Introduction:
While palliative care has long been considered an essential component of care for people with cancer, there has also been exploration of the role it might play for people with disabling and life limiting progressive neurological diseases (PND) (Travers, Jones, & Nicol, 2007). Despite much rhetoric and many reports suggesting that palliative care should be accessible and available for all people with life limiting illnesses there has been little evidence of change in practice (Booth, Fallon, & Hollis, 2016). People with PNDs, including Huntington’s Disease (HD), which will be the focus of this report, frequently live for many years, progressively becoming more physically and cognitively disabled and enduring a decreasing quality of life (Gofton, Jog, & Schulz, 2009). While there is little written specifically about end stage HD, an improved understanding of this stage is essential to ensure existing health services are able to support people with HD in preparing for their end of life (EoL), earlier in their disease trajectory. Recent literature demonstrates a growing interest and awareness of incorporating a palliative approach into neurological care (Oliver et al., 2016). This paper will explore EoL issues and provide recommendations regarding advance care plans for people with HD.

Method:
A literature search was conducted in the electronic databases using the following search terms: ‘Huntington’s Disease’ AND ‘end of life OR palliative care OR late stage OR advanced’. Forty two articles were identified for review.

Discussion: People with Huntington’s Disease think about their end of life wishes, but often do not discuss these with their doctors. Incorporation of an early palliative approach into a multi-disciplinary neurology service is widely regarded as best practice and should include the early introduction of end of life planning, however many clinicians are not comfortable with discussing end of life care. This paper synthesises the available information and makes recommendations regarding advance care plans for people with HD.

Conclusion: Initiation of a palliative approach early in Huntington’s Disease has the potential to improve management of symptoms, increase the likelihood of advance care plans being developed and improve the overall quality of life throughout the duration of the illness; therefore, all clinicians working with people with Huntington’s Disease should have an understanding of palliative approaches.

Key Words
Huntington’s Disease, Palliative Care, Advance Care Planning, End-of-Life care
Background: Huntington’s disease:

Huntington’s disease is a rare, genetic, neurodegenerative disorder that clinically presents as a triad of symptoms occurring in the motor, cognitive and psychiatric domains (Nance et al., 2011). Internationally, prevalence of HD is estimated at 6/100,000 (Oliver et al., 2016). First symptoms commonly appear between 35-50 years of age with slow and insidious deterioration culminating in severe incapacity prior to death, which is usually 10-20 years after symptom onset (Nance et al., 2011). Despite differences in age of death and length of illness reported in the literature, HD does consistently result in a reduced life span. The advanced stage of HD has not been extensively studied, resulting in limited knowledge regarding best practice EoL care for people with HD and their families (Rodrigues et al., 2017).

Late Stage/Advanced HD:

The disease trajectory in HD is complex and varies strikingly, even between affected people in the same family (Macleod et al., 2017), making it difficult to know when to instigate an EoL approach (Gofton et al., 2009). Some suggest that the beginning of the ‘end of life’ in HD is the time when the individual is no longer capable of independent living (Dubinsky, 2004), a stage also referred to as ‘late stage HD’ or ‘advanced HD’, which may last more than 10 years. Others, however, suggest EoL is further into the late stage, when a person is bedbound, unable to communicate, eat or drink on their own and display severe chorea or extreme dystonia (Dellefield & Ferrini, 2011; Huntington Society Canada, 2016).

Terminal stage in HD:

There is very little written about the terminal stage of HD and how to recognise it. The lack of evidence based criteria for determining the terminal phase and specifically the six month prognosis in HD (Johnson, Frank, Mendlik, & Casarett, 2018), is a significant factor in people with HD having difficulties accessing palliative care services. Tarolli, Chesire, and Biglan (2017) suggest the terminal phase in HD tends to be unpredictable, with significant variation and fluctuation between patients. Some people with HD can appear to be in a terminal phase, but then recover and they may do this several times (Tarolli et al., 2017). While there remains difficulty in identifying the terminal phase of the disease, people with HD may also die unexpectedly, leaving families unprepared (Hussain, Adams, Algar, & Campbell, 2014).

