

# END-OF LIFE DECISION MAKING IN AMYOTROPHIC LATERAL SCLEROSIS

Wendy Johnston, MD, FRCP(C)

University of Alberta  
Edmonton, AB, CANADA

## Introduction:

Amyotrophic Lateral Sclerosis (ALS) is a relentlessly disabling neurological disorder that progresses to respiratory failure. Without respiratory support the 5 year survival is approximately 20% with 50% succumbing within 3 years from diagnosis. Symptomatic measures, including improved nutrition and non-invasive respiratory support, as well as the modest benefit of the one FDA-approved drug, Riluzole, have been associated with better survival and quality of life. None-the-less, patients with ALS have limited options with respect to treatment and inevitably face decisions about accepting or forgoing life-sustaining therapies. Management of ALS therefore requires developing individualized advance care planning, particularly addressing specific decisions regarding respiratory and nutritional support. The medical, psychosocial and spiritual context for each individual and family must be recognized and these factors as well as physical and financial resources will influence both the decisions and their implementation.

Initially, management of ALS emphasizes knowledge acquisition, adaptation and rehabilitation. From the time of diagnosis, maintaining quality of life should be emphasized; dimensions beyond the physical domain must be recognized.

The principles of palliative care are applicable to the management of ALS from the time of diagnosis, but assume greater prominence as the disease progresses. For the majority of those with ALS who seek neither to hasten nor delay death, palliative care is the only option. It remains the underlying principle of care for those who would prolong or hasten death. What constitutes the best end-of-life care, and how we can improve it, was the focus of the Robert Wood Johnson Foundation Promoting Excellence in End of Life Care ALS Peer Workgroup Project, which provides both a guide to current management and direction of research and policy development (Mitsumoto et al, 2004). Recent publications offer guidelines for palliative care in ALS, including symptom management, as well as recommendations for breaking the news, the timing of decision-making, psychosocial and spiritual care (see resources).

The controversies surrounding end-of-life decision making in ALS have been featured prominently in both the medical literature and in the general media. Whether in the form of legal appeals, or in televised coverage of actual deaths, patients with ALS have appeared at the forefront of the debate on physician-assisted suicide (PAS) and euthanasia.

The spectrum of apparent choice for patients with ALS ranges from seemingly indefinitely postponing death with long term assisted ventilation through hastening death by physician-assisted suicide and euthanasia. Regardless of the two extremes, whether by choice or chance, or lack of recourse, most patients with ALS die without availing themselves of either, but as a consequence of disease progression. In the terminal stage, when palliative measures come to the forefront of care, fear of the potential double-effect of using sedatives and painkillers necessary for symptom management, yet that might hasten death, can hamper adequate symptom management at the end of life.

## **Advance Care-Planning: *Timing of discussion of end-of-life issues: Framing, denial and cultural sensitivity***

Choice of Life Sustaining Treatment

I. Passive: no plan

II. Active planning for intervention

Monitor swallowing, pulmonary symptoms and function,  
offer PEG when comfort with eating or nutrition compromised, or FVC 50% predicted  
offer NIV (discuss LTV) when symptomatic or FVC 50% predicted

### III. Active planning to forgo interventions (may follow II above)

Explicitly discuss symptoms and scenarios at end of life, plan hospice referral, involve proxy decision-maker

The timing of the discussion of end-of-life issues and advance care-planning with patients and their families has to strike a balance between the desire to know on their part, and the need to make timely decisions about life-sustaining therapies. Advance care-planning should be considered from the time of diagnosis. In breaking the news about the diagnosis of ALS, planning for interventions should be included as part of disease management, which may lead naturally to discussion of end-of-life issues. Recommendations for the breaking the news are included in the AAN Practice Parameters (Miller et al 1999).

The timing and manner of disclosure of the diagnosis can have a lasting impact on the patient and family. In one survey more than 50% were dissatisfied with how they were informed of the diagnosis (Borasio et al, 1998). A recent U.S. survey (McCluskey et al 2004) revealed that 46% of patients and 52 % of caregivers retrospectively rated the clinician breaking the news as good or excellent. Basing survey categories on the same SPIKES (setting, perception, invitation, knowledge, exploration and strategy/summary) protocol used as the basis of the Practice Parameter recommendation, McCluskey and colleagues found those clinicians viewed as having done a good to excellent job of breaking the news were more likely to have scored well in the SPIKES subcategories. Those who rated good or excellent were recalled as having spent more time than those judged average to poor. However, specifically addressing end-of-life issues did not differentiate between good-excellent vs. average-poor.

While there are expert opinions as to when to address EOL issues in ALS, no study has assessed timing for its impact on outcomes (such as unwanted invasive ventilation) nor quality of life of patient or caregivers. The ALS Peer Workgroup identified six triggers for discussion of EOL issues (Mitsumoto et al, 2004):

1. The patient or family asks
2. Severe psychological, social or spiritual distress or suffering
3. Pain requiring high doses of analgesic medication
4. Dysphagia requiring feeding tube
5. Dyspnea or symptoms of hypoventilation, or FVC of 50% or less
6. Loss of function in 2 body regions

End of life care should be addressed routinely as part of discussions about prognosis, or when considering interventions that could have a low probability of success. More urgent discussions would be prompted by concerns that death is imminent (as perceived by the physician, patient or family) or that disease progression has been severe. An expressed desire to die, queries about assisted suicide and interest in hospice or palliative care must be dealt with immediately. Severe suffering is an emergency that demands medical intervention (adapted from Borasio and Miller, 2001).

