HD Lighthouse (an Internet

site), the HDSA annual meeting, and the University of Connecticut Health Center. To ensure comparability of the samples, in this report, only the US participants who identified themselves as carers of a relative with HD are included in the analysis.

Data collection

Measures—The Community Health Care Services Scale is used to measure family members' perceptions of the availability and adequacy of community resources to support health care of the person with HD. The scale has been validated in the USA with HD experts and a subset of families who participated in focus groups: this process will be reported elsewhere. It includes 14 closed ended items and two open-ended questions that were included to enable respondents to register their strongest concerns and record concerns not addressed in the questionnaire. Each closed-ended item had a range of response options: 0 (Strongly disagree), 1 (disagree), 2 (agree) and 3 (strongly agree). For each item, respondents indicated whether they disagreed or agreed that the statement about healthcare service was relevant to their situation as the carer of a person with HD. We refer to this later in the paper as 'agreement'. In a second column, they were asked to indicate the extent to which that issue bothered them on a scale of: 0 (not at all), 1 (a little), 2 (moderately), 3 (very much) and 4 (extremely). We refer to this as 'degree bothered'.

To ensure applicability to the UK sample, the US version of the questionnaire was reviewed by group of UK adult family caregivers through a cognitive interviewing process (Drennan 2003). This involved a series of five interviews with current caregivers. The participants were asked how they interpreted the meaning of each question and, where there was a discrepancy between the meaning intended by the researcher and the meaning as interpreted by the participants, alternative acceptable forms of wording were discussed. There were a small number of minor changes. The wording of one demographic question was altered to reflect the different educational systems in the UK. The term 'physical therapies' was changed to physiotherapy and 'HD Center staff' was changed to 'Special HD Clinic staff'. The intent or basic structure of all the items remained consistent with the US version of the questionnaire.

Collection procedure—These data were collected in the USA from March to November 2006 and in the UK from October 2006 to September 2007. A conscious effort was made to enrol as many people in the study as possible. Potential participants were asked to contact the researchers by email, phone or letter to request a copy of the questionnaire, which was sent to them by post with a reply-paid envelope. The completed questionnaires were returned to the researcher by post. It was expected that the sample sizes would give a power of at least 0.80 for the primary comparisons. This would have been attained had there not been missing responses. However, the minimum power for all statistical significance tests was 0.75, with most being above 0.80. The contribution of free text comments was optional, and the amount of text written by respondents varied from a few lines to over four pages.

Ethical considerations

Approval for this study was obtained in the UK and in the US from the appropriate ethics committees. Personal details were not requested on the questionnaires to enable anonymity to be preserved. Participants involved in the cognitive interviewing were asked to comment on the wording and relevance of the questions but were not asked to provide information about their personal circumstances.

Data analysis

For the demographic variables with categorical responses, the distributions for UK and US respondents were compared using chi-square tests of proportions. For demographic items

with continuous responses, means for the UK and US groups were compared using independent *t* tests. On each of the items for the concerns and bothered response sets, the UK and US groups were compared using chi-square tests of proportions. There were variable numbers of respondents for the comparisons because of non-response on some items, particularly if participants thought that they did not apply.

Results are presented by each factor and for score for the concerns items and for the intensity of being bothered items; this was done by comparing means for the UK and US samples that were based on summing the individual items on the factors and for total score. Independent *t* tests were used for this comparison, with a level of statistical significance of 0.05. The homogeneity of variance assumption was checked and met in every comparison except one, and in that case the unequal variance *t* estimate was used. Responses on the concerns and bothered items were collapsed into fewer categories and the proportions were compared using chi-square tests of homogeneity of proportions with the level of statistical significance set at 0.05.

The free text data were analysed using the approach described by Miles and Huberman (1994). The data were read and coded according to topic. Matrices of codes were constructed and these were grouped under categories and themes. To ensure rigour, a second researcher reviewed the analysis and a consensus was reached concerning the coding and assignment of codes to categories and themes.

RESULTS

The US recruitment yielded 241 returned questionnaires and UK recruitment yielded 120. After extracting questionnaires completed by those identifying themselves as primary carers, there were 119 US and 108 UK participants. The data from these questionnaires are reported here.

