The Intersection between Voluntary Assisted Dying, Suicide and Advance Care Planning in Huntington's Disease

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Abstract

Huntington's Disease (HD) is a complex neurodegenerative disorder, with a life expectancy 10-20 years after symptom onset which is usually in middle age. Symptoms of HD include physical deterioration, cognitive impairment and a high incidence of suicide and suicidal ideation, which may cause intolerable suffering. While the strong association between depression and other psychiatric manifestations with suicide is discussed, this paper explores whether suicide can ever be deemed rational. By the time a person with HD reaches an advanced and terminal stage, their decision-making capacity will be so impaired as to make them ineligible for Voluntary Assisted Dying (VAD) under all current and proposed legislation in Australia and New Zealand. Given that Advance Care Planning (ACP) can provide an opportunity to refuse all medical treatment, except palliative treatment, could this provide an acceptable alternative to suicide and VAD? Nurses need to consider these ethical dilemmas and remain informed on the debates. They may experience moral conflict when a person wishes to discuss their view on rational suicide or seek to implement refusal of treatment in an advanced care directive. They must be aware and consider how their professional obligations and personal opinions interact with this complex topic.

KEYWORDS: Huntington's Disease, Advance Care Planning, Voluntary Assisted Dying, Suicide, Rational Suicide, advanced care directive, end of life.

Introduction

Huntington's Disease (HD) is an incurable progressive disease, that will advance to a stage that may cause intolerable suffering that is unable to be relieved in a manner considered tolerable by the individual (all criteria for Voluntary Assisted Dying in Australia and New Zealand)'; however, by the time a person with HD reaches an advanced and terminal stage, their decision-making capacity will be so impaired as to make them ineligible for Voluntary Assisted Dying (VAD) under all current and proposed legislation in Australia and New Zealand. Juxtaposing this ineligibility for VAD is the widely reported high incidence of suicide and suicidal ideation amongst people affected by HD. While the strong association between depression and other psychiatric manifestations with suicide has been explored, this paper explores whether suicide

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can ever be deemed rational and suggests that ignoring this possibility may result in a paternalistic approach that fails to meet the needs of the patient. In addition, health professionals can be left inadequately prepared and morally compromised. Given that Advance Care Planning (ACP) can provide an opportunity to refuse all medical treatment, except palliative treatment, which could result in an earlier death in a context of intercurrent illness or accident, could early and robust ACP discussions provide an acceptable alternative to suicide and VAD? Suicidal ideation in HD can be viewed as arising from multifaceted matrices including the impact of genetic testing, knowledge of the expected disease progression, including future neurological deterioration, psychiatric history as well as personal and family histories. When viewed this way, rather than just in terms of rationality or pathology, we may be in a better position to provide the best care and support to people with HD, including through ACP. Nurses need to consider these ethical dilemmas and remain informed on the debates as they may be at the forefront of a moral conflict between a person proposing rational suicide or seeking to implement refusal of treatment in an advanced care directive (ACD), and they must be aware and consider how their professional obligations and personal opinions interact with this complex topic.

Overview of Huntington's Disease

Huntington's disease (HD) is an uncommon, autosomal dominant neurodegenerative disorder presenting with a triad of symptoms in the motor, cognitive and psychiatric domains (Nance et al., 2011). People with HD have a high symptom burden and are diagnosed at a younger age than people with Parkinson's Disease (El-Sourady et al., 2022), with symptom onset commonly occurring in the 40–50-year age range (Nance et al., 2011). Progression of symptoms tends to be slow and insidious, with death typically occurring 10-20 years after symptom onset (Langbehn, 2022). Prevalence of HD is cited at 5.70 per 100,000 in Australia (Pringsheim et al., 2012) although it is believed that the actual prevalence may be much higher. As an autosomal dominant disorder, each child of an affected parent has a 50% chance of inheriting the faulty gene. The disease trajectory in HD is complex and can vary significantly even between affected people in the same family (Macleod, Jury, & Anderson, 2017). The clinical course is characterized by a high symptom burden, significant stress on individuals and caregivers and gradual loss of independence (El-Sourady et al., 2022) and decision-making capacity (Ekkel et al., 2021). There is an elevated risk of suicidal ideation, suicide attempts and completed suicide in people with HD (Kachian et al., 2019). By the late stage, a person with HD will be unable to walk, communicate, eat or drink on their own, will have a severe cognitive impairment and may experience severe chorea or dystonia (Dellefield & Ferrini, 2011), which, for some, is perceived to be a future of intolerable suffering (Regan et al, 2018).

Voluntary Assisted Dying

In most states of Australia and in New Zealand legislation is in place, or is being introduced, to enable people who meet specific criteria to access Voluntary Assisted Dying (VAD).

