

Section 2: Empirical Paper

**The views of adults with Huntington's disease on assisted dying: a psychological
exploration**

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Abstract

In several countries the legalisation of assisted dying is frequently debated. However, little empirical research or weight is given to the views of people with life-limiting conditions, who might be more likely to desire hastened death. The purpose of this study was to explore the views of people with Huntington's disease (HD) on assisted dying. Using thematic analysis methodology seven semi-structured interviews were conducted with people with HD or who were gene-positive to understand their views on the issue. Four themes were extracted: 1) Autonomy and kindness in assisted dying; 2) HD threatens identity but is part of life; 3) Dilemmas in decision-making on assisted dying: "There are no winners"; and 4) Absence of explicit discussion on dying and HD: "Elephants in the room". The theoretical, clinical and research implications are discussed.

Keywords: Huntington's disease, neurodegenerative disease, assisted dying, euthanasia, assisted suicide

The views of adults with Huntington's disease on assisted dying: a psychological exploration

With the increasing age of populations in developed countries, chronic illness has become the leading cause of death (WHO; World Health Organization, 2004). Palliative care is increasingly promoted for a range of chronic conditions such as chronic obstructive pulmonary disease (Murray, Pinnock, & Sheikh, 2006), human immunodeficiency virus (Harding et al., 2005) and motor neurone disease (MND) (Borasio & Voltz, 1997). However, cancer patients continue to receive the majority of palliative care provision (Hanks, Cherny, Christakis, & Kaasa, 2011) despite cancer only accounting for an estimated 25% of deaths (Murray, Mason, & Donaldson, 2010). Moreover, there are significant challenges in implementing this approach for conditions such as dementia where disease trajectory is less predictable (Murray et al., 2010). Indeed, Fitzsimons et al. (2007) reported that the clinical needs of people dying with chronic illness are largely not met.

One such condition not usually considered for a palliative care approach is Huntington's disease (HD): a genetic neurodegenerative condition characterised by motor and coordination problems and in later stages cognitive impairment. HD is an inherited condition caused by an increased number of trinucleotide repeats in the coding region of the huntingtin gene (Kirkwood, Su, Conneally, & Foroud, 2001). This mutation is autosomal dominant so each child of an affected person has a 50% chance of inheriting it. Of the major neurodegenerative diseases, HD is unique in this respect as individuals with a diagnosis will usually have seen a parent affected by the condition. Its prevalence is estimated to be between four and eight people per 100,000 (Ehret, Day, Wiegand, Wojcieszek, & Chambers, 2007). Symptoms vary between individuals but disease trajectory is predictable; during the condition's later stages, the individual usually has

dementia, marked decline in physical and cognitive functioning and as a result is totally dependent and requires full nursing care. Age of diagnosis of HD is typically around 35-55, although physical symptoms may emerge at any time and life expectancy from time of diagnosis is approximately 20 years (Walker, 2007).

A major area of concern is whether end-of-life (EOL) care for people with HD is managed adequately (McGarva, 2001). As the course of HD progresses it becomes more difficult to meet the basic needs of the affected individual and institutionalised care is often necessary. HD typically has a long illness trajectory and care is often required for many years. An exploratory analysis by Dubinsky (2005) reported that people with HD are usually hospitalised during the later stages of their disease, are often severely disabled and commonly discharged to long-term care facilities, rarely going home. Fitzsimons et al. (2007) suggest that people with chronic conditions often reach the final stages of their life lacking many of the physical and social resources other dying patients may have. People with HD may therefore typically have different palliative care needs to cancer patients and, as a result, hospice care based on the cancer model may not be ideal for people with HD. Equally, older adult residential care may be inappropriate as many people affected by HD are relatively young (Dawson, Kristjanson, Toye, & Flett, 2004). To illustrate, a systematic review of the service needs of younger people accessing dementia care suggested that young people may have different needs and presentations to those of older people, which are rarely met by services (Beattie, Daker-White, Gilliard, & Means, 2002).

Many people with HD are aware that they will functionally decline years before symptoms manifest, particularly with the advent of genetic testing. Despite much opportunity to plan supportive care in advance families often complain of a lack of joined-up, adequate care,

reporting that it is often reactive rather than proactive (Simpson, 2007). For example, although different specialists are commonly involved in HD care it is rare that multidisciplinary teams provide coordinated care (Phillips, Shannon, & Barker, 2008). This is generally problematic in palliative care, especially outside cancer, where there is a failure to integrate services (e.g. Gore, 2000).

Some studies have concluded that insufficient palliative care may be linked to desire for hastened death (E. J. Emanuel, Fairclough, & Emanuel, 2000). Indeed, suicide rates have been reported to be as high as 13% in people with HD, seven to twelve times higher than the general population and higher than rates of people with other medical and neurodegenerative diseases (Di Maio et al., 1993; J. S. Paulsen, Hoth, Nehl, & Stierman, 2005). Similarly, Robins Wahlin et al. (2000) reported a higher risk of attempted suicide for those who received positive genetic tests, five times greater than the general population.