Socio-demographic profile of respondents

A socio-demographic profile of the respondents is presented in Table 1. There were no differences between the two national groups in terms of age of respondent, sex or ethnicity. The numbers married or living as married were also comparable. The mean age of US respondents was 51.15 years (*SD* = 12.265) while for UK respondents it was 52.63 (*SD* = 13.359).

In both samples, the vast majority of respondents described themselves as White. In the US sample 3.4% (*n* = 4) were native Americans, 2.6% (*n* = 3) Hispanic and 1.7% (*n* = 2) of other ethnic origin. Of the UK respondents, 1.9% (*n* = 2) described themselves as of Asian origin.

However, there were statistically significant differences in the sizes of communities in which respondents lived ($\chi^2 = 26.404$, d.f. = 3, $P < 0.001$), with a greater proportion of US respondents living in rural areas, although in both samples over 40% of respondents lived either in small towns or rural communities. The educational level of US respondents was also statistically significantly higher than that of UK respondents ($\chi^2 = 23.964$, d.f. = 4, $P < 0.001$), with 52.3% (*n* = 62) of US respondents stating they had received some college education or higher, compared with 22.5% (*n* = 23) of UK respondents.

There were striking differences in self-reports of respondent health status ($\chi^2 = 17.656$, d.f. = 2, $P < 0.001$). Of UK respondents, 38% reported their health as fair, and 13.9% as poor. Only 22.9% US respondents felt that their health was fair and 3.4% that it was poor.

Data were collected on the relationship of the carer to the affected person. In the US, 65.5% ($n = 76$) were spouses or partners; this was highly comparable to 64.5% ($n = 69$) in the UK. Other carers were parents, siblings, children or other relatives. There were statistically significant differences in the number of carers who were parents of the affected person ($\chi^2 = 11.801$, d.f. = 3, $P = 0.019$). In the US 19% ($n = 22$) carers were parents, compared with 8.4% ($n = 9$) in the UK. In the UK, the mean age of the person being cared for was older ($t = 2.464$, d.f. = 225, $P = 0.015$).

Survey findings

Confidence intervals for the internal consistency reliability coefficients were found using ScoreRelCI (Barnette 2005). The agreement scale had an internal consistency reliability of 0.89 [95% confidence interval (CI): 0.84, 0.93] and the degree bothered scale had an internal consistency of 0.93 (95% CI: 0.91, 0.96). Principal components with Varimax rotation factor analysis of the disagree-agree scale for the US sample revealed three factors that included 12 of the 14 items and accounted for 62.8% of the variance. Three factors were identified: a factor labelled 'community resources' with six items accounting for 36.7% of the variance with loadings ranging from 0.46 to 0.89 (mean = 0.72), a factor labelled 'Individualized Care' with three items with loadings from 0.58 to 0.83 (mean = 0.75) accounting for 14.0% of the variance, and a third factor labelled 'knowledge of HD' with three items with loadings from 0.51 to 0.85 (mean = 0.73) that accounted for 12.1% of the variance. Subscale internal consistency reliabilities for the three subscales were: Community Resources with six items 0.91 (95% CI: 0.88, 0.93), Individualized Care with three items 0.79 (95% CI: 0.72, 0.85) and HD Knowledge with three items 0.63 (95% CI: 0.54, 0.71). The items included in each factor are presented in Tables 2 and 3.

Table 2 presents the comparisons on the concerns items. It is clear that the most agreement regarding concern is on the Knowledge of HD items (2.65/3 for UK and 2.63/3 for US), with both groups giving ratings in the strongly agree range. There were 'agree' ratings by both groups on the Community Resources items (2.33/3 for UK and 2.16/3 for US) and relatively neutral ratings on the Individualized Care items (1.52/3 for UK and 1.31/3 for US). There were no statistically significant differences in factor scores or total scores between UK and US on the concerns items, based on the t tests of the summed scores on each scale.

Table 3 presents comparisons between the UK and US groups by item on the concerns scale. Responses were collapsed into two categories. Strongly disagree and Disagree were merged into a 'disagree' response and Agree and Strongly Agree were merged into an 'agree' response. The proportions in these categories were compared using chi-square tests of proportion differences. There were statistically significant differences on two of the concerns items, both related to finances. On 'difficult to get affordable quality care' item, the percentage agreement for the UK group was 65.7%, statistically significantly higher than for the US group at 42.2%, $P < 0.001$, with a *post hoc* power of 0.80. On the item 'difficult to obtain financial benefits from disability services' item, the percentage agreement for the UK group was 42.9%, statistically significantly higher than the US group at 11.8%, $P < 0.001$, with a *post hoc* power of 0.99.