Pre-manifest	**Early Stage**	**Mid Stage**	**Late Stage**
Known to carry the HD gene mutation but symptoms are not yet apparent	Largely functional: May continue to work, drive, handle money, and live independently.	Loss of ability to work or drive, manage their own finances or perform their own household chores. Can usually still manage to eat, dress, and undertake own personal hygiene	Require assistance in all activities of daily living.
Subtle changes in mood and cognition may be reported or observed	Symptoms may include: minor involuntary movements subtle loss of coordination, difficulty thinking through complex problems, possible depression, irritability, or disinhibition.	Chorea may be prominent. Difficulties with: swallowing, balance, increased risk of falls, weight loss and problem solving Likely depression, irritability and apathy	Chorea may be severe, but more often it is replaced by rigidity, dystonia, and bradykinesia. Generally non-verbal

Table 1. Stages of Huntington's Disease (Nance et al, 2011)

In Australia and New Zealand criteria that must be met include:

- being 18 years of age or over
- being a citizen or permanent resident in the relevant state or country (usually for at least 12 months before the request is made)
- having decision-making capacity in relation to voluntary assisted dying
- having an incurable disease, illness or medical condition that:
    - is advanced, progressive and is expected to cause their death within six months (or within 12 months for patients with a neurodegenerative medical condition)
    - is causing suffering to the person that cannot be relieved in a manner that the person considers tolerable (White et al, 2022).

While HD is a life limiting disease, with irreversible decline in function that some may view as intolerable; by the time a person with HD reaches an advanced and terminal stage, their decision-making capacity will be so impaired as to make them ineligible for VAD under all current and proposed legislation in Australia and New Zealand (White et al, 2022).

Vignette 1.

A woman in her mid-30's has just watched her father die with HD. She is gene positive for HD, but not symptomatic.

During a consultation she stated that after telling a friend of her diagnosis, the friend had said it must be important to plan for her future. She reported initially feeling quite confronted. She then stated 'I intend to use VAD'.

Do we tell her she will not be eligible for VAD?

After assessing her mood and undertaking a suicide risk assessment, it was deemed appropriate to. She became distressed and agitated when advised that VAD would not be an option under the current legislation. ACP was offered as an alternative option and she is considering this.

Suicide rates in HD

When George Huntington first described Huntington's disease in 1872, he remarked on the suicide rate. 'The tendency to insanity, and sometimes that form of insanity which leads to suicide, is marked' (Huntington, 1872). Most literature on HD refers to an increased rate of suicide and historically there has been considerable research on the topic. Honrath et al. (2018) report suicide as the leading cause of death in HD, after pneumonia. Early retrospective studies examining cause of death in people affected by HD demonstrated a higher rate of suicide amongst people with HD compared to the general community and suggested if anything, the suicide rates reported by Di Maio, Squitieri & Napolitano (1993), of 7.3% and Farrer, Opitz & Reynolds (1986) of 5.7% were an underestimate. Another retrospective study examining the deaths of people with HD in Hungary over a 77 year period revealed a suicide rate of 10.1% (Baliko, Csala & Czopf, 2004). In the most recent study, Alothman et al. (2022) examined 18 years of linked electronic medical records in the UK and concluded that people with HD were nine times more likely to die from suicide than people who do not have HD. They also described people with HD dying from suicide were much younger than those with HD who died of other causes. No relationship has been established between age of symptom onset and suicide or between stage of HD and suicide risk (Farrer, Opitz & Reynolds, 1986). In a systemic review in 2019, Kachian et al reported that 7-10% of people with HD attempt suicide at some stage in their life compared to a 1-3% rate of suicide attempts in the global population. The suicide rate in HD is also higher when compared to other progressive neurological conditions including motor neurone disease, Parkinson's disease and Alzheimer's disease (Kachian et al., 2019).

Suicidal Ideation

Suicidal ideation is reported by 20–30% of people with HD during their lifetime, including in pre-manifest individuals (Kachian et al., 2019), with rates varying between stages (Honrath et al, 2018), but present throughout the course of the disease (Kachian et al., 2019). Evidence for elevated risk of suicidal ideation in specific stages of HD is mixed (Kachian et al., 2019).

A higher incidence of suicidal ideation has been reported at the first appearance of HD symptoms, immediately prior to diagnosis (Di Maio, Squitieri & Napolitano, 1993; Paulsen et al, 2005) and again in stage two of HD, when independence is diminishing (Paulsen et al., 2005); however, both historical and current suicidal ideation rates reported are higher amongst people with symptomatic HD compared to those in the pre-manifest stage (Honrath et al., 2018). It is not clear the extent to which suicidal ideation is in itself a risk factor for suicide attempts or completed suicide (Hubers et al., 2013). Regular assessments of suicidal ideation are an essential part of good practice and are routinely conducted throughout the course of the disease. Suicide questionnaires are included as part of almost all research in HD and, of note, the frequently used Problem Behaviours Assessment rates suicidal ideation as 'questionable' if there is a plan for suicide at a later date when the disease is more severe (Kingma et al., 2008).

