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DISPROPORTIONATE USE OF AID IN DYING AMONG PEOPLE WITH ALS:

WHY ALS AID-IN-DYING REQUESTS ARE COMMON WHILE ALS IS RARE

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ABSTRACT: People with amyotrophic lateral sclerosis (ALS) disproportionately use aid in dying. We explore aspects of the ALS illness experience that may help explain the higher rates of aid-in-dying requests in this disease relative to others. In particular, the desire to maintain control is prominent in the face of a relentlessly progressive disease that results in substantial disability. We also describe how the requirement for self-administration of aid-in-dying medications impacts people with ALS. We conclude with suggested next steps to further our understanding of aid in dying in people with ALS so that we can better provide them with comprehensive, person-centered care.

KEYWORDS: aid in dying, amyotrophic lateral sclerosis, ALS, medical aid in dying

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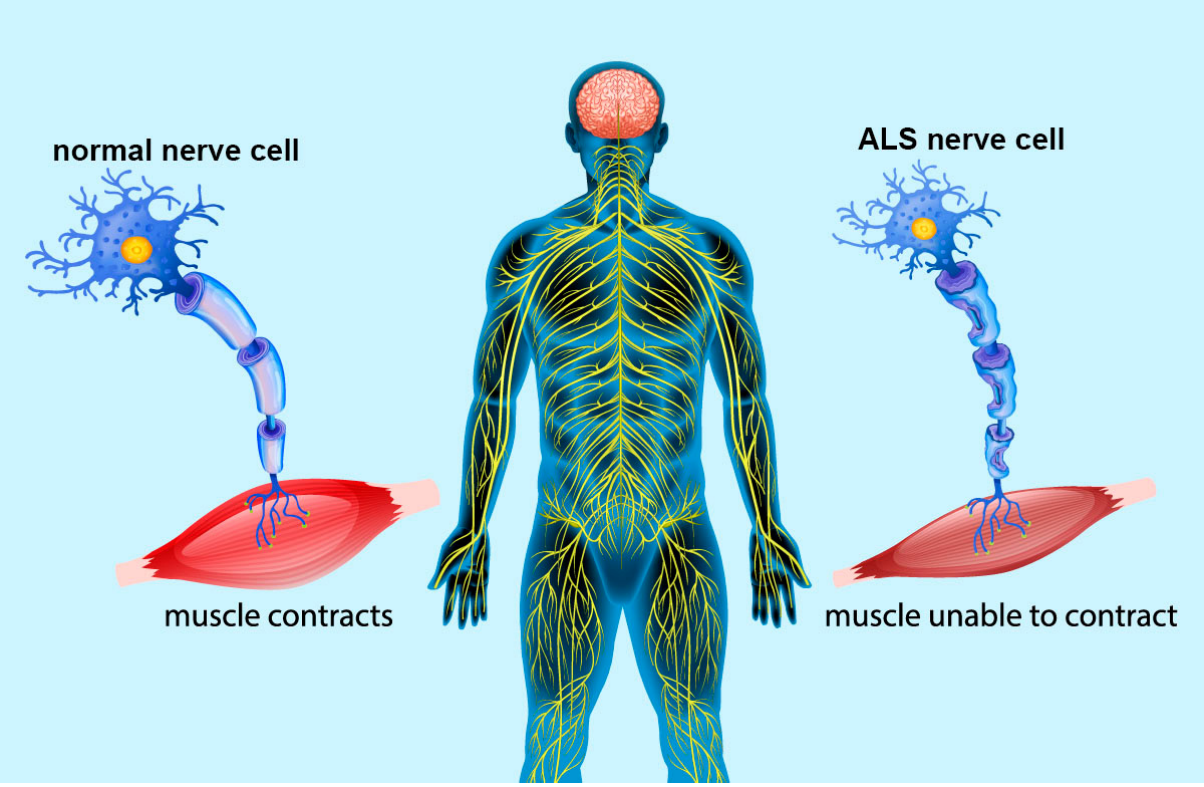