Psychological and social factors such as depression and hopelessness (Breitbart, 2000), anxiety, lower levels of social support and feeling like a burden (Kelly et al., 2003) have been linked to desire for hastened death in a number of studies of terminal cancer patients at the end of life. Given the cumulative difficulties and life changes often imposed on people with HD, such as loss of identity, job, mobility, independence and personal relationships, as well as advanced awareness of these difficulties, it is possible that desire for hastened death is similarly a prominent issue for this population. Many people at various stages of HD (including those who carry the gene but are pre-symptomatic) experience emotional difficulties, (Craufurd, Thompson, & Snowden; Julien et al., 2007). For example, Paulsen, Ready, Hamilton, Mega and Cummings (2001) found that dysphoria, agitation, irritability, apathy and anxiety were prevalent in over 50% of people with Huntington's disease surveyed. The authors also suggest that these mental

health problems are predominantly separate from the cognitive and motor symptoms of HD. This is supported by a cross-sectional study by Zappacosta et al. (1996), which found no correlation between disease severity and mental health problems such as anxiety and depression in HD.

Suicide is usually regarded as irrational and the product of mental illness (Leeman, 2009). In congruence with this, suicide prevention strategies are enshrined as a priority in guidelines for public health and mental health services (e.g. UK National Suicide Prevention Strategy, Department of Health, 2012). This is an ethical aim, as people may have conflicting wishes, fleeting or impulsive suicidal urges, or be in a depressed state and have lost insight that meaningful recovery is possible (Clark, 1992). However, Leeman (2009) argues that the assumption that suicide is always irrational is crude. It is possible that given the suboptimal EOL care often received by people with HD suicide could be a rational, well-reasoned decision to prevent deterioration and future dependence. This is congruent with the respect for patients' autonomy and choice, which is central to modern healthcare (Tauber, 2005).

Hendry et al. (2013) suggest that the term 'assisted dying' encompasses both 'assisted suicide' and 'euthanasia'. The former was defined by the European Association for Palliative Care as "a doctor intentionally helping a person to commit suicide by providing drugs for self-administration, at that person's voluntary and competent request" (Materstvedt et al., 2003, p.98) whereas the latter refers to a doctor administering drugs to intentionally kill a person at their voluntary and competent request. There is increasing current debate in developed nations over whether assisted dying should be legalised. This is a contentious issue; in many European countries legalisation is opposed by a range of professional bodies representing doctors and nurses, and also by palliative care organisations (Materstvedt et al., 2003). Contrastingly, there is wide public support for legalisation. In the UK, for example, a YouGov survey for Dignity in

Dying (2013) found that 76% of people were in favour of the legalisation of assisted dying provided that appropriate safeguards were put in place. In the United States, approximately 66% of the population reportedly favour assisted dying (O. D. Duncan & Parmelee, 2006). Research has attempted to understand the views of physicians, nurses and carers on assisted dying but the perspectives of people most affected by the issue, people with terminal or life-limiting conditions, are seldom investigated (Wilson et al., 2007).

Given the increased mental health difficulties, desire for hastened death and suicidality experienced by people with HD, assisted dying is an important issue for this population. Research suggests that people who have life-limiting conditions generally favour assisted dying. For example, Chapple, Ziebland, McPherson and Herxheimer's (2006) study on the views of people with a terminal illness found that most participants thought that UK law should change to allow assisted dying; participants who had seen others die held particularly strong beliefs on an individual's right to die. Additionally, people often have prior knowledge of the existence of HD in their family. As a result they may have ample time to consider their end of life and come to decisions informed by their families' experiences.

In the Netherlands, in line with the Euthanasia Act (Netherlands, 2001), assisted dying is a viable option for people with HD in the absence of any effective treatment. People with HD are increasingly considering these options, with between six and ten assisted dying requests from people with HD granted each year in The Netherlands between 2007 and 2011 (Booij, Rödij, Engberts, Tibben, & Roos, 2013); however, little is known of how assisted dying is understood and positioned psychologically, socially and existentially by individuals with HD.

The only known study exploring the thoughts and attitudes of people with HD on assisted dying and the use of advance directives in HD is by Booij et al. (2013). The authors reported that

the most important frame of reference for EOL wishes was experience with an affected parent. Participants' knowledge of the law was reportedly limited. This study's design was to provide a grounded theory from the interview data collected. However, a theoretical framework was not evident from the results. There appears to be limited exploration of how affected individuals construct the meaning of HD within their lives and how these understandings shape their views on assisted dying. This gap in knowledge remains. Furthermore, there is no extant research exploring the views and beliefs of people with HD about the concept of assisted dying in countries where this option is unavailable.

Consequently, there is further need for exploratory research in this area. Qualitative research can yield rich and detailed insights into people's views and experiences (Smith, 2007). A qualitative design was thought to be most suitable for providing an in-depth analysis of how assisted dying is understood and positioned within the lives of individuals and the context of having HD. This study therefore aims to use qualitative methods to achieve greater theoretical understanding of the views of people with HD on assisted dying within the context of their own lived experiences.

Method

The research question was to explore what people with HD think about assisted dying and what informs these views.

Design

Semi-structured interviews were conducted and analysed using thematic analysis. This approach is theoretically flexible and focuses on drawing patterns of meaning (Braun & Clarke, 2006). It lends itself to exploratory research (Boeije, 2009) and there is a robust precedent for its use in

healthcare research (Braun & Clarke, 2014). It has previously been used to interpret HD patients' experiences of predictive genetic testing (R. E. Duncan et al., 2007).

Procedure

Ethical approval was sought and acquired from the researcher's academic institution's Research Ethics Committee. To recruit participants, a purposive strategy was employed. This included posters displayed at local meeting places for people with HD, advertisements on social media and on the Huntington's Disease Association website. The purpose of the study was made clear and people who were interested in taking part were able to contact the lead researcher directly for more information or opt to take part. Approximately 18 people inquired about the research and, of these, seven met the following inclusion criteria: participants had a self-reported confirmed diagnosis of HD or were pre-symptomatic gene positive; they were aged 18 or over; and they spoke English to a sufficient level to be interviewed. No participants had symptoms severe enough to make interview impossible. At opt-in potential participants were intentionally screened to ensure they had heard of assisted dying and were happy to offer their opinions on the topic with reference to their own experiences. Following this, interviews were arranged.