Risk factors for suicide in HD

While a higher rate of suicidal ideation and actual suicide amongst the HD community has been established, the contributing factors are less clear. As Farrer et al. (1986, p. 310) put it, '*it is unclear whether it represents a rational but extreme response to an intolerable situation or a manifestation of dementia*'. Psychiatric symptoms are a core feature of HD and are a robust risk factor for suicide (Alothman, 2022). Depression is identified as the major predictor for suicide both in the wider community and in HD (Kachian, 2018). Depression occurs in people with HD at twice the rate of the general population, with 50.3% having sought treatment and 40% identified as currently depressed (Paulsen et al, 2005). Further study into the underlying mechanisms of depression is required, given that the nature of depression and suicidality is poorly understood and it remains unclear whether depression is due to biological changes in the basal ganglia or due to anticipation of the pending disease (Paulsen et al., 2005) or other factors. A depressed mood is identified as predictive of suicidal attempts and completed suicide (Hubers et al., 2012) while a small relationship between suicidal ideation and depression has been noted (Hubers et al., 2013) which increases with higher alcohol use (Wettzel et al., 2011). Although depression appears strongly associated with HD suicidality, it does not appear to explain all suicidal ideation, which can still be detected in cohorts of non-depressed individuals with HD (Halpin, 2012). A relationship has been identified between suicidal ideation and aggression and irritability (Kachian, 2018), along with anxiety and psychosis in people with symptomatic HD (Honrath et al., 2018), whereas higher rates of apathy are associated with lower rates of suicidal ideation (Honrath et al., 2018). More rapid disease progression is correlated with increased suicidal ideation (Hubers et al., 2013). Sociodemographic variables do not appear to predict suicidal ideation in people with symptomatic HD, however older age and being single or unemployed were associated with a presence of suicidal ideation in premanifest individuals (Honrath et al., 2018).

Living within an HD family may be itself a risk factor. It is possible that watching family members progress through an incurable illness, while recognising this represents your own future, might instigate suicidal thoughts (Alothman, 2022). A familial disposition to

depression and suicide may be a factor (Epping et al., 2011), with an increased risk of suicide reported in families where one member has committed suicide (Di Maio et al., 1993; Farrer et al., 1986). An alternative perspective presented in a qualitative study of suicidality, concluded that the HD community did not connect suicide to depression or mental illness but instead saw it as a response to the realities of a deteriorating disease and concerns over future care (Halpin, 2012). Whilst the sample was small, it is worthy of consideration given its personal narratives and the themes that arose.

Rational Suicide

A clear link has been established between HD and suicide and between suicide and depression and key periods of suicide risk have been suggested. The focus of this paper now moves to perspectives regarding the act of suicide. Attitudes to suicide vary within the HD literature; however, the majority advance an argument of reversibility, with most studies recommending a need for increased vigilance and monitoring of suicidal ideation and risk, for example, '*suicide is one of the few potentially preventable causes of premature death in this disorder'* (Wetzel et al., 2011 p. 2). They did however conclude that the absence of psychiatric symptoms does not indicate lack of suicidal ideation, so it may not be fully preventable.

Vignette 2.

A woman in her late 20's with early-stage HD was referred to our service after an attempted suicide. She was unable to articulate what lead her to that suicide attempt or what she had been thinking at the time or in the lead up. She was commenced on antidepressants.

Ten years later her husband has watched her gradual decline. She can still walk, but with supervision, needs assistance with personal care, can talk but is very withdrawn and does not speak much. She is severely underweight, eats very little and has taken to staying in bed all day and never leaves the house.

Her husband, who is her medical treatment decision maker, believes she has 'given up' and that she is too exhausted to fight anymore. He resists suggestions to re-structure her day to set expectations that she gets up and eats. He is inclined to think she should just be allowed to die, which is what he believes she wants.

We insist on investigations to rule out other contributing factors and encourage him to accept a trial of up-titrating her antidepressants, which he agrees to.

Two months later she is getting up and following a structured routine, eating well and has gained some weight, is going out for walks (in the wheelchair) and socialising again.
They have smiles on their faces when we see them now.

Paulsen et al. (2005 p. 729) took this further suggesting the need to '*separate suicide associated with major depressive disorder from rational suicide in persons with a known terminal illness'*. Amongst the HD specific literature Halpin (2012) presents the most compelling arguments for accepting the possibility or concept of rational suicide, stating that to ignore it may result in paternalism, in inadequately prepared and morally compromised health professionals and pessimism and hopelessness amongst people with HD. Furfari et al. (2015) present a case study of a woman with HD treated in hospital with intubation and intensive care after a suicide attempt, who clearly demonstrated lack of coercion and no mental illness and who sought discharge to complete her suicide and end her suffering. She had previously documented an advance directive requesting no active treatment. The authors report the moral distress for all concerned and suggest that in this example the principle of beneficence might in fact be allowing relief of suffering (through death) in a situation where the woman had already identified and documented her condition and quality of life to be unbearable.