Table 4 presents the comparisons on the extent bothered items. It is clear that respondents were bothered most on the Knowledge of HD items (3.10/4 for UK and 2.86/4 for US), with both groups providing ratings in the bothered 'very much' range. There were differing degree of bothered ratings between the groups on the Community Resources items (2.90/4 for UK and 2.38/4 for US) and 'moderately' bothered ratings on the Individualized Care items (2.02/4 for UK and 1.53/4 for US). There were statistically significant differences on two of the factor scores and the total score between UK and US on the bothered items, based on the summed items scores. Mean bothered ratings on the Community Resources scale (UK

= 2.90/4 and US = 2.38/4, $P = 0.006$ with a *post hoc* power of 0.80), Individualized Care scale (UK = 2.02/4 and US = 1.53/4, $P = 0.013$ with a *post hoc* power of 0.76), and total scale were statistically significantly higher for the UK group compared with the US group (UK = 2.67/4 and US = 2.22/4, $P = 0.001$ and a *post hoc* power of 0.93).

Table 5 presents comparisons by item on the degree bothered items. Responses were collapsed into three categories: Not at all, Little or Moderately, and Very Much or Extremely. Based on the chi-square test of proportion homogeneity, there were statistically significant differences on four items, and on each item of these the degree bothered was statistically significantly higher for the UK group. These items were: 'difficult to get affordable quality care' (UK = 79.7% and US = 57.1% very much or extremely bothered with $P = 0.005$, with a *post hoc* power of 0.80), 'difficult to find appropriate activities' (UK = 76.5% and US = 57.1% very much or extremely bothered with $P = 0.014$ and a *post hoc* power of 0.75), 'most HCP (healthcare professionals) do not have enough HD education' (UK = 82.6% and US = 64.3% very much or extremely bothered with $P = 0.006$ and a *post hoc* power of 0.83), and 'Difficult to obtain financial benefits from disability services' (UK = 59.3% and US = 34.2% very much or extremely bothered, with $P = 0.006$ and a *post hoc* power of 0.88).

Factor 1: community resources

Factor 1 was the factor on to which the highest number of items loaded. As can be seen from Table 2, there was agreement across the cohorts on items related to insufficient number of resources available to support care, difficulty finding trained staff, facilities close to home and facilities that would accept the affected person. The textual data collected supported the quantitative findings:

(There is a) lack of understanding/training in care homes so they accept someone with HD – only to realise they can't manage. Yet another move for the HD sufferer. (UK P16)

What gives me most stress about coping with my husband's HD is the continual struggle to get the care workers to look after him properly and timely...I am repeatedly training new people and reminding regular people what they should be doing and trying to be polite and pleasant about it! (UK P22)

Due to my son's early onset of HD, it is especially difficult to find services, which would help him, that he would be willing to try. Adult day care and Assisted living facilities are full of old people. (US P103)

We would like to move him closer, but because of anger outbursts no one wants to take him. (US P120)

In respect of care/residential homes – I found it irritating when seeking a suitable plan for respite care that manager of homes did not seem to understand that people with HD do have special care...they were then not inclined to either seek training or listen to advice from family member who is normally caring full time. (UK P48)

Factor 2: individualized care

There was agreement by caregivers in both countries about the three items that loaded on to the second factor; these were about healthcare professionals not taking account of the personal preferences of the affected person, not addressing the affected person's concomitant health problems and not giving the health care required:

Nursing home was completely insensitive to her inabilities. They forgot to feed her, family stayed with her from 8 am to 8 pm to make sure she was fed, got medications. (US P113)

Politics between social services and health service may mean her being taken away from home and placed in residential care. (UK P115)

Doctors are not up-to-date on HD and do overlook other medical problems. (US P179)

Factor 3: knowledge of Huntington disease

The three items that loaded on to factor 3 were related to the knowledge of HD amongst the general public, knowledge amongst healthcare professionals, and perceived lack of media attention focussed on the disease. There were no differences in mean scores for the US and UK cohorts for these items regarding the relevance of the items or the extent of concern about them:

My main concern is the lack of HD ignorance/experience displayed by health professionals. i.e. doctors, dentist – some social workers. In some cases I have known more about HD than some doctors. (UK P62)

I just wish the public understood more about HD. Continue to bring awareness to this horrible disease. Continue to offer as much resources as possible on HD. (US P505)

HD is forgotten about. Other illnesses promoted by ‘celebrities get far more attention and funding. (UK P103)

DISCUSSION

Study limitations

The respondents for this study were drawn from two countries, and therefore the data reflect the views of carers from a wide geographical area and living in varied communities. However, the method of recruitment relied upon access to those who attended specialist clinics, were in contact with the support group or accessed an HD specialist website. Because of this recruitment strategy, it may be that the views of those who are less aware of the support groups or who do not have access to specialist care may not be represented, and this needs to be addressed in further studies.

Discussion of findings

Our findings indicate that family carers of individuals affected by HD are considerably concerned about accessing health services for the affected person. They believed that healthcare professionals do not have the requisite knowledge about the condition and this may well have an impact on the difficulties they experience in finding affordable and appropriate care. In our study the mean age of the person being cared for was 51 years, well below the age at which gerontological services for people with long-term conditions are normally provided. Rurality has been shown to be a factor in access to healthcare services in many countries (Iredale *et al.* 2005, Smith *et al.* 2008), and the level of concern about appropriate services may also reflect the fact that over 40% of respondents in both samples lived outside a major metropolitan area, meaning that specialized services could be harder to find.

Despite the existence of the National Health Service in the UK, carers there were statistically significantly more concerned about finding affordable care that met the needs of the affected person, as well as about obtaining disability benefits. Initiatives have been undertaken in the UK (Department of Health 2005) to improve the quality of care of people

with long-term neurological conditions and their carers; however, there appear to be important shortfalls in the care provided. Concerns were also expressed by those in the US, and assisting people and families remains a priority in the mission of the HDSA (2009). However, the quality of links between specialty services in HD Centers and local healthcare providers may vary across communities.

The relationship between the carer and the person with HD may be an issue in how challenging the carer finds the process of dealing with healthcare providers and care systems. Pearlin and Aneshensel (1994) describe caregiving as an unexpected 'career', and it may be that spouses are more likely to accept such a career as part of their commitment to their partner. A case study of a daughter of a woman with dementia has highlighted the daughter's reluctance to accept the role change required to care for her mother (Goldsteen *et al.* 2007) because it required a shift in her own identity as a daughter. In our study, around a third of those who responded were not spouses or partners, and this may help explain the high levels of concern indicated in the results. Where the carer is a parent, they may be worried about their own ability to continue to care for the lifetime of their affected son or daughter. Depp *et al.* (2005) examined the influence of kinship status for daughter versus wife caregivers of people with dementia and found that daughters reported higher self-efficacy when indicating level of confidence in ability to manage a caregiving issue; it may be that parental carers and carers who are sons or daughters of the affected person may have very different concerns. An additional factor that should be borne in mind when considering the needs of carers is the risk of developing HD in biological relatives, which may increase the difficulties in the carer's abilities to carry out these responsibilities. From the present study, it is not possible to determine how much a person's own at-risk status influences the intensity of response to questions about the individual for whom they provide care, and this merits further investigation.

CONCLUSION

Carers in from both countries in this study perceived a lack of knowledge in healthcare professionals. While it is not feasible for all healthcare professionals to have competence in caring for patients with a wide range of conditions, it is essential that those caring for a person with a long-term neurodegenerative condition such as HD develop the requisite skills and acquire the appropriate knowledge to develop competence. There is an urgent need for formal routes of preparation for HD practitioners that would encompass standards for education, specific training and professional development of those who are responsible for providing quality day-to-day care for people with HD. The model of practitioner accreditation provided by the National Council of Certified Dementia Practitioners (2009) could be an appropriate resource for addressing this issue. Furthermore, better policies are needed to facilitate effective consultation with experts when required.

It is clear that where a person is affected with a long-term condition in young adulthood or middle age, the pressure on families will be increased because of the additional responsibilities inherent in those life stages, such as the need to work for an income and the raising of children. These factors in themselves require a different approach to carer support and assuring the fit of community-based health and social services than is required for conditions of old age. Healthcare professionals need to be aware of the potential stressors on carers in younger age groups.