The failure to address advance care-planning leads to unplanned interventions. Whether through ignorance or denial, 66% of patients on long-term ventilation (LTV) recently surveyed in Germany were not aware of their impending respiratory failure before emergency intubation (Kaub-Wittermer et al, 2003) and 81% did not give informed consent for the procedure. Although 79% of those on LTV surveyed in six U.S. states were informed about mechanical ventilation, only 21% had chosen it in advance (Moss et al 1993). Oppenheimer (1994) found only 5% had chosen mechanical ventilation in advance.

Overall, the use of tracheostomy and mechanical ventilation is low in the U.S. (Bradley et al, 2001, Albert et al, 1999), at 4% or less. In spite of the advent of Practice Parameters advocating the use of Non-invasive Ventilation (NIV) for symptomatic management of respiratory failure, use of even NIV remains low (Bradley, 2001). This could reflect a lag in knowledge, but could also reflect the values and attitudes of the medical practitioners providing the care. Variability in rates of mechanical ventilation in ALS patients in Illinois was related to physician attitudes in the medical centers where the patient was managed (Moss et al 1993). Physicians "frame" the discussion of mechanical ventilation in chronic lung diseases in either a positive or negative way (Sullivan et al, 1996) depending on their perception of the patient's quality of life, and the potential reversibility of respiratory failure. This has not been evaluated for ALS patients. The variability in use of riluzole (RiluteK®) in different centers as reported in the ALS CARE database likely reflects framing of the information presented for ostensibly patient-centered decision making. It is possible that the physician's own attitudes to NIV and LTV may be the main determinants of the likelihood of use of these interventions for most patients with ALS.

Patient autonomy in decision making can only be assured when the available information is presented neutrally, but fully. The resources available to support the decisions must be understood, as well as the family and community support. Advance care planning must be firmly grounded in the values of the individual, who in turn must understand the consequences of the decisions, both for themselves and their family. Values of the individual may not reflect the mainstream; exploring the spiritual and cultural values of the person with ALS and their family should be integral to the decision making process, and should be established before crises occur. When cultural differences appear to preclude patient-centered decision-making, or appear at odds with the practitioners, consultation with an institutional ethics committee, community leaders or spiritual counselors of the individual may resolve potential conflict.

### **Physician-Assisted Suicide and Euthanasia: *Ethical, legal and practical concerns***

The re-emergence of the “right-to-die” social movement in the mid-20th century, and its rise to prominence, parallels the development of successful medical interventions to extend life, as well as the legal milestones sanctioning the right of individuals (or their proxy decision makers) to refuse or withdraw life-sustaining measures (McInerney, 2000). Patients with ALS have featured prominently in legal challenges, as well as individuals represented in the press and on television.

Patients with ALS seem to be more likely to request and complete assisted suicide or euthanasia than those with other terminal diseases such as cancer (Ganzini and Block, 2002). Interest in assisted suicide is high (Ganzini et al, 1998) and sustained (Ganzini et al 2002b) in Oregon where PAS has been legal since 1998. In a survey of U.S. neurologists, of those that considered themselves specialist in ALS, 41% had received at least one request for assisted suicide, with two-thirds of requests occurring in the last 6 months of life (Carver et al, 1999). Recent studies in New York (Albert et, 2003) and Germany (Neudert et, 2003) indicate a high level of interest in hastening death even in those patients enrolled in a hospice, or under the care of physicians in a palliative care unit.

The debate about the ethics of PAS and euthanasia center on interpretation of basic principles of medical practice that superficially do not conflict: the imperatives to relieve suffering, respect patient autonomy and to do no harm. Medical ethicists have written in support of PAS and euthanasia for patients with ALS (Loyal, 2000) and in opposition (Bernat, 2001). It is therefore not surprising that a survey of neurologists reveals a range of attitudes, as well as persistent concerns about the morality and legality of withdrawal of life support and the use of medications that sedate or potentially depress respiratory function (Carver et al 1999).

The American Academy of Neurology and other professional organizations specifically condemn PAS and euthanasia (American Academy of Neurology Ethics and Humanities Subcommittee, 1998). Yet 44% of neurologists surveyed indicated willingness to perform PAS if legalized, and 13% would do so under current conditions (Carver et al, 1999).

In the wake of legalizing PAS in Oregon, studies of the apparent motivation of those requesting or completing PAS suggest that loss of autonomy, control and independence, and the inability to pursue pleasurable activities play a role, more than physical symptoms (Ganzini et al 2000, Sullivan et al 2001).

The results of surveys of ALS patients and their caregivers in Oregon and SW Washington also support these core concerns, but in addition, fear of future suffering and higher levels of hopelessness (but not depression) were associated with interest in obtaining a lethal prescription. Fixed characteristics of the individual, including male gender, higher educational and socioeconomic status, and potentially modifiable factors including religiosity were also significantly associated with interest in PAS (Ganzini et al 1998). There was a mild negative correlation between self-reported Quality of Life and interest in PAS, but prevalence and severity of physical symptom did not differ significantly between groups. In contrast, a request for PAS in the last month of life did correlate with higher pain scores and insomnia (Ganzini et al 2002a). Interest in, or requests for PAS may reflect a number of concerns. These queries should be approached as the opening to discuss end-of-life issues in general.

An ongoing study of ALS patients in the hospice setting (Albert et al, 2003) revealed a significant interest in hastening death in 23%. The decision to hasten dying was expressed consistently before death. Those who hastened dying reported poorer mood and less religiosity; they are more likely to have depressive symptoms of clinical significance, feel less in control and more hopeless. Although the numbers are small (11 of 47 deaths were hastened, one by suicide, 10 by sedation for the imminently dying) this careful prospective study is the first to identify factors that might predict interest in hastened death, and factors that may be modified by improved care.