The strongest view against rational suicide is put forward by an eminent psychiatrist, in the HD field, Adam Rosenblatt,.

It cannot be overstressed that suicide in HD is a preventable manifestation of disease. The concept of a "rational" suicide in someone with HD, a person who dispassionately "chooses" to "take his own life" before symptoms become too advanced, is a myth. Suicide is devastating to the people left behind and increases the risk of suicide in the next generation. It must not be rationalised, romanticized, or accepted (Nance et al., 2011 p.68).

Physicians generally hold the most negative view of suicide and VAD because it opposes the norms and ethics of their practice that focuses on treating and curing (Halpin, 2012). By contrast a 70-80% acceptability rate for rational suicide amongst health professionals involved in end of life (EoL) care has been reported (Werth & Holdwick, 2000).

If it is acknowledged that suicide and depression are not synonymous and that some people with suicidal ideation do not have a mental illness, then it can be considered that suicide is not always an irrational decision (Cutcliffe & Links, 2008; Ho, 2014). The prevailing libertarian view in most Western cultures is that the individual owns their body and therefore, with an emphasis on autonomy, the individual can choose where and when to die (Cutcliffe & Links, 2008; Ho, 2014). This perspective was a significant driver for development of VAD legislation. Where access to VAD is not an option, suicide may be seen as the only alternative way to control the circumstances of one's death (Kresin et al., 2021).

The most widely cited author on rational suicide is a psychologist, James Werth. He developed criteria for rational suicide (see Table 2), a version of which is almost always cited in the literature on this topic and which provides a framework for evaluating whether a suicide is rational or not. Rational suicide can be defined as the taking of one's own life that is characterised by reason or by making sense to others. Essential characteristics of rational suicide include being free from psychological distress, having a realistic assessment of their life situation, contemplating the decision for some time and ideally involving the individual's significant others (Werth & Holdwick, 2000).

Criteria for Rational Suicide

The person considering suicide has an unremitting "hopeless" condition. Hopeless conditions include, but are not necessarily limited to, terminal illnesses, severe physical and/or psychological pain, physically or mentally debilitating and/or deteriorating conditions, or quality of life no longer acceptable to the individual.

The person makes the decision as a free choice (i.e., is not pressured by others to choose suicide).

The person has engaged in a sound decision-making process. This process should include the following:

- Consultation with a mental health professional who can make an assessment of mental competence (which would include the absence of treatable major depression)
- Non-impulsive consideration of all alternatives
- Consideration of the congruence of the act with one's personal values
- Consideration of the impact on significant others
- Consultation with objective others (e.g., medical and religious professionals) and with significant others

Table 2. Source: Werth & Holdwick, 2000

The opposing view on rational suicide is presented as a largely philosophical debate, tending to use examples of healthy people choosing suicide, in an endeavour to counter the viewpoints of the rational suicide proponents (Pilpel & Amsel, 2011). They suggest that, 'The 'rational suicide' proponents are in a dilemma: either accept there is nothing wrong with this suicide, or they must make certain substantive value judgments...' (Pilpel & Amsel 2011). However, exponents of rational suicide contend that it needs to be assessed on the merits of each individual case and may in fact be judged differently by different practitioners (Cutcliffe & Links, 2008; Ho, 2014).

From a Catholic and some other Christian perspectives, belief in the sanctity of human life means that neither VAD nor suicide are seen as acceptable means of ending life. This belief system sees life as a gift from God that is precious and should always be protected and can only be ended by God. Therefore, to prematurely and unnaturally end life is contrary to love of self and love for God. Some Catholic teachings also encourage embracing suffering, as a means of being better able to identify with the suffering of Christ.

Vignette 3.

A man in his late 60's with advanced HD has been living in a residential aged care facility for several years. He is completely estranged from his family, largely due to the behavioural changes that resulted from HD over the years. He has no contact with anyone outside of the aged care facility and his HD treating team. He decides that his life is no longer bearable and announces that he wants to die. He ceases eating and drinking. He is assessed by the aged mental health team psychiatrists on two separate occasions and is found not to be depressed but repeating a consistent desire to die because his quality of life is so poor. He is deemed to understand the consequences of refusing to eat and drink. He remains in the aged care facility and dies two weeks later, never having waivered in his refusal of food and fluids.

Is this rational suicide?

Should he have been admitted to a psychiatric unit as an involuntary patient and forcibly kept hydrated and fed?

Is there an alternative way to view this?