Symptoms that might indicate the approach of the terminal stage include; inability to walk, speak and eat, weight loss (Huntington Society Canada, 2016; Klager et al., 2008), new-onset screaming occurring in an otherwise mute and bed bound individual (Nance, 2012;

Further relevant articles were found from the reference lists of selected papers. Article titles and abstracts were reviewed and papers were included if they made reference to palliative care and PND, including HD, even if these were not the main focus of the article. Articles were excluded if their focus was Juvenile HD or another PND such as Motor Neurone Disease, because of their vastly different disease trajectories to HD.

A total of forty-two articles have been included, 39 from scholarly journals, one report from a working party, one fact sheet retrieved from an HD organisation’s website and one chapter from a management guideline available on the internet. Of the 39 journal articles, 15 related broadly to palliative care in neurodegenerative disorders, 16 to HD and palliative care, five were general articles on HD that mentioned terminal or end of life care and three were primarily focussed on euthanasia and HD. The majority of articles (22) were ‘expert opinion’ review pieces, there were three literature reviews, one book review, seven retrospective cohort studies, one report on a current cohort of patients and eight papers with mixed method qualitative research. The expert opinion articles were written from the perspective of neurology (13), palliative care (4), gastroenterology (1), neuro-palliative services (3) and other (3).

Major themes arising from the literature that will be explored in this paper include; definitions of palliative care, service delivery models for HD, barriers to accessing palliative care, advance care directives, tube feeding and end of life care.

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Nance et al., 2011), isolated episodic fevers and deep sleeping for most of a 24 hour period (Huntington Society Canada, 2016; Mowatowitz & Marier, 2001). Identified triggers have been suggested for the recognition of end of life for patients with PND generally in a retrospective cohort study (Hussain et al., 2014); however, these require further validation and it should be noted that only four people with HD were included in the study (Hussain et al., 2014; Oliver et al., 2016). This could be a useful area of further research. Johnson et al. (2018) identified that the most common reported symptom for people with HD accessing a hospice service was pain, which is not otherwise a commonly reported symptom associated with HD, while Mestre and Shannon (2017) report that pain was present in less than 10% of a cohort of people with advanced HD living in a residential facility. Pain is otherwise scarcely mentioned in the literature reviewed.

**Causes of Death:**

Cause of death in HD is typically due to medical complications (Huntington Society Canada, 2016; Nance et al., 2011), most commonly pneumonia due to aspiration (Huntington Society Canada, 2016; Macleod et al., 2017), but possibly including dehydration, malnutrition or choking (Huntington Society Canada, 2016). Rodrigues et al. (2017) found that the most frequent cause of death was pneumonia (19%), followed by other infections, then suicide and cancer, with cause of death being undetermined in 23% of deaths. Solberg et al. (2018) similarly found respiratory diseases, including pneumonia, were the most common cause of death (44%), followed by cardiovascular disease, HD itself, injuries and suicide, while Booji, Rödig, Engberts, Tibben, and Roos (2013) report pneumonia as the primary cause, followed by suicide.

People with late stage HD may have a series of sudden deteriorations or acute illnesses such as aspiration and it can be difficult to predict if and when they will respond to treatment and therefore to accurately identify the dying phase (Hussain et al., 2014; Wilson, Seymour, & Aubeeluck, 2011). This can inadvertently lead to someone’s end of life wishes being breached, particularly if a decision is made to seek hospital care (Wilson et al., 2011). The majority of people with HD admitted to hospitals via emergency departments are discharged to long term care facilities (Simpson, 2007). A good death is widely considered to be pain free, calm and peaceful, in a familiar place, surrounded by familiar people, whether staff or family and where individual preferences are known and respected (Huntington Society Canada, 2016; McClinton, 2010; Wilson et al., 2011). People with HD typically spend their final years in care facilities, most commonly residential aged care (Tarolli et al., 2017) and the literature shows that the most common place of death is hospital or care facility, which differs from other hospice patients who are more likely to die at home (Johnson et al., 2018; Rodrigues et al., 2017). Mestre and Shannon (2017) suggest that stronger models of palliative care focussing on community support may enable people with HD to remain at home longer as their condition advances.