Both PAS and euthanasia have been sanctioned, though only recently legalized, in the Netherlands. In the period 1994 to 1999, 20% of patients with ALS, according to the physicians surveyed, died as a result of PAS (3%) or euthanasia (17%). The response of the physicians to the survey was high (responses accounted for 72% of patient deaths). The choice of physician assisted death was positively associated with dying at home, and negatively associated with anxiety and importance placed on religion. Other variables (age, income, educational level, disease or care-related) were not associated with choosing physician-assisted death. "The frequency of feelings of pain, despair, fear, choking and anger were felt to be similar in the two groups of patients" (Veldink et al, 2002). The study used medical records as well as the physician survey; it is not stated from which source the symptom information was extracted. Two deaths by euthanasia were of unconscious patients who had not explicitly requested it, although one had an advance directive requesting physician assisted death. The study did not directly survey patients, using the physician's recollection and records.

The study by Veldink et al yields other interesting information about death and ALS. No end-of-life decision was made in 54 (27%) of those who died, and in 37 (18%) "such decisions could not be made because the patients died suddenly", although 9/37 had advance directives requesting physician assisted death. Therefore 40% of patients studied died without having made any decisions. Although tracheostomy was present in 3% and NIV used in 16% no comment is made about withdrawal of respiratory support nor how such cases were classified. Guidelines for withdrawal of respiratory support from ventilator-dependent patients provide specific recommendations that include both sedation and analgesia (Borasio & Voltz, 1998, Miller et al, 1999) however it is possible that euthanasia may have been used instead.

In general, requests for assisted suicide and euthanasia do not persist, but persistent requests are very challenging for physicians, even when a legal framework for the query exists. Interviews with physicians in Oregon who received these requests demonstrated that they are emotionally difficult both for physicians who might participate in PAS as well as those who feel they cannot (Ganzini et al 2003, Dobscha et al 2004). The physician should be ready to listen thoroughly and assure the patient that no matter what the final decision, the physician is available to the patient through the illness, even if he or she cannot prescribe a lethal medication. Some physicians reported a sense of hopelessness and failure after receiving a request. At other times, too much empathy and identification with the patient will lead to failure to thoroughly look for alternatives. In our experience, patients who persist in wanting assisted suicide have strong needs for control, negative views of the future, and strong dislike of being dependent on others—all areas in which ALS particularly affects patients. There is the risk that too much medical intervention may result in the patient feeling more dependent. Every effort to improve the patient's independence and avoid institutionalization should be made, even if safety in the home is not optimal (Ganzini et al 2003)

### ***End-of-life Care: When to refer to hospice; symptom management in terminal ALS; fear of the double-effect***

Management of ALS throughout its course is shared by many. The need for multidisciplinary care is even greater as the disease approaches its end. The Practice Parameters recommend involvement of specialist palliative care services, which in many instances are provided by hospice services. In the U.S. Medicare Guidelines restrict referral to hospice services to those with remaining life estimated at less than 6 months; the specific requirements for ALS (see table below) are felt to be onerous and restrictive (McCluskey and Houseman 2004).

### **Medicare Criteria: Determination of Terminal Status in ALS**

Patients will be considered to be in the terminal stage of ALS (life expectancy of six months or less) if they meet the following criteria. (Must fulfill 1, 2 or 3)

1. Patient must demonstrate critically impaired breathing capacity.
  - a. Critically impaired breathing capacity as demonstrated by **all** of the following characteristics occurring within the 12 months preceding initial hospice certification:
    - i. Vital capacity (VC) < 30% of normal;
    - ii. Significant dyspnea at rest
    - iii. Requiring oxygen supplementation at rest
    - iv. Patient declines invasive ventilation.
2. Patient must demonstrate **both** rapid progression of ALS **and** critical nutritional impairment:
  - a. Rapid progression of ALS as demonstrated by **all** of the following characteristics occurring within the 12 months preceding initial hospice certification:

- i. Progression from independent ambulation to wheelchair, or to bed bound status
  - ii. Progression from normal to barely intelligible or unintelligible speech
  - iii. Progression from normal to pureed diet
  - iv. Progression from independence in most or all activities of daily living to needing major assistance by caretaker in most all
- b. Critical nutritional impairment as demonstrated by **all** the following characteristics occurring within the 12 months preceding initial hospice certification:
- i. Oral intake of nutrients and fluids insufficient to sustain life
  - ii. Continuing evidence of weight loss
  - iii. Dehydration or hypovolemia
  - iv. Absence of artificial feeding methods
3. Patients must demonstrate **both** rapid progression of ALS **and** life-threatening complications.
- a. Rapid progression of ALS, see 2.a. above
  - b. Life-threatening complications as demonstrated by **one** of the following characteristics occurring within the 12 months preceding initial hospice certification:
    - i. Recurrent aspiration pneumonia (with or without tube feedings)
    - ii. Upper urinary tract infection, e.g. pyelonephritis
    - iii. Sepsis
    - iv. Fever recurrent after antibiotic therapy
    - v. Decubitus ulcers, multiple stage 3-4, particularly if infected

The necessity for modifying the criteria for referral to Hospice was recognized earlier (Del Bene et al, 1999) and the following criteria proposed:

**Hospice Qualifying Symptomatology (Columbia University):**

(A) or (B) must be present. (A) does not require associated symptoms. If just (B), at least 2 other Respiratory Indicators must be present or 1 respiratory and 1 Nutritional Indicator present.