Participants

Seven people participated in the study. This comprised five women and two men whose age ranged from 27-47 ($M= 35$). Four participants had children, three were single, three were married and one was separated. All participants identified as British and were either pre-symptomatic or had begun to experience symptoms but were at an early stage of the disease trajectory.

Participants were assigned a pseudonym to maintain anonymity.

Interviews

Five interviews were conducted by phone and two interviews were conducted face-to-face based on participant preference.

An interview schedule was used to guide questioning but allow the lead researcher to follow areas of interest to the participants and be receptive to new ideas. A digital Dictaphone was used to record interviews. At interview, the lead researcher discussed consent and confidentiality with the participants and it was explained to them that they were able to withdraw their participation, or their data, at any point. Any further questions were answered and written consent was obtained.

Interviews lasted between 37 and 66 minutes ($M= 47$). Sensitivity to participants' anxiety levels and language skills were monitored throughout each interview to ensure inclusive practice. Participants were debriefed following interview. Interviews were transcribed verbatim by the lead researcher as soon as possible after interview and stored on a secure computer. All identifiable information was anonymised.

Analysis

Inductive thematic analysis was used to analyse the data from the interviews. The aim of analysis was to identify patterns of meaning across the dataset about people's views on assisted dying.

Analysis was entirely data-driven, with no pre-existing theory or framework applied.

An idiographic approach to the data was taken, with the first interview analysed in detail before moving on to the next, case by case. Analysis ran simultaneous to data collection after the first two interviews were transcribed to gain familiarity with the data. Therefore, the analytical process began during the data collection phase, because each interview conducted and transcribed influenced subsequent ones.

The five steps of thematic analysis as outlined by Braun and Clarke (2006) were used; (1) familiarising self with the data, (2) generating initial codes, (3) searching for themes, (4) reviewing themes, and (5) defining and naming themes.

The analysis initially involved familiarisation with the data through transcription. Each transcript was read and re-read to increase familiarity with and immersion in the data. The transcripts were systematically annotated, with points of interest or particular significance made and returned to at later phases of analysis. The generation of initial codes was achieved through line-by-line coding of each transcript. Initial codes from the dataset were inputted into a table, supported by relevant extracted quotes. Consideration was then given to how codes could be compared and grouped according to patterns across the data that were relevant to the research question. These items were then examined and emerging themes identified. A summary list of themes based on all the transcripts was constructed and superordinate themes were obtained. The transcripts were constantly re-examined to ensure that the themes still related to the original texts.

Quality appraisal

Arguably, it is less easy to evidence quality and reliability in qualitative research than it is in quantitative (Morse, Olson, & Spiers, 2002). The analysis focused on sensitivity to context, rigour, coherence, transparency and importance: core principles of validity and quality in qualitative research (Yardley, 2008). These key indicators are considered more suitable than criteria traditionally applied to quantitative research to assess quality.

Reflexivity

I was reflexively aware of my own strong views in favour of the legalisation of assisted dying, which will have inevitably affected the way I understood the data. For example, I strongly believe that not every death can be ‘good’, in accordance with the aim of palliative care, so as a

civilised society, we should legalise assisted dying. However, through good use of research supervision and keeping a reflexive log I recognised and “bracketed” my views and perceptions (Lester, 1999, p.1) as much as possible. Additionally, I have made them available for scrutiny and consequently believe that the analysis presented remains compelling and trustworthy.

Results

Four themes were identified in the analysis of the interview data: 1) Autonomy and kindness in assisted dying; 2) HD threatens identity but is part of life; 3) Dilemmas in decision-making on assisted dying: “There are no winners”; and 4) Absence of explicit discussion on dying and HD: “Elephants in the room”.

Autonomy and kindness in assisted dying

This theme summarises the two central beliefs expressed by participants: that an individual has a right to autonomy and that assisted dying is as an act of kindness.

Participants viewed assisted dying as a way of taking control of their situations and exercising their right to autonomy.

It is enabling somebody to escape illness. To be able to end a life in a dignified manner... rather than having to go through the pain and suffering that you may have had to go through otherwise... And just be able to be in control of your final moments rather than it controlling you. (Claire)

The “right to die” (Peter) was conceptualised as a freedom that is yet to be won. Peter drew parallels to the debate around abortion, where freedom of choice is similarly juxtaposed with arguments to protect life.

Assisted dying was deemed a conditional right, participants acknowledging that safeguards must be in place to protect people and prevent abuse. Although they demonstrated

awareness of arguments against assisted dying, overwhelmingly participants believed that personal autonomy was paramount and that individuals were best placed to make such decisions about their own deaths provided they had the capacity.

[The] biggest concern is people in a vulnerable position, you know, if they're really ill and they're not able to make their own decisions. But surely if someone is sound of mind... and people can understand that person's wishes, I think quite strongly that it should be their right. (Mary).

Most participants feared prolonged suffering at the end of their lives and talked about a point beyond which life would not be meaningful or when suffering would be too much to bear. This point varied but all participants emphasised the importance of choosing that point for themselves. Assisted dying represented freedom from suffering.

I don't have any suicidal ideation or a plan. But I do have intent or a desire to take my own life at some point to prevent myself from being at a point of suffering and losing my dignity with this condition. (Anna).