The intersection between VAD and suicide

People affected by HD and their families, often express an understanding and acceptance of their family member's suicide, relating it to prevention of further suffering and degeneration. Members of the HD community, while recognising that depression can be a related factor, generally see suicide for themselves or their family members as rational (Halpin, 2012). This is a stark contrast to the psychiatrist's view quoted earlier (Nance et al, 2011 p.68). Kresin et al (2021) postulate that access to VAD may reduce the incidence of suicide by giving people incentive to live longer, past the point of incapacity, secure in the knowledge that autonomy in death will be possible, even when they can no longer carry out the act themselves. They identify the perspective that VAD provides a safe, regulated medical procedure as opposed to the often violent and uncertain outcome of a suicide attempt and may therefore be seen as more acceptable. This argument has only recently appeared in the discourse on VAD in Australia and may have emerged due to a national focus on suicide prevention (Kresin et al, 2021).

In any discussion about rational suicide, VAD is perhaps the 'elephant in the room', with opponents viewing VAD as sanctioned suicide. Euthanasia laws in the Netherlands, which allow for preparation of VAD wishes in advance, have similar criteria to Werth's for rational suicide, in that any request must be voluntary and carefully thought through and that the person's condition must have no prospect of improvement, there are no logical treatment alternatives and that their suffering

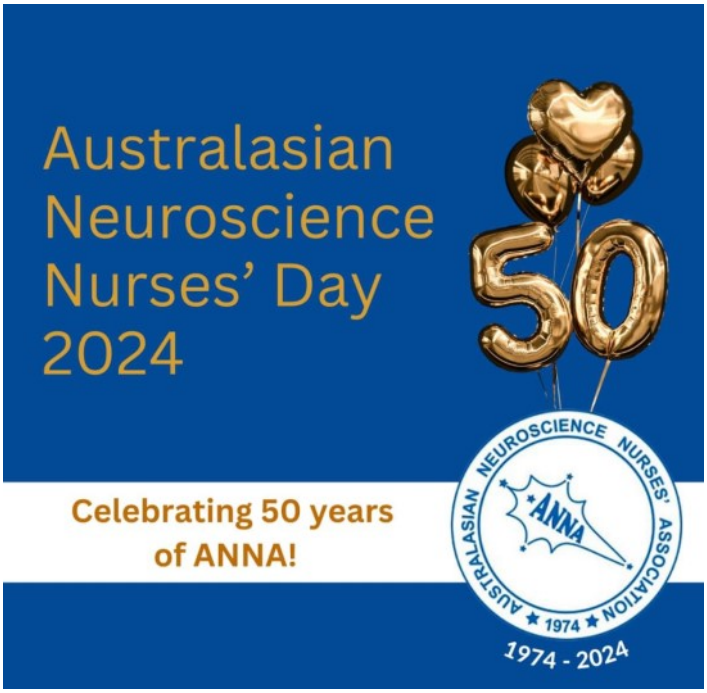
is unbearable (Booij et al., 2013). Figures from the Netherlands describe the granting of six to ten euthanasia requests a year for people with HD in a total of 50-60 deaths per year of people with HD (Booij et al., 2013). This computes to approximately 14% of all HD deaths compared to a rate of approximately 2% of all deaths in the Netherlands, occurring as a result of euthanasia. This figure is much lower than the suicidal ideation rates discussed earlier and prompts the question as to whether legalising euthanasia has any effect on the incidence of suicide in the HD population.

Because many people with HD clearly understand their future as HD progresses, it follows that many of them think about and plan for end of life (EoL) (Booij et al., 2013, Booij et al., 2014).

People with HD's views on VAD are strongly shaped by their clear understanding and experience of what their future with the disease will be like, and fear of loss of physical function and self-identity (Regan, Preston, Eccles, & Simpson, 2018). They therefore have a tendency to view VAD as desirable or necessary to alleviate suffering and where it is not legally available, suicide by other means might be considered a rational option (Regan et al., 2018) and may occur earlier in the disease trajectory, due to fear of being physically unable to complete this when they most desire it. People with HD, when considering EoL, tend to focus more on what they would perceive as unbearable, rather than on what could be done to best support them (Ekkel et al., 2021). Regan et al. (2018) found that people with HD emphasised their right to autonomy and perceived VAD as 'an act of kindness'. Given all of this, a focus on early and robust ACP may provide an alternative to contemplating suicide as the only other option and may also provide reassurance and psycho-education to meet the needs of people with HD who want to discuss EoL issues but are unsure how to (Regan et al., 2018).

Advance Care Planning

Two separate studies (Ekkel et al., 2021; Regan et al., 2018) showed that people with HD, similarly to the wider community (Bernard et al 2020), have varying approaches to the future, with some wanting to make



Vignette 4.