(A) FVC less than or equal to 30% predicted

(B) FVC less than or equal to 60% predicted with a steady decline over past 2-3 months

Other Respiratory Insufficiency Indicators:

- Shortness of Breath
- Shallow Breathing
- Paradoxical Breathing
- Inability to be supine due to diaphragmatic weakness
- Non-explosive cough
- Inability to blow nose
- Breath Support poor as evidenced by accessory muscle use
- Endurance poor (excessive fatigue)
- Significant insomnia related to diaphragmatic weakness

Nutritional Insufficiency Indicators:

- Excessive oral secretions with dysphagia
- Nutritional Compromise/Dehydration (with or without feeding tube)
- Weight Loss >10% of body weight and refusing PEG
- Aspiration or choking (on food or liquid)

Standard Requirements

- Prognosis of 6 months or less
- Patient election of Hospice services
- Patient election of DNR status
- Patient refusal of Invasive/Biomedical Intervention
- Patient is Home Care Eligible

In a retrospective study of 97 consecutive patients with ALS referred to hospice, McCluskey and Houseman (2004) reported that only 5 met the Medicare criteria for admission to hospice, yet 91% stayed under hospice care for less than 180 days with a mean of 84 (range 1 – 534) days (98% met the Columbia criteria). They comment

that while there was no difficulty in referring any in this group from their tertiary care ALS clinic, community based physicians may face greater restrictions on hospice availability for their patients. Based on this concern, a working group consisting of representatives of the American Academy of Neurology, the ALS Association and the national hospice and palliative care organization (NHPCO) has developed criteria for hospice referral as follows (McCluskey 2005, personal communication):

### **Proposed Penn ALS Hospice Criteria**

Patients are considered eligible for hospice care if they do not elect for tracheostomy and invasive ventilation and display evidence of critically impaired respiratory function (with or without use of NIPPV) and / or severe nutritional insufficiency (with or without use of a gastrostomy tube):

A. Critically impaired respiratory function is as defined by:

1. FVC < 40% predicted (seated or supine) and 2 or more of the following symptoms and / or signs

- Dyspnea at rest
- Orthopnea
- Use of accessory respiratory musculature
- Paradoxical abdominal motion
- Respiratory rate > 20
- Reduced speech / vocal volume
- Weakened cough
- Symptoms of sleep disordered breathing
  - Frequent awakening
  - Daytime somnolence / excessive daytime sleepiness
- Unexplained headaches
- Unexplained confusion
- Unexplained anxiety
- Unexplained nausea

2. If unable to perform the FVC test patients meet this criterion if they manifest 3 or more of the above symptoms / signs.

B. Severe nutritional insufficiency is defined as:

Dysphagia with progressive weight loss of at least five percent of body weight with or without election for gastrostomy tube insertion.

These revised criteria rely less on the measured FVC, and as such reflect the reality that not all patients with ALS can or will undertake regular pulmonary function tests.

If a hospice resists referral of ALS patients, it may reflect concern over the need to meet existing Medicare Criteria. Lack of familiarity with ALS and its management, or concerns over the costs of interventions may also be barriers to acceptance. Hospice care is covered by global budgeting, thus the cost of insertion of PEG, institution of NIV or use of riluzole may be viewed as burdensome. It is worthwhile to address the concerns, provide expert support and even in-service presentations to the appropriate hospice providers to facilitate palliative services to ALS patients.

The intensity of symptom management required by many ALS patients in the terminal phase is best supported by hospice services. Symptoms that dominate include excess secretions (saliva, bronchial secretions), dyspnea, insomnia and pain (Ganzini et al 2002a). Hospice also recognizes and supports the caregiver and provides psychosocial and spiritual support for the patient and family. Many have bereavement services as well. It is important to continue to remove barriers to hospice care for ALS patients and their families

The end-of-life decision most frequently made in the Dutch study was the use of medications in doses that "probably shortened the patient's life". Given the context of the study, with categories that reflected the intention of the physician to end life explicitly, this category does not likely reflect intentional overdose, but may reflect concern on the part of the physician that the use of morphine and sedatives such as benzodiazepines would

shorten life. Such concerns were reflected in the survey of U.S. neurologists (Carver et al 1999), where 39% equated the use of morphine in treating dyspnea sufficient to depress respiratory drive, with euthanasia. (Moreover, 22% agreed that an intentional morphine overdose was the most humane treatment for terminal ALS.)

Even if pain or dyspnea is acute or severe enough that the doses required could result in clinically relevant respiratory suppression, the prevention of suffering is the more important goal. The principle of "double-effect" refers to such a scenario – when an action can have two morally opposite effects. The criteria used by philosophers and theologians (succinctly discussed in Bernat, 2001) support the use of narcotics and anxiolytic agents at doses to relieve suffering even at the risk of hastening death.

These studies reflect the prominence given to the concern of probable life shortening effects of palliative medications. The respiratory depressant effects of medications used for relief of pain and anxiety are susceptible to the development of tolerance; thus early and appropriate use of these medications can lead to doses that might be lethal to a naïve patient, but are well-tolerated by the patient well-palliated. Studies in terminal cancer patients, including those with lung involvement, have shown that morphine can relieve symptoms without altering respiratory parameters, even in the elderly. (summarized in Borasio et al, 2004). Thus, when titrated for symptom relief, opiates are unlikely to hasten death.

Insomnia was a significant symptom in the last month of life (Ganzini et al, 2002a) and may reflect nocturnal hypoventilation (Borasio et al, 2004). Symptomatic relief may be obtained with the use of nocturnal non-invasive ventilation (NIV). Tricyclic antidepressant medications (e.g. amitriptyline) may offer benefit, but anxiolytics may be required as well. Reluctance to prescribe hypnotic sedation due to fear of respiratory depression may deprive these patients of a beneficial therapy.