INTRODUCTION

People living with amyotrophic lateral sclerosis (ALS) disproportionately pursue medical aid in dying. While cancer is the illness with the largest number of people who die through aid in dying, this is largely because cancer is so common. ALS is an uncommon illness, but a much higher percentage of people with ALS request aid in dying than cancer and other illnesses. For example, an estimated 3.4–6.7% of people living with ALS received aid-in-dying prescriptions in Washington state between 2009 and 2014, compared to only 0.6% of people who died from cancer and 0.2% of all people who died.[1] In 2023 in Oregon, 5.7% of all people who died via aid in dying had ALS, while ALS had a population prevalence of just 0.08%.[2,3]

The high proportion of ALS patients who contemplate aid in dying is not new; between 1995 and 1997, 56 of 100 people with ALS in Oregon and Washington surveyed said that they would consider what was at that time called physician-assisted suicide if it became legal.[4] Oregon's Death with Dignity Act followed in 1997.[2]

To increase our collective understanding and shed light on this important issue, we convened an interdisciplinary group. The group consisted of ALS and palliative care clinicians (ALS neurologists, palliative and neuropalliative care specialists, an ALS speech pathologist, and a palliative care chaplain) from eight institutions across six states—including both states where aid in dying is legal and where it is not—as well as a person with ALS and a care partner representative. Participants in this focus group re-

sponded to the question, *In your experience, what is driving the relatively high frequency of medical aid-in-dying requests in people with ALS?* The conversation was wide ranging and included not only potential explanations but also a discussion of the nuances of aid in dying in ALS cases and ideas to advance practice in the area. A subset of the focus group collaborated on this article to present our thoughts and propose next steps.

POSSIBLE MOTIVATIONS FOR MEDICAL AID IN DYING IN PATIENTS WITH ALS

It may be tempting to simply assume that because people with ALS suffer more than people with other illnesses, they use aid in dying more frequently. But it is impossible to compare the subjective and multifaceted experience of suffering across different illnesses. Instead, we identify four aspects of the ALS experience that help explain the higher rate of aid in dying. They are: the importance of control in the face of dependence, the relentlessness of the disease course, concern about death from choking or respiratory failure, and worry about being a burden on others.

Control in the face of dependence

In our experience, and consistent with the literature, a major motivation for aid in dying among people living with ALS is the desire to have control over the dying process and the associated desire to preserve autonomy.[1,5,6] Desire for control is a common motivator for people with all serious illnesses to pursue aid in dying.[7–9] However, this motivation may be particularly common in ALS patients, given the nature of the disease.

ALS is characterized by an inexorable series of losses in ability—first to engage in favorite activities, then to perform activities of daily living, then to eat and speak, and finally to breathe unassisted. The loss of the ability to communicate verbally while cognition is preserved is especially difficult for people with ALS and differs from the progression of most other illnesses. With each of these losses, control diminishes and dependence increases. People with ALS often fear the most extreme manifestation of this situation: being "locked in" at advanced stages of the illness, with near complete paralysis. Given how profound the loss of control can be in ALS, it is understandable that people seek a way to gain some agency through aid in dying.

Clinicians in our group observed that simply knowing about the option of aid in dying or having access to aid-in-dying medications can provide comfort to some people living with ALS, even if they have no immediate plan to take the medications. The comfort of knowing aid in dying is available gives patients an increased sense of agency and more options in the face of this illness and can be therapeutic even without

Sandy Morris, an ALS patient, on her porch in the Sierra mountains of California. She had a medically assisted death later that year. Photo: Talia Herman

changes in a person's current physical circumstances.