There are no published guidelines regarding specific treatments to be used in the terminal stage of HD; however, Nance et al. (2011) recommend the following: consider ceasing prophylactic medications and treatments for conditions other than HD except those used for comfort, but continue any psychotropic medications which may be necessary for psychological comfort and control of chorea. In addition they advise the use of long term narcotics, muscle relaxants and anti-anxiety medications, along with anti-cholinergic agents if drooling is excessive (Nance et al., 2011).

**Care Delivery in Late Stage HD:**

It can be argued that all management for HD, from diagnosis to death, is palliative because there are currently no disease altering treatments available (Dellefield & Ferrini, 2011). However there is a common misconception that palliative care is synonymous with end of life care and is primarily related to diagnoses of cancer (Boersma, Miyasaki, Kutner, & Kluger, 2014; Provinciali et al., 2016). It is surprising to note many of the articles reviewed that were related to palliative care contained no definitions of palliative care.

The most commonly cited definition in the remaining papers is that of the World Health Organisation.

There is widespread agreement within the literature identifying barriers to people with PND accessing specialist palliative care services leading to their significant under-representation in these services (Rosenwax, Spilsbury, McNamara, & Semmens, 2016). Palliative care services are generally not funded or equipped to provide support to someone throughout a 20 year illness, so alternative care delivery models must be considered. The therapeutic goals of management for HD align with those of palliative care; mitigating symptoms, enhancing quality of life and responding to the often complex needs of patients and their families (Klager et al., 2008; Wilson et al., 2011). This can be extrapolated as the ‘palliative approach’ which has the support of many (Gadoud & Johnson, 2015; Oliver et al., 2016; Provinciali et al., 2016); combining active treatments specific to the disease with the holistic assessment and response to patients’ needs while providing support to carers, within a philosophy of an open attitude towards death and dying. The effectiveness of the palliative approach in HD will depend on the skills and expertise of clinicians in adapting the concept of palliative care to the idiosyncrasies of HD (Provinciali et al., 2016) and this takes on additional importance in the late stage. Vaughan and Kluger (2018) suggest that a palliative approach may in fact improve quality of life for people with HD; however, they suggest this approach remains in early developmental stages and requires significantly more work to address the barriers. Tarolli et al. (2017) highlight the challenges of specialist neurology services not necessarily being well equipped to manage the non-neurological end of life issues, including advance care planning and psychosocial support, while palliative care services are generally unfamiliar with the specific needs of people with HD. The most effective care may be offered by a multi-disciplinary team, including both neurology and palliative care specialists (Mestre & Shannon, 2017).

Onset of late stage is the time where some consider palliative care should commence (Klager et al., 2008). Others argue it should commence much earlier, for example soon after the time of diagnosis (Dubinsky, 2004; Provinciali et al., 2016), while some, moreover, implore for the incorporation of a palliative approach in the early stages to relieve physical and psychological distress emanating from diagnosis (Kristjanson, Toye, & Dawson, 2003; Provinciali et al., 2016). What is clear is the need to commence a palliative approach early enough in the disease to ensure the person has capacity to make decisions and choices regarding their end of life care (Nance et al., 2011; Travers et al., 2007). If implementation of palliative care is left until the terminal phase, the period of greatest suffering for the patient may be over and with it the opportunity of palliative care to reduce suffering (Booth et al., 2016).

It is interesting to note that while the articles reviewed that were specific to management of HD (Nance, 2012; Nance et al., 2011; Novak & Tabrizi, 2010) did not mention palliative care or a palliative approach, their overarching philosophy and tone indicate that they apply a palliative approach to their management. ‘The aim of treatment is to manage symptoms and improve quality of life’ (Novak & Tabrizi, 2010 p.5). ‘Thoughtful care can reduce discomfort and improve the experience of the patient and family confronting this difficult neurodegenerative disorder’ (Nance, 2012, p. 364) ‘Goals of care remain the same....: reduce the burden of symptoms, maximise function and optimize quality of life’ (Nance et al., 2011, p. 111). Oliver et al. (2016) concur that most guidelines for care of specific neurodegenerative disorders do not provide an overt description of the role of palliative care, perhaps reflecting a level of discomfort with addressing EoL concerns.

