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Abstract

The Oregon "Death With Dignity" Act (DWD Act) allows a terminally ill patient with 6 months to live to ask a physician for medication to end their life. To receive the medication, the DWD Act requires the patient to verbally request the prescription twice 2 weeks apart as well as in writing. Patients with amyotrophic lateral sclerosis have three main barriers to using DWD: (a) the ability to communicate their informed consent as the disease progresses further, (b) the possibility of dementia which may affect their decisional capacity, and (c) given the nature and speed of amyotrophic lateral sclerosis, limited time is available for patients to self-administer the prescription and may rush the time line for the death. This article reviews the current knowledge and addresses the need for adjustments to existing law and recommendations for states considering a DWD law.

Keywords: United States, amyotrophic lateral sclerosis, death with dignity, competency, informed consent, physician-assisted suicide

Introduction

The Oregon "Death with Dignity" Act (DWD Act) allows physicians to prescribe medication allowing patients with a terminal illness end their lives. For patients with amyotrophic lateral sclerosis (ALS), however, the barriers

to using the DWD Act may be difficult to overcome. ALS, also known as Lou Gehrig's disease, is a degenerative disease of the nervous system which affects the brain and spinal cord (National Institute of Neurological Disorders and Stroke [NINDS], 2013) for which no treatment or cure has been identified (Mehta et al., 2016). The degeneration and eventual death of these motor neurons effectively cuts off communication from the brain to the muscles and can lead to the loss of the ability to move, eat, speak, and eventually breathe (NINDS, 2013). The progressive loss of function is fatal (Mehta et al., 2016). Patients with ALS (PALS) have three main barriers to using DWD: (a) the ability to communicate their informed consent as the disease progresses further, (b) the possibility of dementia which may affect their decisional capacity, and (c) given the nature and speed of ALS, limited time is available for patients to self-administer the prescription and may rush the time line for the death. The research presented in this article will address the need for adjustments to be made to the current law in states with DWD laws and recommendations for other states considering a DWD law.

Death With Dignity

History of the Oregon DWD Act

The Oregon DWD Act passed by a small margin in 1994 but not without protest (Cohen-Almagor & Hartman, 2001). Groups such as the American Medical Association and the Catholic Church opposed the DWD Act, the former due to the role the physician played in prescribing life-ending medication and the latter due to the perceived disrespect of the gift of life (Cohen-Almagor & Hartman, 2001). After the DWD Act passed, one of the most important responses was in the form of a lawsuit filed in federal district court which challenged the Act based on the 14th Amendment to the Constitution. The lawsuit claimed violation of due process and equal protection of vulnerable patients who may choose to utilize physician-assisted suicide due to coercion or undiagnosed mental disorders (Cohen-Almagor & Hartman, 2001). In 1997, the Supreme Court ruled there was no constitutional right to assisted suicide, and the Oregon Legislature placed the measure to repeal the Oregon DWD Act on the November 1997 ballot (Cohen-Almagor & Hartman, 2001). The citizens of Oregon voted to keep the DWD Act in place by a wider margin than originally voted for the DWD Act in 1994 (Cohen-Almagor & Hartman, 2001).

Requirements of the Oregon DWD Act

The Oregon DWD Act allows terminally ill patients in the state of Oregon with a life expectancy of 6 months or less to request and receive prescriptions for self-administered life-ending medications from their physician (Cohen- Almagor & Hartman, 2001; OR. Rev. Stat. 127.800-127.995, 1999). An important part of the law is the self-administered requirement—it does not legalize or "authorize a physician or any other person to end a patient's life by lethal injection, mercy killing or active euthanasia" (Cohen-Almagor & Hartman, 2001; OR. Rev. Stat. 127.800-127.995, 1999). A capable and competent resident of Oregon with a diagnosis of a terminal disease (e.g., ALS) can request "medication for ending his or her life in a humane and dignified manner" (Cohen- Almagor & Hartman, 2001; OR. Rev. Stat. 127.800-127.995, 1999). The DWD Act includes a provision protecting physicians from civil and criminal liability in aiding a person in the act of committing suicide, which outside of the DWD Act violates Oregon law (OR. Rev. Stat. 127.800-127.995, 1999; Cohen-Almagor & Hartman, 2001). The DWD Act does not require a physician to write the prescription for the lethal medication; however, if a patient requests the medication and the physician refuses, the patient does have a right to transfer care to another provider and the physician must provide the medical records to the new physician (OR. Rev. Stat. 127.800-127.995, 1999).