Relieving suffering through assisted dying also embodied an act of kindness.

I think if we had had something like this available, although it would have meant we would have lost Nan a lot sooner, I think it would have been kinder to her. Because she was in and out of care homes... they kicked her out because they couldn't cope with her. They weren't designed to deal with Huntington's. (Claire).

Conversely, to leave someone with suffering was thought to be "a great cruelty" and a violation of that person's right to autonomy and dignity (Anna). Peter compared assisted dying and sedation at end of life as methods of ending suffering and acts of compassion: "It is effectively assisted dying by the back door."

Participants lamented the perceived lack of choice and kindness in the legal status quo and used evocative, persuasive language to convey their depth of feeling. “You wouldn’t let your own dog suffer so really why should you put a human that you love through that suffering” (Mary).

Assisted dying was also considered a kindness to families, as the alternative would be for them to bear the burden of witnessing participants suffering, potentially for many years, or for participants to request assisted dying from families, also thought to be a terrible burden.

HD threatens identity but is part of life

HD strongly informed participants’ views on assisted dying: discovering they had inherited the gene or being diagnosed formed their beliefs or reinforced their existing ones.

My thoughts changed when I was diagnosed with Huntington’s, I suppose. I hadn’t thought about it until then, or thought enough about it... I have become more aware, and I’ve become more understanding about why people would want that right. (Lola).

Participants reported that these views had remained stable over time. Consequently, they spoke about HD changing their future self-representations, suggesting that suffering went beyond physical pain to their very self-concept.

People shouldn’t have to die alone and a shell of what they were. Nan was a vegetable by the end of it, bless her, she was literally just a case. There was nothing left inside. We were just keeping her alive but what for? She wasn’t getting any joy out of life. (Claire).

This vivid language demonstrates how previously distant possibilities such as suffering, loss of functioning and death became more proximal for participants. They reported having to question previous assumptions about themselves, the world and the future.

My job as a counsellor is very much about emotional instinct and being emotionally aware. So it's really huge for me that I've been diagnosed with a condition that is likely to influence my ability to be good at what I cherish about my role... It's awful. (Anna)

As illuminated here, potential loss of role, personality and meaning were considered the most disruptive aspects of the disease and therefore more relevant in deciding the point at which assisted dying would be desired over loss of physical functioning. "I think the idea of the physical symptoms scares me a lot less than it used to and the fear of the mental symptoms scares me a lot more." (George). Physical functioning seemed important mainly in an interpersonal context, for example, in not being able to communicate. "Obviously just like being able to function, get out and have conversations with people... cos it can massively affect your relationships with people." (Dawn). The point at which assisted dying would be desirable was therefore connected to loss of self for participants.

Medicalisation of both HD and dying contributed to this threat to sense of self; participants felt that their psychological, social and existential needs were ignored at the expense of their physical needs. Anna noted the incongruence between progress as defined by medicine and what participants ascribed meaning to:

There's moments where [the psychiatrist] says almost that it's good that your mum is still alive, like it's a good thing that we can keep people with Huntington's alive for longer. But they have in no way been able to sustain her quality of life. And, as a result I don't see it as a triumph that she is still breathing. And that doesn't look like progress to me... At the end of it, the horribleness of the disease is just disguised for a while. (Anna)

In this participant's experience, concepts of HD, suffering and progress are rooted in a biomedical framework for healthcare professionals. This is dissimilar to the ways in which

participants made sense of their HD, qualifying these concepts through their own value and belief systems, for example, placing importance on “personal choice and individualisation” (Dawn).

Together with the language of loss and disruption to future selves, participants expressed a sense of continuity. They maintained their respective personal identities by continuing their work, social and family lives and appeared to adapt them around HD to continue living meaningfully. For example, one participant terminated a pregnancy upon discovering that she had passed the gene on to the embryo. Following this experience, she decided not to get pregnant again.

Cos I just thought, for me, I would not put my child through what I'd been through. And since then I've just made the decision I'm just not going to go there. I just don't think it would be fair on me or to bring a child into the world... But I still think I made the right decision and I don't regret having the test or what happened after. (Mary)

Similarly, for many participants, there was an awareness of HD in their families so it formed part of their family narrative and the context of their lives. Although their personal diagnosis of HD was not necessarily anticipated, it was a strong possibility given the genetic basis of the disease. Accordingly, continuity was illustrated with regard to the heritability of HD. “[HD] has always been there in my life one way or another. So it's not all of a sudden you've been diagnosed with something out of the blue. This has always been there.” (Mary). Participants framed their own diagnosis of HD and expectations for the future as congruent with the EOL experiences of affected family members, for example, assuming similar symptoms and trajectory.

The problem with it being – is I'm now watching my mum how my mum watched her mum. She knows what's coming, and I suppose in that sense it makes it worse for my mum, because she already knows. (Lola)

Participants spoke about formative experiences of witnessing family members dying with HD. Their views on assisted dying were embedded in these family experiences. For example, although Peter was in favour of legalisation of assisted dying, this was not something he would ever consider for himself. This was because he saw that his father still had a sense of self and quality of life despite the severity of his symptoms.

He's been bedridden for the last 5 years now, can't speak, can't eat, can't do anything for himself. But he still has a certain quality of life. I took him out in my car the other week for a trip out. But he actually put his arm around me. So he's still in there but he just can't communicate as well... I can sort of observe what's happening to him and make decisions based on that. And that helps inform me [about assisted dying]. (Peter).