The best outcome for those with a terminal disease is a "good death", defined by the National Institutes of Medicine as "...one that is free from avoidable distress and suffering for patients, families and caregivers, in general accord with patient and family wishes and reasonably consistent with clinical, cultural and ethical standards". Even with adherence to the AAN Practice Parameters with respect to PEG, NIV and hospice referral, symptom management was less than optimal in the last month of life (Ganzini et al 2002a); pain and insomnia were prominent. Yet, in another study, most deaths of patients with ALS, as reported by their caregivers, were peaceful (Neudert et al, 2001).

While interest in PAS may be higher in those with ALS than in other terminal disorders, still, the majority do not ultimately avail themselves of this option even where sanctioned. For most, life is too short, and the struggle to maintain quality of life in the face of disability can be overwhelming. Supportive counseling, informed decision-making and aggressive symptom management all have their role in easing the transitions throughout the course of the disease.

### **Summary and conclusions**

The discussion of end-of-life issues would appear to be fraught with ethical challenges and difficult choices. The potential for conflict exists, and may lead clinicians to avoid open discussion and advance care-planning. Yet people with ALS and their loved-ones can achieve good quality of life. Informing and guiding patients and their families through the decision-making process to a peaceful death should be integral to medical practice.

### **References**

1. Albert SM, Murphy PL, Del Bene ML, Rowland LP, Prospective study of palliative care in ALS: choice, timing, outcomes. *J Neurol Sci* 1999;169:108-113
2. Albert SM, Del Bene ML, Rabkin JG, Tider T, O'Sullivan I, Mitsumoto H, The decision to hasten death in people with ALS. *ALS & Other Motor Neuron Dis.* 2003; 4(Supp1):39
3. American Academy of Neurology Ethics and Humanities Subcommittee: Assisted Suicide, euthanasia, and the neurologist. *Neurology* 1998;50:96 – 598
4. Bernat J, Ethical and legal issues in palliative care. *Neurol Clin.* 2001;19:969-87

5. Borasio GD, Sloan R, Pongratz DE. Breaking the news in amyotrophic lateral sclerosis. *J Neurol Sci*. 1998;160(suppl 1):127-133
6. Borasio GD, Voltz R, Discontinuation of mechanical ventilation in patients with amyotrophic lateral sclerosis. *J Neurol*. 1998;245:717-22
7. Borasio GD and Miller RG, Clinical characteristics and management of ALS. *Sem Neurol* 2001;21:155-166
8. Borasio GD, Lyall R, Kaub-Wittemer D, Respiratory Symptoms, in "Palliative Care in Neurology" Voltz R, Bernat J, Borasio GD, Maddocks I, Oliver D & Portenoy R eds, Oxford University Press 2004
9. Bradley WG, Anderson F, Bromberg M, Guttman L, Harati Y, Ross M, Miller RG. Current management of ALS: comparison of the ALS CARE Database and the AAN Practice Parameter. *Neurology* 2001;57:500-5004.
10. Carver AC, Vickrey BG, Bernat JL, et al. End-of-life care: a survey of US neurologists' attitudes, behaviour, and knowledge. *Neurology* 1999;53:284-293
11. Del Bene M, Albert SA, Maxfield RA, Rowland LP. Forced Vital Capacity: Implications for hospice care. Poster presentation 9<sup>th</sup> International ALS/MND Symposium 1998
12. Dobscha SK, Heintz RT, Press N, Ganzini L. Oregon physicians' responses to requests for assisted suicide: A qualitative study. *Journal of Palliative Medicine* 2004; 7:450-460
13. Ganzini L, Block S, Physician-assisted death – a last resort? *N Engl J Med* 2002;346:1663-1665
14. Ganzini L, Nelson HD, Schmidt TA, Kraemer DF, Delorit Mam Lee MA. Physicians' experience with the Oregon Death with Dignity Act. *N Engl J Med* 2000;342:557-563
15. Ganzini L, Johnston WS, McFarland BH, Tolle SW, Lee MA, Attitudes of patients with amyotrophic lateral sclerosis and their caregivers toward physician-assisted suicide. *N Engl J Med* 1998;339:967-973
16. Ganzini L, Johnston WS, Silveira MJ. The final month of life in patients with ALS. *Neurology* 2002a;59:428-431
17. Ganzini L, Silveira MJ, Johnston WS. Predictors and correlates of interest in assisted suicide in the final month of life among ALS patients in Oregon and Washington. *J Pain Symptom Manage* 2002b;24: 312-317
18. Ganzini L, Dobscha SK, Heintz RT, Press N. Oregon physicians' perceptions of patients who request assisted suicide and their families. *Journal of Palliative Medicine* 2003; 6:381-390
19. Kaub-Wittemer D, von Steinbuchel N, Wasner M, Laier-Groeneveld G, Borasio GD, Quality of life and psychosocial issues in ventilated patients with amyotrophic lateral sclerosis and their caregivers, *J Pain Symptom Manage* 2003;26:890–896
20. Loyal L, The case for physician-assisted suicide and active euthanasia in amyotrophic lateral sclerosis. In: Brown RH, Meiningner V, Swash M eds. *Amyotrophic Lateral Sclerosis*. London: Martin Dunitz, :2000:423-439
21. McCluskey L, Casarett D, Siderowf A. Breaking the news: A survey of ALS patients and their caregivers. *ALS and other Motor Neuron Dis*. 2004;5:131-135
22. McCluskey L., Houseman G.: ALS Medicare hospice criteria: A Need for Improvement. *J Palliative Med* 2004;7(1): 47-53
23. McInerney F, "requested death": a new social movement, *Social Science and Medicine* 2000;50:137-154
24. Miller RG, Rosenberg JA, Gelinas DF, et al. ALS Practice Parameters Task Force. Practice parameter: The care of the patient with amyotrophic lateral sclerosis (an evidence-based review). Report of the quality standards subcommittee of the American Academy of Neurology. *Neurology* 1999; 52:1311-1323