The DWD Act provides meticulous detail of the procedures required for both patients and physicians under the Act (OR. Rev. Stat. 127.800-127.995, 1999). Several steps are required prior to the prescription of the life-ending medication. First, the attending physician must make the determination that the patient is terminally ill. The terminally ill diagnosis means the disease is incurable and irreversible, and in the physician's reasonable medical judgment, will cause the patient's death within 6 months (OR. Rev. Stat. 127.800-127.995, 1999). The patient must make an oral request for assistance in ending their life in a humane and dignified way, at which time the patient's attending physician would make the determination of competency, mental health status, and the capability of self-administering the medication (OR. Rev. Stat. 127.800-127.995, 1999). The attending physician counsels the patient to ensure an informed decision, including advising the patient of the risks, results of taking the medication if prescribed, and alternative care options (e.g., hospice, palliative care). The attending physician is required to recommend the patient inform the next-of-kin of their decision to utilize the DWD Act. Furthermore, the physician will refer the patient to counseling if they suspect a mental health condition such as depression or issues with the patient's competency or capability of self-administering the

medication (OR. Rev. Stat. 127.800-127.995, 1999). Next, the patient is required to make a request in writing, witnessed by at least one person who is not related by blood, not the attending physician, health-care provider, owner of a health-care facility, or entitled to any part of the patient's estate (OR. Rev. Stat. 127.800-127.995, 1999). The patient is referred to and seen by a consulting physician to confirm the diagnosis and the length of survival time calculated by the attending physician. The consulting physician verifies the patient is competent, is capable of self-administering the lethal medication, is acting voluntarily, and has made an informed decision (OR. Rev. Stat. 127.800-127.995, 1999). If the consulting physician feels that there is a concern about mental health conditions (e.g., depression, dementia), they would recommend counseling and no medication would be prescribed until the counselor determines the mental health status and competency of the patient (OR. Rev. Stat. 127.800-127.995, 1999). The patient is required to make a second verbal request no sooner than 15 days after the first verbal request (OR. Rev. Stat. 127.800-127.995, 1999). At that time the attending physician must offer the opportunity for the patient to rescind the original request; however, the patient may rescind the request at any point in the process (OR. Rev. Stat. 127.800-127.995, 1999). The prescription for the medication cannot be written any less than 15 days after the first verbal request, and no less than 48 hours after the written request has been completed (OR. Rev. Stat. 127.800-127.995, 1999). The physician then contacts the pharmacist to inform him or her of the prescription, and once received, the pharmacist will dispense the medication either to the patient, the attending physician, or a designated agent of the patient (OR. Rev. Stat. 127.800-127.995, 1999). The patient may decide to use the medication at any time to end their life in a humane and dignified way.

Amyotrophic Lateral Sclerosis

ALS Phenotypes and Diagnosis

ALS is rare, affecting only 5.0 per 100,000 persons per year (Mehta et al., 2016). In limb onset ALS, the affected person has weakness in either the affected hand or foot, leading to tripping (drop foot) or difficult with grasping objects (drop wrist). Bulbar onset ALS leads to difficulties with speech and swallowing (Kiernan et al., 2011; Muscular Dystrophy Association [MDA] 2016a; NINDS, 2013). Furthermore, there are atypical presentations including weight loss (an indicator of poor prognosis), cramping and twitching of the muscles without weakness or wasting, unstable emotions, and frontal–temporal dementia (Kiernan et al., 2011).

ALS diagnosis is made by means of elimination. In addition, because of the uncertain nature of the ALS diagnosis, there are several diagnostic categories that patients may fit into. Arguments have been made that the diagnostic criteria are too restrictive and may have poor sensitivity, limiting diagnosis in the earliest stages when patients would most likely benefit from therapeutic intervention (Kiernan et al., 2011). Late diagnosis can prevent the successful navigation of medical and psychosocial interventions. ALS diagnosis can be a devastating blow to both the patient and family members, and indecision about and insensitivity of the delivery of the final diagnosis may delay coping with and acceptance of the diagnosis (Kiernan et al., 2011).

ALS Prognosis

ALS progression is incessant, with 50% of patients dying within 30 to 36 months of diagnosis, and 20% surviving between 5 and 10 years after diagnosis (Kiernan et al., 2011). Reduced length of survival is associated with bulbar onset, older age, and early involvement of the respiratory muscles (Kiernan et al., 2011). Increase length of survival is associated with diagnostic delay, younger age, and limb onset (Kiernan et al., 2011). Only two drugs, newly released Radicava and Riluzole, have been developed to treat ALS. Riluzole is thought to extend the life span by 3 months (ALS Association, 2016).