A man in his mid-30's is diagnosed with HD – the first in his family, so diagnosis was delayed. Over ten years his symptoms progressed, including cognitive changes, chorea and parkinsonism and depression, until he was starting to lose his independence.

His family were aware that he had been a longstanding supporter of euthanasia (this was prior to VAD legislation being introduced) and were supportive of his stance.

Fourteen years after diagnosis he had a suicide attempt resulting in a psychiatric inpatient stay, where his anti-depressants were up-titrated. Follow up assessment showed adequately managed depression, and he was not expressing suicidal ideation: however, he had another suicide attempt six months later which resulted in another psychiatric inpatient stay.

Within the next few months, he had a fall with head strike and was shown to have bilateral hygromas. These were managed conservatively, but CT scans two months later show they have increased in size. Surgery is recommended. He refused and stated this will be an easier way to die. His GP and neurosurgeon were not comfortable with his decision. He was advised the hygromas may remain stable and he may not die from them.

He was assessed by a psychiatrist – no signs of depression. He was assessed by a neuropsychologist and deemed competent to make his own decisions in relation to treatment and that he understood the possible outcome of no treatment. ACP discussions were held and he was clear he wanted no invasive medical interventions.

Four months later he had another fall with head strike and refused intervention, including CT scan. Two months later he had another suicide attempt and a psychiatric inpatient stay. Again, he was assessed as not depressed and still competent to make his own decisions.

Two months later he died alone from suicide. His family expressed relief that his suffering was over. Every suicide attempt used a different means. He never discussed his plans, just his desire to die.

plans and arrangements and others avoiding thinking about the future or not worrying about it yet. However, it appears that the majority of people with HD do have thoughts about EoL (Booij et al., 2014) and would like to discuss EoL care, but don't know how, when or with whom to have these conversations, or even what options are available (Regan et al., 2018). Familiarity with their own anticipated future, from witnessing other family members with HD, results in people with HD having more detailed and specific thoughts about EoL, but not necessarily thinking any more about EoL than other people (Booij et al., 2014). Health professionals have a legal, moral and professional obligation to discuss EoL issues with people with HD; however, in a scoping review of the literature only about a

Vignette 5.

Two people in their 50's, unknown to each other and who have just been diagnosed with HD both contemplate their future in a similar way. One was an educator and extremely active; a keen athlete. They both have early-stage HD – that would not be noticeable to anyone but those with a particular interest in HD. One, while participating in a research study, was asked if she had ever thought about suicide. She hesitated and clearly had an internal struggle, before deciding to answer, stating that she had every intention of taking her own life before the HD takes over. She stated however that she is now focussed on doing and achieving all the things she still wants to do with her life. She is clearly not depressed and is very much enjoying her life, working through the many items on her 'bucket list', involving significant travel. It is highly possible that she will take her own life and the planned timeline is when she can no longer live independently. This time is approaching.

The other is a businessman and father of three adult children. He requested ACP and stated that he has achieved all he wants to in life and could happily die now, feeling successful. He is adamant that he is not prepared to suffer the way he watched his mother suffer in end stage HD, in an aged care facility. He is very clear in his intent to take his own life, "before I am no longer able to". His instructional directive includes no CPR or medical interventions from this time forward. He has indicated he is involved with a voluntary dying service and has the means to end his life. He has discussed this in detail with his family who support his decision. He is on antidepressants and has periodically requested review and up-titration when he recognises his mood is lowered.

Both people have had neuropsychiatric assessment and are not depressed. On most measures in research on suicide, their statements would constitute "suicidal ideation". This creates a range of moral and ethical questions, particularly when considering a response to this potential 'rational suicide' in a country where VAD is not accessible to them. Does the health professional hope that as the time gets closer these patients choose not to discuss their plans or ideas? If they do mention it, will the health professional have an obligation to take some action to try and prevent this? Does this impact on the way the health care team can best support them? What if the health professional is uncomfortable with their plans? Can they maintain open and respectful conversation while disagreeing with the decision?

third of people with HD were found to have an advance care plan (El-Sourady, 2022).

The term Advance Care Plan or Advanced Care Directive is used to cover a variety of documents that people may use to express their values and preferences for care and treatment. ACP involves reflection, discussion, determination and documentation of an individual's values, goals and preferences in relation to their future medical treatment and care, to guide clinical decision making (Bernard et al., 2020). It usually also involves the appointment of a substitute decision maker in case they are unable to make decisions for themselves in the future (Ekkel et al., 2021). Ideally ACP is undertaken directly with the person with HD themselves, along with their family and future substitute decision makers, while they are still able to articulate their preferences (Keijzer-van Laarhoven et al., 2020). ACP can enhance quality of life in

Vignette 6.