In contrast to most medical conditions, where the burden of symptoms increases as the disease progresses, HD can present with severely debilitating physical and psychiatric symptoms early in the disease, thus highlighting the need for an early palliative approach (Tarolli et al., 2017). Whether these symptoms are best managed through a palliative care service or a multi-disciplinary neurology or specialist HD service remains unclear in the literature. Existing models of care generally result in few people with HD accessing specialist palliative care services even as they approach EoL (Vaughan & Kluger, 2018). Care throughout the disease, up to and including EoL is therefore largely provided by multi-disciplinary neurology teams using a palliative approach. Although there is limited supporting evidence, Oliver et al. (2016) conclude that a palliative approach is desirable during the progression of the disease and that specialist palliative care may have less contribution until the condition advances. Specialist palliative care is provided by services who assume responsibility for physical, psychological, social and spiritual care of a dying person (Boersma et al., 2014; Travers et al., 2007) and should be referred to when patients or carers have complicated needs beyond the capabilities of their treating team (Boersma et al., 2014; Brown & Sutton,
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therefore vital for clinicians to facilitate open community (Kavanaugh et al., 2016). It is both within their families and in the broader context around the diagnosis and illness itself, (Booij et al., 2014; Kavanaugh, Noh, & Zhang, 2016). Many individuals and families affected by HD experience stigma and secrecy around the diagnosis and illness itself, both within their families and in the broader community (Kavanaugh et al., 2016). It is therefore vital for clinicians to facilitate open discussions around EoL issues to try and minimise the distress and reduce the burden of potentially having to make EoL decisions for a family member without ever having had a discussion about their wishes (Kavanaugh et al., 2016).

End of Life Planning:

Advance Care Plans- In Victoria the term Advance Care Plan is used to cover a variety of documents that people may use to express their values and preferences for care and treatment (The Victorian Department of Health, 2014) whereas others may be more familiar with the term Advanced Care Directive. Advance Care Planning is identified in The Victorian Health Priorities Framework 2012–2022 as an essential growth area in order to enhance the health experience of every Victorian (The Victorian Department of Health, 2014) and involves discussion and documentation of an individual’s values, beliefs and preferences in relation to their future health and care needs to guide clinical decision making. It also involves the appointment of a substitute decision maker in case they are unable to make those decisions for themselves in the future (Hagen et al., 2015; The Victorian Department of Health, 2014).

Early incorporation of a palliative approach has the potential to improve EoL planning, promote the development of ACP and improve the quality of life for people with HD (Oliver et al., 2016; Provinciali et al., 2016). Given the long illness trajectory and the inevitability of the terminal phase there is ample opportunity for ACP to be made (Dawson et al., 2004; Nance et al., 2011). There is some contention in the literature as to when EoL planning should commence for people with HD. There is general consensus that it should be initiated prior to cognitive decline and loss of communication skills and the phrase ‘as early as possible’ is frequently used (Oliver et al., 2016; Provinciali et al., 2016). Others suggest it should be at the commencement of late stage or before institutionalisation occurs (Gadoud & Johnson, 2015; Wilson et al., 2011), while Booth et al. (2016) suggest planning should occur when good symptom control has been achieved. Ideally it should be initiated as early as possible in the illness and certainly before cognitive decline and deterioration of communication; however with predictive gene testing, many people with HD may receive a ‘diagnosis’ years before symptom onset and initiating EoL planning at this stage is contentious (Tarolli et al., 2017).