Anticipating their disease trajectories, participants adapted their perspectives on life and plans for the future. Some participants spoke about a life-affirming aspect of HD and reported "a new way of looking at life" (Lola).

I guess part of me, knowing my diagnosis, has made me seize the day a bit. It sounds a bit corny, but it's kind of about making the most of all the good days, being thankful, being mindful, taking notice of things that bring me great joy. And that sense of responsibility of living a good life has also made me think about that moment when that good life is becoming diminished. (Anna).

Anna anticipated that she would be unable to maintain her quality of life as a result of HD symptoms in the future, at which point she would desire assisted dying.

HD therefore formed part of the biographical narratives of participants and influenced their views on assisted dying.

Dilemmas in decision-making on assisted dying: “There are no winners”

This theme encapsulates the personal and practical dilemmas inherent in participants’ situations, in particular around their stance on assisted dying. Assisted dying was considered to be an imperfect solution to a real world problem. Some participants were optimistic about medical advances; a cure was the “ultimate and desired end goal” (George) and would obviate the need for assisted dying in HD. However, given this is not a reality, assisted dying was thought to be necessary. The only alternative was to live with suffering or commit suicide by other means.

I don’t want it there just as an escape route but for those who genuinely do need it. And the only way out is either continue as you are until something happens to you or starve yourself or have to take your own life (Claire).

Claire elaborated on this view, saying that “you lose your dignity” in violent suicide, or voluntary refusal of food and fluid - as in the case of her grandfather.

Another dilemma across participant accounts was the conflicting views of family. Often there were multiple stakeholders in EOL decision-making and participants reported times where different views and rights came into conflict.

I just don’t wish to go through that process of decline. I don’t wish it for my mother, but I appreciate that other people are taking those decisions... I think my dad would feel very guilty if there were to be any intervention to end my mum’s life sooner than it naturally would (Anna).

Linked with first theme, *Autonomy and kindness in assisted dying*, freedom of choice was most important for participants. However, this was frequently balanced with the wants and needs of family.

It might be, for example, it is not worth it for me directly but for whatever reason I still keep my wife company. And you might drag it out another year or something... I guess it wouldn't be just about me, but at the same time it is not about many people other than me. (George).

Another predicament identified by some participants is that the point at which suicide may be desirable would be the point at which it was no longer an option for them because they would be too impaired physically or cognitively.

Sometimes with the assisted dying thing I think, "Well, it's pretty easy. All I have to get myself a packet of paracetamol or whatever, and plan things in advance." And the day I think that, I am going to be in a position where I can't take my own life. (George).

Similarly, Anna acknowledged this tension and wondered at what point she should document her wishes formally.

When I read the HD literature and it talks about kind of losing the ability to initiate decisions or take action – so, again, that alerts me to, well hang on a minute, if I'm declining gradually, am I losing the ability to make those decisions? So do I need to make that decision long before I'm symptomatic in order to ensure that I don't miss my opportunity? (Anna).

Claire contemplated going to Dignitas (a Swiss assisted dying organisation) in the future assuming no change in the legal status of assisted dying in the UK. However, she highlighted the difficulty of being well enough to travel but ill enough to warrant treatment.

To do that you have still got to be well enough to travel so it is a Catch-22 situation. You have got to be well enough to travel but then you have to be poorly enough to warrant going somewhere like the clinics. (Claire).

Additional considerations for her were having the funds to travel to Switzerland and the place of death being a strange country, without many of her family and friends.

Some participants reflected on the 'battle' narrative that permeates living with life-limiting illness. Anna felt constrained by this metaphor and rejected it. "There's this kind of thing about people bravely fighting on and things like that, and battling, and all those words associated with terminal cancer. But I don't want to fight that fight." She elaborated on this unsuitable metaphor with regard to the prolonged battle that people with HD must face. This was felt to be particularly important in relation to the conservative proposals to changes in UK legislation.

They talk about people within six months at the end of their life. With Huntington's that's not applicable. That wouldn't be of any benefit, because the decline is such a long, drawn-out process. I don't think any doctor can predict where six months off death is for somebody with Huntington's. Because in my experience of my mum's condition, she has been awful for years and close to death for years yet sustained by tiny amounts of food and nutrition (Anna).

It can be seen here how contemporary debate on assisted dying was irrelevant to participants, as it tends to exclude conditions such as HD or dementia.

Congruent with the battle metaphor, Lola admitted feeling that accepting death was tantamount to "admitting defeat" and cited this as a reason why her mother would be likely not to accept assisted dying. This demonstrates how value-laden this particular metaphor is and how it privileges certain ways of coping and living with chronic illness.

Absence of explicit discussion on dying and HD: “Elephants in the room”

Emergent in the data was the idea that both assisted dying and HD were difficult and often unmentionable topics of conversation. Although many participants described their families as supportive, there was reportedly little discussion about death or wishes for EOL care.

Participants considered the cultural taboo of death as one of the reasons behind this reticence.

I said to my mum before about the fact that I think it should be somebody’s free choice to choose something like that, but not in depth. I suppose I don’t want to say that sort of stuff. It’s a bit morbid isn’t it, to your parents? (Laughs). (Lola).

Views on EOL care were also often not talked about explicitly with healthcare professionals. Participants spoke about the need for a balancing act between feeling supported and feeling distressed or offended by conversations about EOL issues.

It is a really difficult balance. And I know that’s the same with family and friends as well as medical professionals. It’s kind of like people wanting to talk to you about it enough so that you feel heard, but not wishing to distress you by bringing it up at times when it feels uncomfortable. (Anna).