A woman in her late-60's with mid stage HD lives alone in her own unit and is supported by National Disability Insurance Scheme and her adult children. She is starting to lose her independence, now unable to manage any of the domestic chores herself, such as cooking, cleaning or shopping. She is needing support to manage her medications reliably and has had a couple of episodes of getting lost when out of the house, needing to be assisted by neighbours. Discussions have started about whether she needs to move into aged care as she transitions into the advanced stage of HD.
She becomes acutely unwell and is taken to her closest emergency department, where she is found to have aspiration pneumonia.

She has an Advance Care Plan and her son is her Medical Treatment Decision Maker. According to her values statement, quality of life would no longer be bearable when she loses her independence. It indicated she would consider treatment for reversible conditions, but would not want CPR or ICU management.

The son advises that they have decided against treatment and have requested her transfer to our unit for palliative care. We discuss this in more detail to ensure he understands that with antibiotics she may recover and return to her previous baseline, where she was still enjoying aspects of life, and that without them, she will likely die. He reports that over the last few months she had been saying she had had enough and was tired of the battle with HD.
He believes she would not want interventions, apart from palliative care. She dies peacefully five days later, with her family around her.

advanced HD and provide reassurance to relatives (Ekkel et al, 2021) while reducing potential misunderstandings between health professionals and families (Lennard, 2018). A clearly written Advance Care Plan is likely to result in less aggressive interventions (Boersma, Miyasaki, Kutner, & Kluger, 2014; Gadoud & Johnson, 2015; The Victorian Department of Health, 2014) which could otherwise prolong dying and diminish quality of life.

ACP is a dynamic process and should be embedded as part of routine clinical practice, with consideration given to clinic visits dedicated just to ACP in order to overcome the most common barrier of insufficient time (Cooper, 2022). While some people may not want to undertake ACP, there is no way to predict those who will and those who will not, so health professionals must initiate these discussions (El-Sourady, 2022), encouraging people with HD to discuss and review their preferences at identified points along their illness trajectory, perhaps annually or in conjunction with functional changes (Vaughan & Kluger, 2018). People with HD report that they are generally able to return to living their regular daily lives once they have discussed ACP, without this causing lingering distress (Ekkel et al, 2021). Follow up ACP discussions and treatment options, in relation to emerging symptoms, are generally viewed as part of expected guidance from treating health professionals throughout the disease trajectory (Seeber et al, 2019).

ACP should include developing a values directive, which describes a person's views, values and preferences to guide alternative decision makers and doctors when needing to make medical decisions for them. An instructional directive is an optional component, is legally binding, and should unambiguously outline specific treatments that a person consents to or refuses and the specific circumstances under which they would want or not want those treatments (The Victorian Department of Health, 2014). The full implication of stated preferences and potential scenarios where these might be applied, should be discussed with the individual as part of the process. ACP for a person with HD should include but is not limited to : appointing substitute decision makers and powers of attorney, enduring guardian, decisions regarding resuscitation, types of treatment desired or refusal of treatment (such as antibiotics, ventilation, intensive care interventions), decisions regarding hospitalisation and preferred place of death and tube feeding (Hosken, 2019). ACP discussions and documentation should include treatments both now and into the future, with clearly identified stages a person may like to refuse specific treatments (Hosken, 2019). In the author's clinical experience, people with HD are often not aware that they can refuse medical treatments, such as antibiotics in the event of pneumonia for example, which may result in their death in the setting of an intercurrent illness. Within the author's service, a survey of people with people with HD and their caregivers, regarding their EoL fears and wishes, found that participants overwhelmingly wanted HD specific advance care planning information to help support them in this process.

An argument is sometimes made that ACP is flawed in that it requires individuals to anticipate what their future may hold, a state they have not previously experienced, without knowing how they may adapt to their condition in the future or if their values might change (Lennard, 2018). However, most people with

Vignette 7.

A woman in her late 50's with advanced stage HD lives with her husband and 2 teenage children. She is dependent on others for all aspects of her life and is no longer able to communicate verbally. She does not have an advanced care directive.

She develops aspiration pneumonia and has a traumatic experience in a tertiary hospital, where typical protocols mean she is made nil orally and the husband feels pressured to consider a naso-gastric tube or PEG, which he resists. He feels that he is being reprimanded by staff for 'force feeding her' as the clinical team have assessed having oral intake high risk. She is treated with antibiotics and transferred to our unit where oral feeding is recommenced while monitoring closely for re-feeding syndrome. She recovers.

Discussions are held with the family regarding the likelihood of further aspiration pneumonia, given her progressive dysphagia. Advance care planning discussions are held, with no outcomes decided upon at that time. If they had clear wishes these could have been written into a 'statement of choices' document.

Two months later she becomes acutely unwell again. The family choose not to seek medical intervention and she dies at home.