The profile of carers in the two countries was different in terms of relationship to the person with HD, carer health and age of the affected person. Whilst this may have been an artefact of our study, it could also indicate that different approaches may be necessary according to the carer profile in each country, and this is an area where further research is needed.

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What is already known about this topic

People with long-term neurodegenerative conditions require complex care from a range of professionals from many disciplines.

Family carers provide much of the care required by people affected by long-term neurodegenerative conditions.

Both positive and negative aspects of taking responsibility for care have been reported.

What this paper adds

Family carers of individuals affected by Huntington disease were concerned about accessing appropriate and affordable health care for the affected person.

Family carers were concerned that healthcare professionals do not have the requisite knowledge about the condition to offer appropriate care.

There were different challenges for carers when the affected person had a long-term neurodegenerative condition with onset in early or middle adulthood, because the carer was also likely to have responsibilities for earning and caring for children.

Implications for practice and/or policy

Healthcare professionals who provide services for people affected with Huntington disease should have the specific preparation to ensure they have the knowledge and skills required to provide care.

Further facilities and resources are needed to support families affected by long-term complex conditions.

Healthcare professionals need to be aware of the health needs of carers as well as those of the affected person.

Table 1

Demographic profile of respondents

	United States of America, <i>n</i> (%)	United Kingdom, <i>n</i> (%)	Total, <i>n</i> (%)
Sex	119	108	227
Male	38 (31.9)	39 (36.1)	77 (33.9)
Female	81 (68.1)	69 (63.9)	150 (66.1)
Marital status	117	108	225
Single	7 (6.0)	7 (6.5)	14 (6.2)
Married or living as married	92 (78.6)	93 (86.1)	185 (82.2)
Separated	0 (0)	2 (1.9)	2 (0.9)
Divorced	9 (7.7)	2 (1.9)	11 (4.9)
Widowed	9 (7.7)	4 (3.7)	13 (5.8)
Community size	114	107	221
Metropolitan	43 (37.7)	19 (17.8)	62 (28.1)
Urban	18 (15.8)	43 (40.2)	61 (27.6)
Small town or sub-urban	31 (27.2)	37 (34.6)	68 (30.8)
Rural	22 (19.3)	8 (7.5)	30 (13.6)
Self-reported health of respondent	118	108	226
Good	87 (73.7)	52 (48.1)	139 (61.5)
Fair	27 (22.9)	41 (38.0)	68 (30.1)
Poor	4 (3.4)	15 (13.9)	19 (8.45)
Mean age of person with HD	119	103	227
SD	13.32 (49.34)	12.66 (53.66)	13.15 (51.40)

Table 2
Factor and total score comparisons on disagree-agree items – extent to which the respondent was concerned

Factor	Items	Score range	United Kingdom			United States of America			<i>t</i> Value	<i>P</i> value
			<i>n</i>	Mean (SD)	Mean on scale	<i>n</i>	Mean (SD)	Mean on scale		
1. Scale: 0 = strongly disagree, 1 = disagree, 2 = agree, 3 = strongly agree.										
Community resources	6	0–18	92	13.98 (4.48)	2.33	102	12.96 (4.42)	2.16	1.60	0.112
Individualized care	3	0–9	103	4.57 (2.48)	1.52	108	3.94 (2.33)	13.1	18.9	0.061
Knowledge of Huntington disease	3	0–9	108	7.94 (1.19)	2.65	119	7.89 (1.30)	2.63	0.32	0.749
Total	14	0–42	108	30.26 (7.09)	21.6	119	28.38 (7.37)	2.07	1.95	0.052