The earliest stages of ALS often happen prior to diagnosis (MDA, 2016b). Muscles begin to either atrophy or spasticity begins to develop, and cramps or fasciculations begin. The patient may notice fatigue, balance problems, slurred speech, trouble gripping objects, and trouble with tripping. As ALS progresses into the middle stages, symptoms become more prevalent and paralysis of muscle groups begins, while other muscles begin to weaken and atrophy and fasciculations continue (MDA, 2016b). Contractures can develop in unused muscles; swallowing can become more problematic and lead to choking and difficulty eating. Greater assistance is needed; driving is often given up and falls may occur from which the patient cannot return to standing without assistance. The respiratory muscles begin to weaken, leading to shortness of breath, especially when lying down. In late stage ALS, paralysis is widespread requiring assistance for most activities and needs (MDA, 2016b). Eating, drinking, and speech may become impossible. Respiratory muscles are compromised, which can cause sleepiness, unclear thinking, headaches, and an increase risk of pneumonia. Most deaths from ALS are due to respiratory insufficiency leading to respiratory failure which progresses over the course of the disease (MDA, 2016b). Less commonly, death may occur due to aspiration of food or saliva, pulmonary embolism, cardiac arrhythmias, and malnutrition (MDA, 2016b).

The Statement of the Problem

There is concern that the Oregon DWD Act (as well as others) may be hastening the death or unintentionally extending the life PALS due to the requirements set forth in the Act itself. The DWD Act requires that the patient ingests the medication on their own either through swallowing the medication or through a percutaneous endoscopic gastronomy or radioscopically inserted percutaneous tube. In addition, the DWD Act requires the patient to verbally request the prescription for the lethal medication twice 2 weeks apart as well as in writing in-between the two verbal requests. Furthermore, the DWD Act requires the patient to have less than 6 months to live.

The requirements for physician-assisted suicide are difficult to meet for PALS, who have a varied disease course and an uncertain time line to death. PALS must rely on computer-generated speech as the disease progresses, due to the weakening of the facial muscles, which if not successfully mastered may render the patient unable to verbally communicate their desire for physician-assisted suicide. Furthermore, PALS lose the ability to swallow and face progressive paralysis which can hamper both the ability to request physician-assisted suicide in writing and self-administer the medication. The loss of the ability to swallow and move may happen prior to the 6-month guideline provided in the DWD Act. Moreover, PALS may develop cognitive impairment as the disease progresses (e.g., frontotemporal dementia, language deficits), which would no longer allow them to be deemed competent to request the medication under the DWD Act. Research estimates that cognitive impairment may develop in 35.6% to 50% of PALS over the course of the disease (Khin, Minor, Holloway, & Pelleg, 2015). Cognitive impairment does not necessarily mean a patient is no longer competent; however, the multiplicative losses over the course of the disease may make the determination of competency in the last 6 months of life difficult or impossible (Khin et al., 2015). To further complicate the determination of competency in PALS, research indicates that physicians do not use standardized methods of assessing decision-making capacity in Alzheimer's patients, which may indicate discrepancies in assessing competency in PALS (Khin et al., 2015; Volicer & Ganzini, 2003).

There are concerns about the desire for physician-assisted suicide regarding PALS. PALS commonly report psychosocial distress and show symptoms of depressed mood, hopelessness, anxiety, and confusion (Connolly, Galvin, & Hardiman, 2015; Ganzini & Block, 2002; Gill, 2004; Monforte-Royo et al., 2011). PALS report feeling as though they are a burden to their families and caregivers and a significant increase in loneliness as the disease progresses (Achille & Ogloff, 2004; Monforte-Royo et al., 2011; Stutzki et al., 2014). The feelings of depression, loneliness, isolation, anxiety, and that the patient is a burden on their caregivers have been associated in previous research as

increasing the "wish to hasten death" through the use of physician-assisted suicide (Achille & Ogloff, 2004; Stutzki et al., 2014). Opponents of the DWD Act believe that these feelings may hasten death unnecessarily (Ganzini & Block, 2002).

Moreover, PALS may feel extreme anger if denied physician-assisted suicide due to suspected mental instability, resulting from the patient feeling powerless rather than their life is valued and hampering opportunities to resolve unrelated emotional issues (Ganzini & Block, 2002). Families may feel that the physician, acting in the role of the gatekeeper, is an insurmountable barrier to receiving a prescription and misinterpreting their concern for the patient as the patient being ignored or dismissed (Ganzini & Block, 2002). Physician–patient communication may be too focused in the here and now rather than on future needs, neglect the negative aspects of disease progression, and who the communication needs to be initiated by (e.g., neurologist, primary care, and palliative care) may not be clear to the care team, increasing the risk the patient feels unheard (Moss et al., 1996). Physicians and insurance companies also act as gatekeepers of the treatments and equipment desired by PALS, and fighting the system of bureaucracy to obtain needed items may feel like an added burden to the patient and their family, increasing the desire for a hastened death (Reagan et al., 2003). Physicians further express concerns that patients who request physician-assisted suicide later in the disease course are doing so because of poorly managed symptoms and fear of death (Ganzini & Block, 2002).