25. Mitsumoto H, ALS Peer Workgroup Members. ALS: Completing the continuum of ALS care: A consensus document. Missoula, Montana: Promoting Excellence in End-of-Life Care, a national program of The Robert Wood Johnson Foundation, 2004 [http://www.promotingexcellence.org/als/als\\_report](http://www.promotingexcellence.org/als/als_report)
26. Mitsumoto H, Bromberg M, Johnston W et al. Promoting excellence in End-of-life care in ALS. ALS and other Motor Neuron Dis. (in press)
27. Moss AH, Casey P, Stocking CB, Roos RP, Brooks BR, Siegler M. Home ventilation for amyotrophic lateral sclerosis: outcomes, costs and patient, family and physician attitudes. *Neurology* 1993;43:438-443
28. Neudert C, Oliver D, Wasner M, Borasio GD. The course of the terminal phase in patients with amyotrophic lateral sclerosis. *J. Neurol.* 2001;248:612-616
29. Neudert C, Wasner M, Borasio GD. Attitudes towards life-prolonging treatments and active euthanasia in German patients with amyotrophic lateral sclerosis. *ALS and other Motor Neuron Dis.* 2003; 4(Supp1):41
30. Oppenheimer EA. Respiratory management and home mechanical ventilation in amyotrophic lateral sclerosis. In Mitsumoto H, Norris F eds. *Amyotrophic Lateral Sclerosis*. New York : Demos, 1994:139-162
31. Sullivan AD, Hedberg K, Hopkins D. Legalized physician-assisted suicide in Oregon 1998-2000. *N Eng J Med* 2001;344:605-7
32. Sullivan KE, Hebert PC, Logan J, O'Connor AM, McNeely PD. What do physicians tell patients with end-stage COPD about intubation and mechanical ventilation? *Chest* 1996;109:258-64
33. Veldink JH, Wokke JHJ, van der Wal G, de Jong JMBV, van den Berg LH. Euthanasia and physician-assisted suicide among patients with amyotrophic lateral sclerosis in the Netherlands. *N Eng J Med* 2002;346:1638-1644

Resources:

"Palliative Care in Amyotrophic Lateral Sclerosis" Oliver D, Borasio GD, Walsh D eds. Oxford University Press 2000

"Palliative Care in Neurology" Voltz R, Bernat J, Borasio GD, Maddocks I, Oliver D & Portenoy R eds, Oxford University Press 2004

"Completing the continuum of ALS care: A consensus document". Missoula, Montana: Promoting Excellence in End-of-Life Care, a national program of The Robert Wood Johnson Foundation, 2004 [http://www.promotingexcellence.org/als/als\\_report](http://www.promotingexcellence.org/als/als_report)

# End-of-life Decision-making in Amyotrophic Lateral Sclerosis

---

**Wendy Johnston MD, FRCPC**  
**Associate Professor**  
**University of Alberta**  
**Edmonton, Alberta**  
**Canada**

## Amyotrophic Lateral Sclerosis

---

- Mortality
    - 50% survival at 3 years
    - 20% survival at 5 years
    - 10% survival at 10 years
  - Most with ALS die as a result of the disease
  - Some measures improve survival, treatment is overall palliative
-

## End-of-life Decision-making

---

- Decisions with respect to life sustaining interventions
    - “Advance care planning”
    - “Advance directives” a.k.a. “Living Will”
  - Decisions about care at the end of life
    - Timing, manner and location of dying
- 

## Parallel Social/Medical Movements

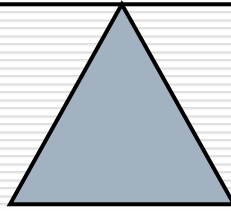
---

- | I                                       | II  | III                                      |
|---|---|--|
| □ Advances in life sustaining treatment | □ Resurgence of right-to-die societies      | □ Emergence of modern hospice movement   |
| □ Development of intensive care units   | □ Right to refuse life-sustaining treatment | □ Palliative care as a medical specialty |
| □ Respiratory support                   | □ Legalization of assisted death            | □ Comfortable death as a medical goal    |
| □ Enteral feeding                       |   |  |
-

# Breaking the News

## HOPEFULNESS

- Be hopeful yourself
- Stepwise disclosure
  - "Warning shot"
  - Allow time for questions
- Identify short term goals
- Explore emotional impact
  - Provide for quick follow-up
- Define direction and plan

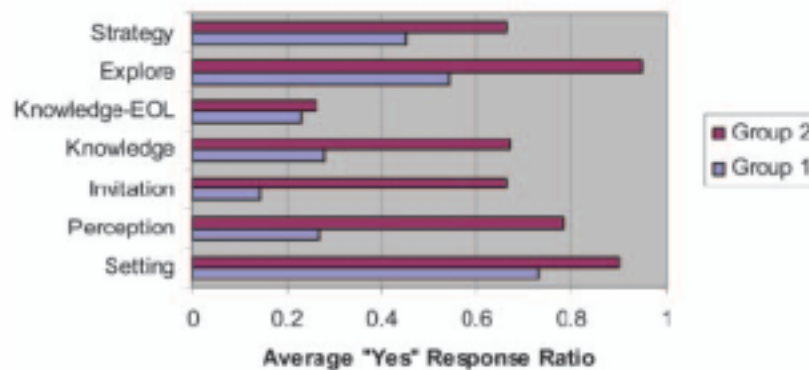


## REALISM

- Ask about current knowledge & correct misinformation
- Give clear information using common terms
- Acknowledge serious long term issues,
- Avoid absolute timelines
- Decisions to be made as conditions evolve