Impact of the familial nature of HD:

Memories of how relatives lived their final days and months and how they died will increase the likelihood of people with HD having thoughts about EoL decisions and will often result in individuals not wanting to prolong the advanced stage of the disease (Booij, Tibben, Engberts, Marinus, & Roos, 2014; Hubers et al., 2016). Booij et al. (2014) concluded that although people affected by HD do not think about EoL issues any more or less than other people, their EoL thoughts are more detailed because of their familiarity with their own anticipated future as seen through the journey of relatives. Macleod et al. (2017) reflect that families may be scarred by generations of witnessing debilitating disease and difficult deaths. Regan, Preston, Eccles, and Simpson (2018), in a qualitative study on views of people with HD on assisted dying, report that most people’s views were strongly influenced by having witnessed the effect of HD on their family members; although, this did not mean there was a consensus viewpoint amongst family members. The anticipated loss of dignity in the future is a major instigator of EoL thinking (Booij et al., 2014).

The familial nature of HD increases the need to ensure that EoL planning results in effective and respectful care as this can have a long term influence on how caregivers make decisions about their own end of life care (Booij et al., 2014; Kavanaugh, Noh, & Zhang, 2016). Many individuals and families affected by HD experience stigma and secrecy around the diagnosis and illness itself, both within their families and in the broader community (Kavanaugh et al., 2016). It is therefore vital for clinicians to facilitate open discussions around EoL issues to try and minimise the distress and reduce the burden of potentially having to make EoL decisions for a family member without ever having had a discussion about their wishes (Kavanaugh et al., 2016).
Thoughtfully prepared ACP can enhance quality of life during late stage HD and provide reassurance to relatives (Simpson, 2007). Written ACP tend to result in less aggressive treatments (Boersma et al., 2014; Gadoud & Johnson, 2015; The Victorian Department of Health, 2014) which could otherwise prolong dying and diminish quality of life (Huntington Society Canada, 2016). Autonomy is promoted through ACP and the individual 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 et al., 2018), leading to greater satisfaction with their care overall (Boersma et al., 2014; The Victorian Department of Health, 2014). Given that advance care planning is a dynamic process, once embedded as part of routine clinical practice, people with HD should be encouraged to discuss and review their preferences at identified points along their illness trajectory, perhaps annually in conjunction with functional changes (Vaughan & Kluger, 2018).

**Talking about EoL** wishes with an HD patient is part of the legal, professional and moral responsibility of the physician (Booij, Engberts, et al., 2013) and is desired by most people affected by HD; however, they often do not know how, or when, to discuss this, or even what options are available for EoL planning (Regan et al., 2018). Only 31-38% of people with HD have documented ACP (Booij et al., 2014; Downing et al., 2018), which is low given the prolonged and severe disease trajectory.

**Barriers to EoL Planning** - Many barriers to EoL planning have been identified in the literature. Reluctance to undertake ACP is a significant barrier, with Boersma et al. (2014) suggesting that neurologists have a propensity to broach the subject too late in the illness, or not initiate the conversations at all (Booij et al., 2014), while Tarolli et al. (2017) suggest a lack of comfort, time constraints and lack of a defined process contribute to this. Booij et al. (2014) found that less than half the people with HD who had EoL wishes had discussed these with their doctor, potentially leading to their wishes not being heard and treatment being provided that is not in keeping with their wishes, with a significant number not being aware that an ACP could be drawn up (Booij, Rödig, et al., 2013). Other barriers to ACP include lack of engagement or understanding of the process in the community (Hagen et al., 2015), patients’ denial and fear of the future (Dawson et al., 2004; Klager et al., 2008), a sense that planning EoL is equated with giving up hope (Booij, Rödig, et al., 2013), family conflict possibly arising from feelings of guilt (Klager et al., 2008), and cognitive and communication decline resulting in reduced capacity to participate (Klager et al., 2008). Systemic barriers include insufficient time and competing priorities, inexperiance or personal discomfort (Blackford & Street, 2013; Hagen et al., 2015), insufficient training and the belief that it is the responsibility of another health professional (Blackford & Street, 2013). Clinicians may feel that ACP discussions may lead to patient distress by forcing them to confront their inevitable mortality (Tarolli et al., 2017). However Booij, Engberts, et al. (2013) suggest that the discussions can result in peace of mind and reduce time spent worrying about it, while Hubers et al. (2016) found it may actually reduce suicidal ideation, although this was in a context where euthanasia is legally permissible.