Previous Policy Recommendations and Alternative Solutions

In recognition of the difficulties faced by PALS, Oregon House Bill 3337 (2015) sought to modify the language of the DWD Act to extend the expected time until death from 6 months to 1 year. House Bill 3337 did not make it out of committee. Utah, while largely copying the Oregon DWD Act when proposing the state's own DWD Act (Utah House Bill 391, 2015), removed the expected time until death language and replaced it with the phrase 'suffering from a terminal disease; or an intractable and unbearable illness' (Utah House Bill 391, 2015). The Utah proposal, however, did not pass when voted on. Opponents of the proposed changes to the Oregon DWD Act and the Utah DWD Act felt that a 6-month time until death was an important safeguard for the patient requesting access to a prescription for life-ending medication. While unsuccessful, the bills introduced in Oregon and Utah have highlighted important limitations of DWD laws.

In addition to legislative efforts to modify DWD policy, physicians and others have suggested that improvements in both hospice and palliative care should alleviate the need for the DWD Act. Physicians believe hospice and

palliative care will allow patients to receive adequate pain control, counseling for the patient and their caregivers to alleviate fear and anxiety, and access to home care staff to relieve the perceived burden of the patient's illness (Achille & Ogloff, 2004; Ganzini & Block, 2002; Monforte-Royo, 2011; Moss et al., 1996; Reagan et al., 2003; Stutzki et al., 2014). Proponents of the DWD Act disagree, as it removes the patient's choice to have a dignified death of their choosing, rather than the unbearable suffering brought on by terminal illness.

Policy Recommendations

While the extended time line to death, as seen in the proposed amendment to the Oregon DWD Act (2015), and the language change to "suffering from a terminal disease; or an intractable and unbearable illness" suggested in the proposed Utah DWD Act (Utah House Bill 391, 2015) have not been implemented, these changes are necessary to ensure equal access for PALS to physician-assisted suicide. Given the uncertainty of ALS progression and when the ability to speak, write, and swallow will be lost to the PALS, ensuring adequate time to request and receive the medication from the doctor while a patient's health is still intact is vital for this group. In addition, extending the time line or modifying the language of such bills allows for patients with other conditions with uncertain progression paths (e.g., Alzheimer's) to use the DWD Act. Modifications may be suggested for specific disease diagnoses to leave in place safeguards for patients with terminal illness that are more predictable in nature. In addition, conversations between lawmakers and PALS may allow lawmakers to better understand this extremely rare disease. Moreover, policy makers need to understand ALS more thoroughly to better assess future amendments to DWD Acts currently in place as well as DWD Acts under consideration. For PALS, hastening death out of fear that the medication may not be self-administrable is a real concern. Clarifying the language of DWD Acts to allow for percutaneous endoscopic gastronomy or radioscopically inserted percutaneous tube administration, as well as the ability for the patient to begin administering the medication and allow for a trained person to finish administering it, may allow patients fearful of missing their window for physician-assisted suicide more peace of mind and allow for them to delay death for additional time with their loved ones. It may also allow for additional time to provide intervention such as palliative care, home health care, and hospice or respite care for caregivers to be put into place, possibly increasing the quality of life for PALS and decreasing the use of the life-ending medication in this population.

Given the issues of physician–patient communication, medical schools need to require additional training for their staff and students on how to discuss terminal illness and the various pathways a terminal illness may take. For example, discussion of the typical causes of death (e.g., respiratory failure) versus the feared cause of death (e.g., choking) may allow patients to feel more in control of their treatment course. Exploring options such as noninvasive versus invasive ventilation, forgoing the percutaneous endoscopic gastronomy or radioscopically inserted percutaneous tube to allow a natural death by withholding nutrition and hydration, and acknowledging the patient's possible desire for physician-assisted suicide versus palliative care may allow the patient to feel more connected with their physician. The opportunity for PALS to see their physician as a partner, rather than a gatekeeper, may in turn allow PALS to be more open to possible options for care. Moreover, ensuring that physicians understand the patient needs, and deals with them proactively by ordering equipment (e.g., wheelchairs, computer-assisted voice programs), ahead of the point of need may help the patient retain autonomy and control over their situation. These actions may reduce the need for, as well as the use of, physician- assisted suicide. Physicians, however, must be aware in the states where DWD Acts have been passed that their patients may request physician-assisted suicide and should prepare themselves to listen to the patient's request and handle it appropriately.

Implementing a standard assessment of cognitive abilities, thereby ensuring patients are evaluated equally by physicians, is an important aspect of improving the DWD Act in Oregon and other states that have implemented similar laws. The use of a standardized system of evaluating competency will reduce some of the pressure felt by physicians to make the correct decisions regarding the request for life-ending medications. In turn, a standardized method to assess competency may allow the patient to avoid feelings of anger and allow them to work through the emotional and psychosocial issues barring them from receiving their prescriptions. Future research around ALS and DWD may need to focus on the areas of competency more thoroughly to better understand the possible implications of competency testing and may help physicians and families better understand the disease progression and issues of competency.

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