# McCluskey et al (2004)

Patient Assessment by Domain

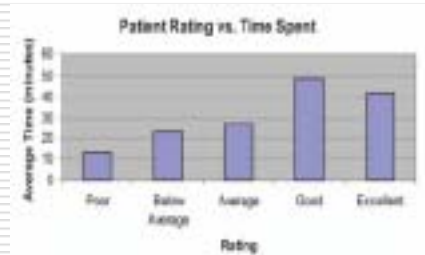


- Comparison of doctors rated as good-excellent (group 2) vs. average – poor (group 1) in breaking the news

## McCluskey et al (2004)

---

- Time spent in breaking the news valuable
- Excellent communication may not take longer



## Six Triggers for Discussion of EOL

---

1. The patient or family asks
2. Severe psychological, social or spiritual distress or suffering
3. Pain requiring high doses of analgesic medication
4. Dysphagia requiring feeding tube
5. Dyspnea or symptoms of hypoventilation, or FVC of 50% or less
6. Loss of function in 2 body regions

---

RWJ working group

## ALS and Life-sustaining Treatments

- ❑ Progressive weakness, including muscles of swallowing and breathing
- ❑ Death caused by respiratory failure
- ❑ Assisted ventilation can extend life without affecting progression
- ❑ Enteral feeding improves quality of life and can improve life expectancy

## ALS and Respiratory Support

- ❑ Death usually a result of respiratory insufficiency (hypoventilation or pneumonia)
- ❑ Non-invasive ventilation (NIV)
  - a palliative measure that may prolong life
- ❑ Tracheostomy and long-term ventilation (LTV)
  - extends life, although not indefinitely

## Choice of Life Sustaining Treatment

I. Passive: no plan

II. Active planning for intervention

- Monitor swallowing, pulmonary symptoms and function,
  - offer PEG when comfort with eating or nutrition compromised, or FVC 50% predicted
  - offer NIV (discuss LTV) when symptomatic or FVC 50% predicted

## Choice of Life Sustaining Treatment

III. Active planning to forgo interventions

- Explicitly discuss symptoms and scenarios at end of life
- plan hospice referral
- involve proxy decision maker

## Barriers to Decision-making

---

- TIME
  - Experience
  - Denial (of diagnosis, of progression)
    - Useful coping strategy?
    - Impediment to care?
    - Executive dysfunction?
- 

## Barriers to Decision-making

---

- Cultural Taboos
    - Disclosure of diagnosis and prognosis
    - Explicit discussions of dying
    - Dying as a "choice"
  - Framing
    - Context and wording of question
    - Values and experience of clinician (explicit or implicit)
-

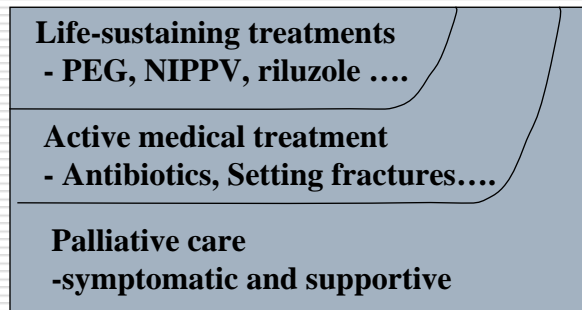
## Advance Care Planning

- Practice parameters: Recommendation that all patients prepare advance directives conveying decisions regarding life sustaining therapies (Miller et al, Neurology, 1999)
- High compliance (90% patients had AD, 97% respected) reported in ALS CARE database (Bradley et al 2001)

## ALS and Palliative Care

- Provision for ALS patients in Cicely Saunders's Hospice from the start
- Development of palliative care field focused mostly on cancer
- Recent recognition of need for improved end-of-life care in non-malignant conditions
- ALS first neurological disorder to receive specific attention in advancing end-of-life care

## Integrated Palliative Care in ALS



Diagnosis

Death

Adapted from Oliver in  
"Palliative Care in Amyotrophic Lateral Sclerosis", Oxford Press, 2001

## Palliative Care and Hospice



## Hospice Referral

---

- Medicare Criteria stringent, complicated and not representative of terminal ALS
  - Rely on FVC
  - 5 of 97 patients referred to hospice met criteria (McCluskey and Houseman 2004)
    - 91% died in < 180 days
    - Average 84 days (1 – 534)
- 

## Pain and Insomnia in ALS

---

- Survey (retrospective) in last month of life (Ganzini et al 2002)
    - 50 patients, high adherence to AAN practice parameters except
    - Levels of pain, insomnia were high
  - Survey of U.S. neurologists (Carver et al 1999)
    - 39% equated the use of morphine in treating dyspnea sufficient to depress respiratory drive, with euthanasia
-

## The Double-effect

- Refers to an action that can have two morally opposite effects
- The criteria used by philosophers and theologians support the use of narcotics and anxiolytic agents at doses to relieve suffering even at the risk of hastening death (succinctly discussed in Bernat, 2001)

## Medicare Criteria: Determination of Terminal Status in ALS

Patients will be considered to be in the terminal stage of ALS (life expectancy of six months or less) if they meet the following criteria. (Must fulfill 1, 2 or 3)

1. Patient must demonstrate critically impaired breathing capacity.
  - a. Critically impaired breathing capacity as demonstrated by *all* of the following characteristics occurring within the 12 months preceding initial hospice certification:
    - i. Vital capacity (VC) < 30% of normal;
    - ii. Significant dyspnea at rest
    - iii. Requiring oxygen supplementation at rest
    - iv. Patient declines invasive ventilation.