Relentlessness of the disease course

ALS is well recognized to be tenacious in its course. Once neuromuscular function is lost it does not return, and a future with increasing disability is certain. Even if a person with ALS takes FDA-approved medications and decides to extend their life with percutaneous endoscopic gastrostomy (common) or tracheostomy (less common), these therapies do not reverse loss of function or stop the ongoing losses.[10,11] Instead, they can enable a person with ALS to live to the point of near-complete paralysis, while generally fully aware of their own limitations.

The course of illness in ALS contrasts with other terminal diseases. In heart failure and cancer, for instance, therapies can extend quality of life and often improve symptoms and function such that overall decline is punctuated by periods of relative recovery and "better days," even close to the end of life. The relative preservation of cognition in ALS also distinguishes the decline from other neurodegenerative conditions like dementia, in which people often have less awareness of their dependent situation.

In our experience, people living with ALS and their care partners understand and anticipate the relentless nature of this terminal disease. A study in the Netherlands found that most (56%) caregivers of

people with ALS who chose to hasten death via euthanasia reported that "no chance of improvement" was a reason for that choice.[12]

Concern about death from choking or respiratory failure

Prior research has found that uncontrolled pain is a less common motivator for aid in dying among ALS patients compared to people with cancer.[1] Respiratory distress, however, is a particularly feared symptom of end-stage ALS. A review of 30 people with ALS considering aid in dying in California found that many worried about "intolerable symptoms" like shortness of breath.[5] In addition, many ALS patients with compromised swallowing function (dysphagia) have had the experience of choking when they try to drink or eat. Choking combined with an inability to cough forcefully because of diaphragmatic weakness can be very frightening. Therefore, people living with ALS may also seek aid in dying because of a fear of dying from choking or respiratory failure, which they often refer to as "suffocation."

Fear of distressing shortness of breath might also be expected in advanced respiratory illnesses like chronic obstructive pulmonary disease or interstitial lung disease. However, these respiratory illnesses often have a more fluctuating course, and there may be a sense that more can be done to treat the underlying illness even at advanced stages (e.g., increased supplemental oxygen, steroids, or antibiotics).

It is important to note that some people with ALS may not know that a death by hypoventilation can often be peaceful and comfortable and that opioids, benzodiazepines, and positive pressure ventilation can help relieve their suffering. ALS clinicians must educate patients about the typical end-of-life course of ALS and the options for palliation of respiratory symptoms. Even after receiving such education, some patients living with ALS may prefer the faster and more controlled process of aid in dying rather than a more prolonged and potentially uncomfortable death by respiratory failure or aspiration.

Worry about being a burden on others

Because people living with ALS are often significantly dependent on others for their physical needs, family members and other care partners typically spend a substantial amount of time providing direct care, attending to household tasks, managing equipment, obtaining medications, coordinating care, and so on. Unlike many other serious neurologic illnesses, such as Parkinson's, patients with advanced ALS have a short prognosis and maintain the cognitive capacity to make them legally eligible for aid in dying. The degree of perceived burden coupled with intact insight may distinguish ALS from other progressive neurological diseases in which people become highly dependent. Ad-

...WE DO NOT MEAN TO IMPLY THESE FEELINGS ARE UNIVERSALLY HELD AMONG ALL ALS PATIENTS.

ditionally, the extent and duration of functional dependence in ALS is greater than for many other serious illnesses, such as cancer, in which patients tend to be more highly functional until the final stage of illness.

In large part because of the caregiving needs associated with the illness, the financial impact from ALS can also be substantial. The literature suggests that some people living with ALS are motivated toward aid in dying in part because they worry about the burden on their family and other care partners, though not all care partners experience their role as burdensome.[1,5,9]

CRITERIA FOR AID-IN-DYING ELIGIBILITY

Though late-stage ALS can impact cognition in up to half of people, at the time when their prognosis is less than six months, which is a requirement for medical aid in dying, the cognitive effects are most commonly not severe enough to impair the capacity to make a decision about aid in dying.[13–16] This situation is in contrast to people with other neurodegenerative diseases with major motor impairment, such as Parkinson's disease, which more commonly than ALS can have severe cognitive impairment before the less than six-month prognosis, precluding those patients from aid in dying.