Many participants expressed the view that this balance was not being achieved, with most having never had conversations about end of life with healthcare professionals. Consequently, some participants were unaware of EOL care options. For example, two participants had not heard of advance directives. Some participants felt this reticence was inadequate and put the onus on healthcare professionals to facilitate conversations about assisted dying. Others thought it more acceptable because of the difficulty of the subject.

I wouldn’t know when the right sort of age to start talking about dying is. It’s a tough one. Some people could be seriously offended by it. (Laughs)... But then I probably would

start talking about it quite early on because you never know how quick Huntington's is going to affect some people. (Lola).

More implicitly, participants communicated that HD itself was also not talked about. Participants largely felt left to manage by themselves and some reported feeling lonely with their HD. "Because I am symptom free, I'm also kind of doctor free... I guess there's time when I feel a bit adrift maybe with it; kind of like a bit left alone with it". (Anna).

A commonly held belief was that services should be more integrated, as participants reported frequent experiences of fragmented and incomplete care. "But with something like Huntington's where you know what the outcome is going to be and where you know what is going to happen, I think there should be more of a support network in place." (Claire).

Some participants talked about not having open discussions with families, instead having "hushed conversations" (Anna) or implicit understandings. For some this was an active choice. For example, George described a significant period where he chose not to disclose to his mother that he was gene positive as: "it's just extra time to worry more." For other participants, the uncommunicativeness within their families was not their decision. For example, Dawn talked about her family support system but acknowledged that her father did not talk about HD and withdrew from the family: "I think that was more that he didn't want us to see him declining, so he kind of distanced himself." Similarly, Anna reported that her father didn't reveal that her mother had HD.

There is this very fractured, vulnerable group that has really been quite shattered by – in my circumstances, it was to do with the secrecy around Huntington's being in our family, and my father feeling unable to reveal that knowledge he had to us all. (Anna).

Anna spoke about the damage that this caused her family, particularly because she was not aware of HD in her family before choosing to have children. She made a conscious different choice to tell her children about HD: “I very strongly feel that part of my role as a parent is to role model being OK with having Huntington’s.”

Many participants echoed the sentiment behind this choice, reporting that communication is important in both offering and receiving the support needed to manage and cope with HD.

Discussion

The study developed rich insight into how people with HD view assisted dying within the context of their lived experiences of having the condition. Participants expressed varying levels of distress, often using poignant language and metaphor to elaborate on their thoughts and feelings on assisted dying. Despite the emotive topic, participants demonstrated well-reasoned decisions and awareness of other perspectives. They reported that their views were stable over time, which accords with a qualitative study by Ho, Gilbert, Mason, Goodman and Barker (2009) that explored the concerns of people with HD and found that the impact of emotional, social and identity issues were most important at pre-symptomatic stage and remained stable throughout the trajectory of HD.

The views and arguments offered by participants on assisted dying are represented in wider literature on assisted dying. Participants all thought that the law should provide for assisted dying. In a comparative analysis of the attitudes of 33 countries to euthanasia, Verbakel and Jaspers (2010) identified two dominant arguments in favour of assisted dying. The ‘autonomy argument’ proposes that individuals should have the ultimate right to make decisions about their life and death (Scherer & Simon, 1999) and is consistent with the discourse of individual choice and autonomy used by participants in this study. Similarly, the discourse of compassion

employed by participants is consistent with the ‘death with dignity argument’ (Scherer & Simon, 1999), which argues that euthanasia relieves suffering and spares people from a drawn-out, painful death. This study and others (Fadem, 2003; Lavery, Boyle, Dickens, Maclean, & Singer, 2001) support the authors’ prediction, that people with personal experiences of witnessing suffering are more tolerant of euthanasia. All participants were in favour of legal provision of assisted dying, similar to other people with life-limiting conditions (Chapple et al., 2006).

Participants drew heavily on family experiences of suffering. This accords with findings by Maxted, Simpson and Weatherhead (2014) that family members shared the impact of HD. As well as being rooted in the past, in terms of their own family experiences, support for assisted dying was framed by participants’ ideas about a hypothetical future. Loss of self is a widely researched concept in relation to chronic illness. Bury (Bury, 1982) described the phenomenon of biographical disruption, whereby chronic illness necessitates a re-evaluation of one’s self-representation and life narrative. Congruently, Charmaz (1983) also suggested that illness becomes dominant and the associated sick role or patient identity overtakes other concepts of self. These ideas have influenced subsequent understandings of coping with chronic illness and been described in association with various illness experiences such as chronic fatigue syndrome, fibromyalgia, cancer and MND (Asbring, 2001; Locock, Ziebland, & Dumelow, 2009; Park, Zlateva, & Blank, 2009).

Participants in this study adhered to some aspects of these ideas, for example, experiencing strong emotions such as fear, sadness and anger in relation to perceived losses. However, participants also maintained their personal identities through perceived continuity with their family members who had HD; this was enhanced by close supportive relationships with their family. This was recognised by participants as hugely influential in their views on assisted

dying. Recent research supports these findings, contradicting the view that loss and disruption are inevitable for people with chronic illnesses. For example, Faircloth, Boylstein, Rittman, Young and Gubrium (2004) explored stroke recovery narratives and found various factors, such as age, previous knowledge of illness experience and pre-illness behaviours, which tempered biographical disruption and created more of a biographical flow. Similarly, Nanton et al. (2016) explored how advancing illness affected individuals' personal identities and found that pain and symptom control as well as connections with life pre-illness were important in maintaining a sense of self.