Table 3

Carers' concerns about healthcare services: item comparisons

	United Kingdom		United States of America		P value
	n	%A	n	%A	
Factor 1: community resources					
There are not enough community resources to help	80	38.8	82	50.0	0.150
Difficult to find facility with trained staff	51	37.3	57	40.4	0.742
Difficult to get affordable quality care	70	34.3	83	57.8	<0.001*
Difficult to find appropriate activities	65	35.4	57	52.6	0.055
Difficult to find facility close to home	49	40.8	52	53.8	0.190
Difficult to find facility that will take him	50	48.0	55	61.8	0.155
Factor 2: individualized care					
HCP do not ask about his treatment preferences	75	82.7	73	86.3	0.542
HCP do not take care of his other health problems	71	83.1	73	89.0	0.303
He is not getting health care he needs	82	65.9	76	77.6	0.101
Factor 3: knowledge of HD					
The general public needs to know more	108	31.5	119	25.2	0.294
There is not enough media attention	105	28.6	117	29.9	0.826
Most HCP do not have enough HD education	101	40.9	110	42.2	0.432
Other items not included in factors 1–3					
Difficult to obtain financial benefits from disability services	84	57.1	68	88.2	<0.004*
Person has experienced discrimination because of HD	56	71.4	67	71.6	0.979

1. Scoring: 0 = strongly disagree, 1 = disagree, 2 = agree, 3 = strongly agree. Collapsed for data analysis into D = 0 or 1 and A = 2 or 3. HCP, healthcare professional; HD, Huntington disease.

2. * $P < 0.05$.

Table 4

Factor and total score comparisons on degree bothered items

Factor	Items	Score range	United Kingdom			United States of America			<i>t</i> -Value	<i>P</i> value
			<i>n</i>	Mean (SD)	Mean on scale	<i>n</i>	Mean (SD)	Mean on scale		
Community resources	6	0–30	89	17.39 (7.26)	2.90	99	142.5 (7.99)	23.8	2.80	0.006*
Individualized care	3	0–15	100	60.7 (4.43)	2.02	106	45.9 (3.93)	1.53	2.52 [†]	0.013*
Knowledge of HD	3	0–15	106	9.30 (2.77)	3.10	117	8.57 (2.85)	2.86	1.92	0.056
Total	14	0–70	106	37.34 (13.67)	2.67	117	31.05 (13.90)	2.22	34.0	0.001*

1. Scale: 0 = not at all, 1 = little, 2 = moderately, 3 = very much, 4 = extremely.

2. * $P < 0.05$.3. [†]Based on unequal variance *t* estimation as homogeneity of variance assumption not met.

Table 5
The extent to which the issue bothered the carer, item comparisons

	United Kingdom				United States of America				P value
	n	%N	%LM	%ME	n	%N	%LM	%ME	
Factor 1: community resources									
There are not enough community resources to help	81	4.9	25.9	69.1	8.2	12.2	25.6	62.2	0.247
Difficult to find facility with trained staff	57	15.8	19.3	6.49	60	16.7	25.0	58.3	0.724
Difficult to get affordable quality care	74	8.1	12.2	79.7	84	9.5	33.3	57.1	0.005*
Difficult to find appropriate activities	68	7.4	16.2	76.5	61	19.7	27.9	52.5	0.014*
Difficult to find facility close to home	55	14.5	18.2	67.3	59	18.6	32.2	49.2	0.129
Difficult to find facility that will take him	57	17.5	17.5	64.9	59	22.0	28.8	49.2	0.208
Factor 2: individualized care									
HCP do not ask about his treatment preferences	83	37.3	27.7	34.9	88	43.2	29.5	27.3	0.543
HCP do not take care of his other health problems	77	35.1	23.4	41.6	88	43.2	28.4	28.4	0.208
He is not getting health care he needs	90	22.2	2.89	4.89	83	32.5	32.5	3.49	0.145
Factor 3: knowledge of HD									
The general public needs to know more	105	2.9	28.6	68.6	116	1.7	34.5	63.8	0.574
There is not enough media attention	104	3.8	25.0	71.2	115	3.5	25.2	71.3	0.989
Most HCP do not have enough HD education	102	3.9	13.7	82.4	112	3.6	32.1	64.3	0.006*
Other Items not included in factors 1–3									
Difficult to obtain financial benefits from disability services	86	18.6	22.1	59.3	76	32.9	32.9	34.2	0.006*
Person has experienced discrimination because of HD	59	32.2	25.4	42.4	73	35.6	23.3	41.1	0.911

1. Scoring: 0 = not at all, 1 = a little, 2 = moderately, 3 = very much, 4 = extremely. Collapsed for data analysis into N, not at all; LM, little/moderately; ME, very much/extremely. HCP, healthcare professional; HD, Huntington disease.

2. * $P < 0.05$.