HD have lived experience of the disease, having witnessed often multiple family members EoL experiences and are therefore in a unique position to understand their likely future and predict their future self (Regan et al 2018). ACP discussions often consist of people with HD seeking reassurance that they will not have to endure the disease until the very end (Ekkel et al, 2021), like they have witnessed in other family members.

Autonomy is promoted through ACP and individuals may feel reassured by the knowledge that their preferences for EoL care will be followed and that the burden for decision making will not be placed on their families (Booij, Engberts, Rödig, Tibben, & Roos, 2013; Huntington Society Canada, 2016; Regan, Preston, Eccles, & Simpson, 2018). However, an ethical dilemma may arise, if an individual's condition has progressed so that they are no longer able to express their viewpoint and others around them consider they are no longer aware of or committed to their previous beliefs expressed in an advance care plan and appear to be currently able to derive enjoyment from life (Keijzer-van Laarhoven et al, 2020; Lennard 2018). In this situation questions may arise as to whether a previous autonomous decision should take precedence over protecting their current presumed wishes (Keijzer-van Laarhoven et al, 2020, Lennard 2018) and will require the treating team and substitute decision maker to interpret the values documented in the ACP. In relation to HD, there is a unique future risk associated with not adhering to the ACP wishes and directives. Given that the nurse-patient relationship is based on a foundation of trust and honesty (Snir et al, 2022), where a family

member may be also a person with HD themselves, lack of adherence to, or respect for, a previous ACP decision may undermine the very nature of that relationship and take away their hope for their own future EoL care. While no link has been found in the literature, the question must be asked as to whether disregard for the Advance Care Plan could then increase the risk of suicide in other family members with HD, as a means to assert some control over their own future.

Implications for Health Professionals

The clearly established links between HD and depression, and between depression and suicide compel health professionals to undertake regular assessments of depression and suicidal ideation with their patients with HD, and ensure that they are offered medication and therapeutic interventions, such as counselling to treat depression (Nance et al, 2011). Nurses are closely involved with people with HD and their families, so they may be the first one to whom suicidal ideation is expressed or the question of VAD is raised (Snir et al, 2022). The nurse should engage in empathic but exploratory discussion with the individual to gain an understanding of the factors contributing to this thinking and questioning, as well as passing this information on to other members of the team (Snir, 2022). This may be the trigger for initiation of ACP discussions. Routinely discussing end of life wishes and assisting patients in ACP is an important part of providing care to people with HD, to enable them to maintain autonomy and quality of life, given they will inevitably have considered EoL at some stage in the disease trajectory (Booij et al, 2014).

Identifying how to respond to rational suicide is a greater challenge, given that most health guidelines, practices and policies are written on a premise of suicide being driven by a psychiatric disorder and as such health professionals are charged with identifying potential risks and a duty of care to try to prevent suicide (Cutliffe & Links, 2008; Ho, 2014). Nurses working with a person with HD who expresses their intent to take their own life are faced with an ethical dilemma, potentially causing moral distress, and need to manage the discomfort and dissonance arising from this. The nurse may hold a belief that the individual is rational and accepts their right to autonomy, yet they may be working within a policy and an organisation that requires them to keep that person physically safe and do all in their scope to prevent their suicide (Cutliffe & Links, 2008, Rich & Butts, 2004). '*Moral progress in nursing necessitates that nurses ponder these ethical uncertainties in supporting or opposing intervention with patients who are contemplating rational suicide'* (Rich & Butts, 2004 p.277), otherwise they may be unprepared psychologically and professionally when faced with this situation (Rich & Butts, 2004). The literature draws out this dilemma however it offers no solutions, with the one exception a proposed model for ongoing research, training and lobbying for policy change on a larger scale (Werth & Holdwick, 2000).

Conclusion

A higher incidence of suicide amongst people affected by HD and the strong association between depression and suicide has been demonstrated and this knowledge is at the forefront of good practice in working with people with HD. Given that VAD is not available for people with HD, awareness of the possibility of rational suicide is important. Whilst

there is a significant body of writing on the topic of rational suicide, this remains controversial, posing unresolved ethical dilemmas for practitioners working with people with HD who are contemplating suicide. Given the close relationship that may exist between nurses and people with HD and their families, they may be the first person with whom these suicidal thoughts or questions of VAD are raised. This provides an opportunity and perhaps an imperative to introduce the concept of ACP, to enable the individual to assert some autonomy over their future, which may provide an acceptable alternative to suicide and VAD. If suicide is viewed as arising from multifaceted matrices including the impact of genetic testing, knowledge of the expected disease progression, neurological deterioration, psychiatric history and personal and family histories rather than just in terms of rationality or pathology we may be in a better position to provide the best care and support to a person with HD (Halpin, 2012), including through ACP. Awareness of these issues, and the ethical dilemmas that may arise from them, is essential for nurses, to reduce the moral distress they may otherwise experience when faced with a conflict between their professional obligations and their own personal views and beliefs.

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