Participants in this study faced personal and practical dilemmas in their views on assisted dying. Dilemmas evident in living with chronic illness are well reported in research. For example, studies have found that people with multiple conditions felt morally obliged to manage well (Townsend, Wyke, & Hunt, 2006) and that people with multiple sclerosis struggled to "pass" as unimpaired and not be perceived as mentally ill, drunk or malingering (Robinson, 1990). Interestingly, participants in this study mainly identified personal and practical dilemmas rather than ethical dilemmas that might typically be the focus of assisted dying debates (Sanders & Chaloner, 2007), bridging the gap between the personal and political.

Comparable to the participants in the Netherlands study by Booij et al. (2013), most participants wanted to discuss HD and end-of-life issues but lacked certainty on how and when to do this appropriately. Participants' experiences of few conversations about death and EOL care align with findings that healthcare professionals tend not to communicate well about EOL issues (Milberg & Strang, 2000). Similarly, participants' experiences of feeling unsupported with their HD fits with research findings that the needs of people with HD, including informational needs, were not met by services which were described at best as ad hoc and reactive (McGarva, 2001).

In the current study some participants were unaware of advance directives. Furthermore, two reflected on the futility of documenting wishes due to their options being restricted. The use of advance directives may be particularly useful within this population; as a result of progressive neurodegeneration in HD and the associated decline in cognitive ability, it can be difficult to comprehend a person's wishes at end of life. Research suggests that despite being widely advocated, advance directives are infrequently used (Barnes, Jones, Tookman, & King, 2007). Congruently, research shows that for people with MND decisions about EOL care are often delayed. This is despite a systematic review by Brinkman-Stoppelenburg, Rietjens and van der Heide (2014) indicating that advance care planning improves the quality of EOL care. Additionally, doctors and patients, including people with neurodegenerative diseases (Poppe, Burleigh, & Banerjee, 2013), seem to be favourably disposed to advance directives (L. L. Emanuel, Barry, Stoeckle, Ettelson, & Emanuel, 1991).

Practical Implications

Consistent with participants' wishes, healthcare professionals should facilitate an open, communicative atmosphere and EOL decisions should be addressed in a timely manner. Participants should also feel supported with their HD with coordinated and integrated care throughout the trajectory of HD. Advance care planning should be facilitated so that the autonomy of people with HD is maximised; barriers to implementation such as lack of resources (Seymour, Almack, & Kennedy, 2010) and perceived difficulty to do (Boddy, Chenoweth, McLennan, & Daly, 2013) should be identified and addressed.

Participants privileged quality of life over symptom severity or closeness to death in arguments for assisted dying and felt that the psychological impact of HD was more threatening than physical changes. This accords with research stating psychological aspects of HD are one of

the dominant factors influencing quality of life, above motor problems or cognitive impairment, and are most strongly associated with stress and impairment (e.g. Ho et al., 2009). Since assisted dying is illegal in the UK, weight should therefore be given to the psychological impact of HD and efforts made to reduce it and enhance quality of life. People with neurodegenerative diseases have expressed an interest in psychological interventions (Aoun, Kristjanson, & Oldham, 2006). Moreover, research in this area has shown that psychological factors such as illness beliefs and coping mechanisms play a major role in people's mental health and wellbeing (Arran, Craufurd, & Simpson, 2013; Kaptein et al., 2007). A systematic review of pharmacological interventions in HD by Bonelli & Wenning (2006) revealed that there is "very low" evidence for the efficacy of medication, with many commonly used antidepressants being considered only "possibly useful". However, psychological therapy has an emerging evidence base for people with neurodegenerative diseases such as Parkinson's (Dobkin et al., 2011; Fitzpatrick, Simpson, & Smith, 2010). Evidence also suggests that the impact of HD extends beyond the affected individual to impact upon the family. For example, it is common for family members to become caregivers and experience stress-related illness, reduced quality of life, negatively affected relationships and psychological problems such as low mood (e.g. Dawson et al., 2004; Maxted et al., 2014; McCabe, Firth, & O'Connor, 2009; Read et al., 2013). A systemic psychological approach (Dallos & Draper, 2010) would therefore be beneficial in assisting the whole family and ensuring that psychological problems are not solely located within the affected individual and social context is not lost. Congruently, healthcare professionals should be mindful of continuity within the context of people's lives, such as role, family and other interpersonal relationships in order to minimise threats to identity.

Although it is important to provide more resources to help people in distress and desiring death, improving palliative care does not eliminate the case for assisted dying. Mental health professionals have an important role in assessing people requesting assisted dying, for example, for lack of competence. However, caution should be exercised in assuming that alternative options would be preferable or that people may be talked out of decisions to end their lives. Similarly, suicide prevention measures should be judiciously implemented and individuals' desire for hastened death should not be pathologised. This study has demonstrated that despite distress associated with impending decline in HD, people can make considered, rational decisions about assisted dying.

Participants' views should therefore be considered and used to inform current debate on the legalization of assisted dying in countries where this is not legal. For example, participants' emphasis on quality of life and their lack of agreement over the point at which assisted dying should be accessible is relevant for legislation. Even if assisted dying were more widely legalised this may not be accessible for individuals with HD. Generally, stringent procedures for euthanasia and assisted dying exclude people who are not terminally ill. For instance, restricting assisted dying to those with less than six months to live (e.g. Assisted Dying Bill, UK Parliament, 2013) excludes many people with chronic, life-limiting conditions such as HD where trajectory is less certain. Findings by Suarez-Almazor, Newman, Hanson and Bruera (2002) suggest that psychosocial factors such as depression, anxiety and poor wellbeing were more important determinants in decision making about assisted dying. Consequently, autonomy should be at the forefront of EOL care.