People with HD have tended to feel excluded from current debates regarding assisted dying as they recognise that in most jurisdictions it does not (or will not) allow for the complexities of conditions such as HD where the final stages involve severe cognitive impairment and difficulty in identifying a six month prognosis (Regan et al., 2018). They, however, may view euthanasia as desirable or necessary to relieve suffering and where it is not legally available, suicide by other means is considered a rational option by some (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. As one of the few conditions that can be identified before its onset and where individuals have a clear understanding and experience of what their future with the disease will be like, people with HD tend to have a significant interest in assisted dying (Booij, Engberts, et al., 2013; Booij et al., 2014; Regan et al., 2018).

**Facilitators of EoL Planning** - Education regarding ACP, along with resources to increase skills and confidence of clinicians, are critical strategies (Blackford & Street, 2013) to embed EoL planning in practice. Triggers for starting conversations about ACP can be identified and shared with all clinicians as a prompt to assist in initiating conversations and further develop required communication skills (Blackford & Street, 2013). Whilst the majority of the articles reviewed were written by physicians and discussed the responsibilities of physicians for initiating EoL discus-
sions, it has also been identified that care for people with HD is best managed in a multi-disciplinary team (MDT). All clinicians in the MDT could be involved in contributing to EoL discussions, particularly around their specific area of expertise (The Victorian Department of Health, 2014), thus reducing the expectation on the physician to initiate the discussion. The role of a case co-ordinator in the MDT for people with HD has been well established (Macleod et al., 2017) and perhaps responsibility for initiation of EoL discussions sits within this role. Booij et al. (2014) recommend that every person with HD should be asked early in the course of their disease ‘their fears, their wishes and thoughts for the future’ and given that ACP is best viewed as a process not an event (Hagen et al., 2015), these questions should be repeated at regular intervals. In a qualitative study, people with HD were found to be aware of the possibility of changing their views regarding EoL decisions (Booij, Rödig, et al., 2013) and confidence in knowing they would be asked to review their ACP regularly may overcome a reluctance to plan due to sensitivity to their possible response shift.

Two additional papers reviewed, that were not included in this report, presented tools developed to examine the end of life concerns and end of life planning for people with HD (Carlozzi et al., 2018; Carlozzi et al., 2016). The use of these tools may assist in addressing clinicians reported difficulties in initiating conversations about EoL planning.

**Recommended Considerations for ACP and Implications for Practice:**

Ideally ACP for a person with HD should include: appointing substitute decision makers and powers of attorney, 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, tube feeding and brain donation (Hussain, Adams, & Campbell, 2013; Klager et al., 2008) as well as concerns, goals and fears of both the individual with HD and their family (Tarolli et al., 2017). Discussions and documentation should include treatments both now and into the future, with clearly identified stages a person may like to refuse specific treatments. It is important that ACP stating the individual’s wishes in relation to tube feeding be completed earlier in the illness when they have decision making capacity (Gadoud & Johnson, 2015; Simpson, 2007) and discussions should be had regarding indications for withdrawal of tube feeding if one is to be inserted (Macleod et al., 2017). Refer to discussion box.

Loss of the sense of self, rather than the severity of the disease symptoms was found to be a significant factor in anticipated reduced quality of life and fears for EoL (Booij et al., 2014; Regan et al., 2016) and may be partially addressed through the development of ‘booklets’ that describe the person and reflect their life and their values before and after onset of HD symptoms (Simpson, 2007). These ‘booklets’ can be powerful reminders of personhood, conversation starters and memory prompts to assist in developing rapport between the person with HD and carers thereby enhancing quality of life.