US laws also require that people self-administer aid-in-dying medications—for instance, orally or by depressing a plunger on a 60-to-100cc syringe attached to a gastric feeding tube or a rectal catheter. Some people with ALS fear that they will lose the strength to do so, denying them the aid-in-dying option if they wait until they are too weak. This has led some ALS patients to pick an earlier date for aid in dying than they would otherwise prefer in order to avoid the window of opportunity closing and being trapped in an intolerable state. In this case, it would be impossible to know whether they would, in fact, have found that state intolerable. It is worth noting here that recent clinical knowledge and innovations have led to a number of methods whereby ALS patients with minimal limb motor abilities can still self-ingest aid-in-dying medications, for example, by clasping and then releasing a valve in their mouth or pushing a syringe with their forehead.

Regardless, concerns about losing access lead to increased stress and worry around the process, which

raises a challenging ethical question: Some claim that the law as currently written discriminates against people with physical disabilities and call for targeted changes to increase access to aid in dying for people with neuromuscular diseases like ALS.[17,18] Notably, in many countries outside of the US where aid in dying is legal, prognostic criteria and the requirement of self-administration are less stringent.[19,20]

IMPLICATIONS AND NEXT STEPS

Aid in dying for ALS is a topic about which people, depending on their values, beliefs, and lived experiences, have a wide range of strongly held opinions. Certainly, not all people living with ALS will consider aid in dying, and in discussing some reasons why people with ALS may consider aid in dying, we do not mean to imply these feelings are universally held among all ALS patients. However, since this is an illness in which requests for aid in dying arise relatively frequently, we feel it is important to understand why and prepare clinicians to address the existential needs revealed by these requests.

Clinicians may feel that if we are unable to ease symptoms or fears enough to obviate a patient's wish for aid in dying, we are failing as healers and falling short of our commitment to alleviate suffering and provide comfort. However, we also maintain a commitment to walk with people through their illnesses no matter where that leads and to address their options for palliation and, in states where it is legal, the potential for aid in dying.

A European review of physician attitudes toward aid in dying in patients with ALS suggested that physicians are more comfortable responding to a request for aid in dying than initiating the conversation.[21] This is also true in our experience—although within our focus group, some clinicians' practices have shifted to introducing the option of aid in dying earlier in advance-care-planning conversations as part of informed consent about their spectrum of end-of-life options before patients address it specifically. Future studies should explore the question of whether and how clinicians and community organizations should or should not proactively educate people living with ALS about the option of aid in dying. Many ALS patients may already have the question of aid in dying on their minds, and an open discussion could address unanswered questions or concerns, bringing ease and reassurance, whether or not an assisted death is pursued.

However, we must also be wary of the Disability Paradox, by which people without a serious disability (such as clinicians or people in early stages of illness) underestimate the quality of life with a disability.[22] Clinicians must take great care when communicating with patients not to suggest that life with ALS will eventually feel not worth living. Most people with ALS do not feel this way, and any implications to the contrary could quickly degrade trust.

Given that a desire for control features strongly in ALS patients who consider aid in dying, further research is needed to explore additional ways that control and agency can be offered to people with ALS—through education, involvement in advocacy, and highlighting strengths and capabilities that persist in the face of physical disability. These actions might also provide comfort and reassurance.

It is imperative that we engage diverse groups of people living with ALS and their care partners. We acknowledge that nobody in this group of authors is living with advanced ALS, and we cannot understand the nuances of these decisions without that lived experience. We also appreciate that each person's experience with ALS is unique and that culture, background, and lived experience influence people's thoughts and feelings about aid in dying. We need a concerted effort to involve a diverse range of people from a wide range of demographic, cultural, and religious groups at varied clinical stages of ALS in further research, which could include qualitative interviews with ALS patients, to understand not only motivations for aid in dying but also how these requests can best be addressed.

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