## Medicare Criteria: Determination of Terminal Status in ALS

---

2. Patient must demonstrate **both** rapid progression of ALS **and** critical nutritional impairment:
    - a. Rapid progression of ALS as demonstrated by **all** of the following characteristics occurring within the 12 months preceding initial hospice certification:
      - i. Progression from independent ambulation to wheelchair, or to bed bound status
      - ii. Progression from normal to barely intelligible or unintelligible speech
      - iii. Progression from normal to pureed diet
      - iv. Progression from independence in most or all activities of daily living to needing major assistance by caretaker in most all
- 

## Medicare Criteria: Determination of Terminal Status in ALS

---

- b. Critical nutritional impairment as demonstrated by **all** the following characteristics occurring within the 12 months preceding initial hospice certification:
    - i. Oral intake of nutrients and fluids insufficient to sustain life
    - ii. Continuing evidence of weight loss
    - iii. Dehydration or hypovolemia
    - iv. Absence of artificial feeding methods
-

## Medicare Criteria: Determination of Terminal Status in ALS

---

3. Patients must demonstrate *both* rapid progression of ALS *and* life-threatening complications.
    - a. Rapid progression of ALS, see 2.a. above
    - b. Life-threatening complications as demonstrated by *one* of the following characteristics occurring within the 12 months preceding initial hospice certification:
      - i. Recurrent aspiration pneumonia (with or without tube feedings)
      - ii. Upper urinary tract infection, e.g. pyelonephritis
      - iii. Sepsis
      - iv. Fever recurrent after antibiotic therapy
      - v. Decubitus ulcers, multiple stage 3-4, particularly if infected
- 

## Hospice Qualifying Symptomatology (Columbia University)

---

- (A) or (B) must be present. (A) does not require associated symptoms. If just (B), at least 2 other Respiratory Indicators must be present or 1 respiratory and 1 Nutritional Indicator present.
    - (A) FVC less than or equal to 30% predicted
    - (B) FVC less than or equal to 60% predicted with a steady decline over past 2-3 months
-

## Hospice Qualifying Symptomatology (Columbia University)

---

- Other Respiratory Insufficiency Indicators:
    - Shortness of Breath
    - Shallow Breathing
    - Paradoxical Breathing
    - Inability to be supine due to diaphragmatic weakness
    - Non-explosive cough
    - Inability to blow nose
    - Breath Support poor as evidenced by accessory muscle use
    - Endurance poor (excessive fatigue)
    - Significant insomnia related to diaphragmatic weakness
- 

## Hospice Qualifying Symptomatology (Columbia University)

---

- Nutritional Insufficiency Indicators:
    - Excessive oral secretions with dysphagia
    - Nutritional Compromise/Dehydration (with or without feeding tube) Weight Loss > 10% of body weight and refusing PEG
    - Aspiration or choking (on food or liquid)
  - Standard Requirements
    - Prognosis of 6 months or less
    - Patient election of Hospice services
    - Patient election of DNR status
    - Patient refusal of Invasive/Biomedical Intervention
    - Patient is Home Care Eligible
-

## Proposed Penn ALS Hospice Criteria

Patients are considered eligible for hospice care if they do not elect for tracheostomy for invasive ventilation and display evidence of critically impaired respiratory function (with or without use of NIPPV) and / or severe nutritional insufficiency (with or without use of a gastrostomy tube):

## Proposed Penn ALS Hospice Criteria

- A. Critically impaired respiratory function is as defined by:
  - 1. FVC < 40% predicted (seated or supine) and 2 or more symptoms and / or signs (list)
  - 2. If unable to perform the FVC test patients meet this criterion if they manifest 3 or more of the above symptoms / signs.
- B. Severe nutritional insufficiency is defined as:
  - Dysphagia with progressive weight loss of at least five percent of body weight with or without election for gastrostomy tube insertion.

## Respiratory symptoms/signs

---

- Dyspnea at rest
- Orthopnea
- Accessory respiratory musculature use
- Paradoxical abdominal motion
- Respiratory rate > 20
- Reduced speech / vocal volume
- Weakened cough
- Symptoms of sleep disordered breathing
- Frequent awakening
- Daytime somnolence / excessive daytime sleepiness
- Unexplained headaches
- Unexplained confusion
- Unexplained anxiety
- Unexplained nausea

Proposed Penn ALS Hospice Criteria

---

## Major Components of a "Good Death"

---

- Effective pain and symptom management
- Ability to engage in clear decision making
- Preparation for death
- A sense of completion
- Continued ability to contribute to others
- Affirmation of the whole person

*Data from* Steinhauser KE, Clipp EC, McNeilly M, Christakis NA, McIntyre LM, Tulsky JA. In search of a good death: observations of patients, families, and providers. *Ann Intern Med* 2000;132:827

---

## ALS and the Right-to-die Movement

- Patients with ALS prominent in court appeals for legalized assisted suicide
- Broadcast of assisted deaths of ALS patients in U.S.A. and Netherlands

## ALS and the Right-to-die Movement

- Studies indicating high (Ganzini et al 1998) and sustained interest (Ganzini et al 2002)
- Disproportionate use of assisted death in patients with ALS where legal or sanctioned (Sullivan et al 2001, Veldink et al 2002)

## Ethics and PAS/euthanasia

- Ethical & legal basis for patient autonomy in medical decision-making
- No legal or moral obligation to provide PAS/euthanasia
- Withdrawal of life-sustaining therapies not PAS/euthanasia

## Ethics and PAS/euthanasia

- Sanctioned physician-assisted death
  - Physician-assisted suicide (PAS) in Oregon, U.S.A.
  - Euthanasia and PAS in Netherlands, Belgium, Switzerland