**Conclusion:**

The current literature on EoL planning in HD primarily reflects expert opinion, although more recently has included reports on the thoughts and feelings of people with HD and describes an increasing interest in the early introduction of a palliative approach into multi-disciplinary neurological care (Gofton et al., 2009; Oliver et al., 2016). Discussion regarding palliative care in broader PND literature continues to develop, however palliative care in HD remains under studied. HD provides a unique opportunity to investigate long term palliative care support for a younger cohort who tend to be institutionalised earlier and for longer periods of time than other people (Moskowitz & Marder, 2001; Nance & Sanders, 1996). Initiation of a palliative approach early in HD has the potential to improve management of symptoms, increase the likelihood of ACP being developed and improve the overall quality of life throughout the duration of the illness (Boersma et al., 2014; Dawson et al., 2004; Gofton et al., 2009; Hussain et al., 2013; Kristjanson et al., 2003; Oliver et al., 2016) and therefore all clinicians in a MDT working with people with HD should have an understanding of palliative approaches (Travers et al., 2007).

Further research is required to see if this has a direct impact on patient care and to produce evidence based data to support this approach (Gofton et al., 2009; Oliver et al., 2016). Additional research is recommended into the development and validation of prognostic predictors and outcome measures (Boersma et al., 2014; Dubinsky, 2004) as well as into the EoL concerns and wishes for people with HD. This paper has synthesised the available literature and concluded that implementing a palliative approach early in the management of Huntington’s Disease is
recommended. All clinicians working with people with Huntington’s Disease should have an understanding of palliative approaches, and the care co-ordinator might be best placed to initiate EoL discussions. Recommendations regarding required components in an ACP for a person with HD have been identified as: appointing substitute decision makers, 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, tube feeding and brain donation as well as concerns, goals and fears of both the individual with HD and their family.

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life: who is still missing out a decade on?


Practice Point – Tube Feeding in HD

Dysphagia, or swallowing difficulties, is a significant problem in HD, resulting in difficulty ensuring adequate nutritional intake to maintain weight (Novak & Tabrizi, 2010) and can have serious sequelae. Dysphagia is the most common cause of aspiration pneumonia in people with HD (Nance, Paulsen, Rosenblatt, & Wheelock, 2011). Consequently, the question of tube feeding may be raised and should be considered as a potentially life prolonging treatment. Expert opinion is divided on the merits and conclude it should be an individual decision. Indications for tube feeding may include: 10% weight loss over a month, inadequate hydration, repeated aspiration and severe swallowing difficulties (Moskowitz & Marder, 2001). Contraindications include: lack of informed consent, lack of capacity for communication and comprehension, or documented refusal (Moskowitz & Marder, 2001), while poorly controlled truncal chorea and behavioural disturbance should also be taken into account. Some suggest that tube feeding may prolong the life of an individual for whom quality of life is already dramatically reduced (Jones, 2010; Klager, Duckett, Sandler, & Moskowitz, 2008), while the reduced personal interaction people in residential care with feeding tubes receive, compared to those who are orally fed may reduce quality of life further (Jones, 2010; Moskowitz & Marder, 2001). While a person with late stage HD will generally meet all the commonly acknowledged indicators for tube feeding (Huntington Society Canada, 2016; Macleod, Jury, & Anderson, 2017) it does not eliminate the risk of choking, or the potential aspiration of colonised oro-pharyngeal material or aspiration of gastric contents. At end stage HD, the body no longer has a requirement for nutrition and loss of appetite is the body starting to shut down naturally (Gadoud & Johnson, 2015; Huntington Society Canada, 2016; Jones, 2010). Dellefield and Ferrini (2011) report that while they advise patients against tube feeding, 23% of their cohort chose to be tube fed. Swallowing can remain safe even in the event of significant cognitive decline when close attention is paid to appropriate food modifications, feeding techniques and environmental factors (Dellefield & Ferrini, 2011; Jones, 2010). When the person has advanced dementia, residential facilities and families should be encouraged to accept the increased risk associated with oral feeding rather than requesting tube feeding (Jones, 2010) and it is conceivable that some people with HD may document a wish to continue eating preferred foods, rather than have modified textures, if they place a higher value on enjoyment of eating over safety (Vaughan & Kluger, 2018). Clear guidelines on the risks and benefits of tube feeding in advanced HD would be beneficial (Simpson, 2007).