Limitations

Plurality of opinion is desirable. Of the self-selected sample all participants supported legalisation and all but one indicated willingness for assisted dying themselves. Additionally, participants did not reflect all stages of HD. Although cognitive deterioration was less likely to have presented an issue for this sample, participants further along in their disease trajectory might offer additional insight.

Future research

Further exploratory research would be beneficial in this area using purposive sampling, seeking to include people with opposing views. This would further illuminate how people construct their views. It is also recognised that views do not necessarily correspond to people's actual future behaviour (de Leeuw, Engels, Vermulst, & Scholte, 2008). Therefore, caution is exercised in interpreting the views of participants as predictive of future behaviour. Future longitudinal research could measure the relationship between attitudes towards assisted dying and future behaviours.

Conclusion

This study has explored the views and beliefs of people with HD about the concept of assisted dying in a country where this option is unavailable. It has identified that people with HD consider historical family experiences and hypothetical futures in constructing their views on assisted dying. They value maintaining control of both their life and their death. Dilemmas in decision making exist and EOL discussions are limited. This study adds to the evidence base and gives voice to the often-overlooked individuals who are most likely to consider assisted dying were it legal. Their views should inform current debate in the UK and elsewhere.

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Appendix 2-B: Excerpt of an Analysed Transcript

L:	Would you start by just telling me what your understanding is of assisted dying?	<p>Laurence 31/5/16 00:27 Comment [1]: Escape illness</p>
P:	<p>To me it is enabling somebody to escape illness. To be able to end a life in a dignified manner. To not have to go to suicide or... Just be able to have the change with your loved one, to have your final moments when you are still here of sound minds. And to be able to die with the respect that you deserve rather than having to go through the pain and suffering that you may have had to go through otherwise.</p> <p>So, it is just enabling you to take control of it yourself I suppose. And just be able to be in control of your final moments rather than it controlling you because... I think especially with my Nan it was a case of got to think if she had had the chance to do the assisted dying it would have meant that she could have died in a lot less pain and without the suffering. And I think that is the biggest thing. It is just to be able to give the control back to the person and for them to be able to have the element there, to be able to say, "Enough's enough I've done what I need to do now." And it is just saying goodbye to friends and family and having that chance to be able to do it in a much more controlled way, which I think is going to be a big thing for people. And not to have to resort to suicide or those kinds of things.</p>	<p>Laurence 31/5/16 00:28 Comment [2R1]: Giving the means</p> <p>Laurence 31/5/16 00:28 Comment [3]: End life with dignity</p> <p>Laurence 31/5/16 00:29 Comment [4]: Awareness of change</p> <p>Laurence 31/5/16 00:29 Comment [5R4]: Significant/loved ones</p> <p>Laurence 31/5/16 00:30 Comment [6]: Cognitive deterioration</p> <p>Laurence 31/5/16 00:30 Comment [7]: Deserving respect</p> <p>Laurence 31/5/16 00:31 Comment [8]: Avoid pain and suffering</p> <p>Laurence 31/5/16 00:31 Comment [9]: Autonomy and control</p> <p>Laurence 31/5/16 00:31 Comment [10]: Reference to family</p> <p>Laurence 31/5/16 00:32 Comment [11]: Avoid pain and suffering</p> <p>Laurence 31/5/16 00:32 Comment [12]: Importance of autonomy and control</p> <p>Laurence 31/5/16 00:33 Comment [13]: Completing life</p> <p>Laurence 31/5/16 00:33 Comment [14]: Saying goodbye</p> <p>Laurence 31/5/16 00:33 Comment [15]: Importance of autonomy and control</p>
L:	I am sure you are aware; it is not currently legal in the UK?	<p>Laurence 31/5/16 00:33 Comment [16]: Suicide a lesser option/last resort</p>
P:	Yes.	<p>Laurence 31/5/16 00:34 Comment [17]: Assisted dying should be legal</p>
L:	What are your thoughts about that?	<p>Laurence 31/5/16 00:34 Comment [18]: Taking control</p>

Appendix 2-B: Example of the development of an analytical theme

Supporting quotes	Codes	Initial themes	Analytical theme
“Huntington’s is one of the big diseases assisted dying would be useful for. Because it affects every aspect of the person’s life, physically and mentally” (Dawn)	Impact of HD HD affects views on assisted dying Loss of dignity Personality changes	HD threatens <u>identity</u> <u>Life affirming aspects of HD</u>	HD threatens identity but is part of life
“I know it would be a death but then you would still be the same person. With Huntington’s your brain changes.” (Lola)	Powerful emotions Quality of life over quantity Suffering is inevitable Loss of role	Family experiences shape views on assisted dying	
“I wouldn’t want to stay beyond the point where I have got no quality of life” (Claire)	Importance of values in decision making Fear for future		
“Do you know even if I get symptoms today, I would still be living in hope.” (George)	Imposition of time frame Seizing the day Living in hope		
“I have no wish to suffer with an illness in the same way I’m watching my mother decline... I think she is alive well beyond a point of any quality of life or any kind of dignity or joy...” (Anna)	We’re all going to die Advantages of HD HD in family Loved ones important Death in family Family history		
“I can always remember my mum being poorly. She died when she was 57 so she was ill a long time. That’s why I have such strong views about assisted dying. Why put somebody through that?” (Mary)	Context of life		