

# End-of-Life Decision Making in 42 Patients With Amyotrophic Lateral Sclerosis

Craig A Munroe MD, Michael D Sirdofsky MD, Tunay Kuru MD, and Eric D Anderson MD

**OBJECTIVE:** To determine when end-of-life issues were discussed with patients afflicted with amyotrophic lateral sclerosis (ALS). **METHODS:** This was a retrospective analysis of ALS patients referred to the neuromuscular clinic at Georgetown University Hospital. Patients were seen by a pulmonologist and a neurologist at the initial diagnosis or referral, and every 2–3 months thereafter. End-of-life discussions were addressed at each visit. Other variables recorded included the amount of time afflicted with ALS, serial pulmonary function test results, and the subjective level of bulbar dysfunction. **RESULTS:** We saw 43 patients (age range 39–94 y) between June 1999 and September 2004. One patient was on a ventilator at the initial visit, and was therefore excluded from the study. Discussion about the patients' end-of-life care preferences were initiated at the first pulmonary visit with 40 patients. With 2 patients, end-of-life decisions were discussed at the second office visit. Twenty-five patients chose do-not-resuscitate and do-not-intubate (DNR/DNI) orders after the initial end-of-life discussion with the pulmonologist. Five other patients chose DNR/DNI orders during subsequent clinic visits. Four patients were still undecided at their last clinic visit. Six patients were lost to follow-up before a decision was made. Two patients requested full ventilatory support. Both the forced vital capacity and the level of bulbar dysfunction were not statistically different between the patients who chose DNR/DNI and the patients who were either undecided or requested full ventilatory support. **CONCLUSIONS:** Decisions about end-of-life care are often delayed in patients with ALS. These patients' final decisions seem to be independent of their level of respiratory insufficiency or bulbar function, and most related to the physician addressing end-of-life care decisions in a timely manner. *Key words:* amyotrophic lateral sclerosis, end-of-life, do-not-resuscitate, do-not-intubate, DNR, DNI, pulmonary functions testing, bulbar dysfunction. [Respir Care 2007;52(8):996–999. © 2007 Daedalus Enterprises]

## Introduction

Amyotrophic lateral sclerosis (ALS) is a debilitating neuromuscular disease that affects both upper and lower

motor neurons. It is the most common form of motor neuron disease, and is thought to affect 1.2–1.8/100,000 individuals.<sup>1</sup> ALS is an age-dependent disorder; the incidence increases with advancing age.<sup>1</sup> Clinically, ALS is characterized by muscular weakness and atrophy. Interestingly, intellect, sensation, and awareness remain essentially unchanged.<sup>2</sup> Death almost universally occurs within 3–5 years and is most typically secondary to worsening restrictive lung disease, with resultant respiratory failure.<sup>3</sup>

Medical advances such as invasive mechanical ventilation, aggressive nutritional interventions (such as percuta-

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At the time of this research, Craig A Munroe MD was affiliated with the Department of Pulmonary, Critical Care, and Sleep Medicine, Georgetown University, Washington, District of Columbia. He is now affiliated with the Department of Medicine, Massachusetts General Hospital, Boston, Massachusetts. Tunay Kuru MD and Eric D Anderson MD are affiliated with the Department of Pulmonary, Critical Care, and Sleep Medicine; and Michael D Sirdofsky MD is affiliated with the Department of Neurology, Georgetown University Hospital, Washington, District of Columbia.

The authors report no conflicts of interest related to the content of this paper.

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Correspondence: Craig A Munroe MD, Department of Medicine, Massachusetts General Hospital, 55 Fruit Street, Boston MA 02114. E-mail: camunroe@partners.org.

nous endoscopic gastrostomy tube placement), and, most recently, noninvasive ventilation measures offer these patients more options at the end of life.<sup>1</sup> Such interventions continue to delay the time to death in patients with ALS, which is increasing the prevalence rate of this disorder.<sup>1</sup>

Though there is a large body of literature regarding end-of-life issues in patients with cancer and human immunodeficiency virus/acquired immune deficiency syndrome, there is little published information regarding end-of-life decisions in patients afflicted with ALS. There remains no consensus on when and how end-of-life issues should be addressed with these patients. The relatively gradual progression of ALS allows the patient and his or her family time to contemplate and participate in end-of-life decisions, especially when these issues are addressed in a timely manner.<sup>3</sup> Smyth et al found that approaches to end-of-life decision making in patients with ALS differ because of historical, cultural, and economic differences between patients.<sup>3</sup> However, there has been little mention in the medical literature of when end-of-life issues should be addressed with patients afflicted with ALS.

### Methods

We conducted a retrospective chart review of all patients referred to the Muscular Dystrophy Association multidisciplinary clinic at our hospital, which is a tertiary-care referral center. The study was approved by the institutional review board of Georgetown University.

The diagnosis of ALS was confirmed by attending neurologists. At each office visit, patients were evaluated by a single pulmonologist from the Division of Pulmonary, Critical Care, and Sleep Medicine. Patients were seen at the initial diagnosis (or referral to the Muscular Dystrophy Association clinic with previously documented ALS) and every 2–3 months thereafter. During these office visits, documentation of discussion of end-of-life issues, mechanical ventilation preferences, and code status (do-not-resuscitate [DNR] and/or do-not-intubate [DNI]) was recorded in each patient's chart. Other factors recorded included the amount of time the patient had been afflicted with the disease, pulmonary function as documented by forced vital capacity (FVC), and the patient's bulbar function at the time of their decision. Bulbar function was defined as firm glottis closure that allowed near-normal speech and swallowing, and was measured subjectively via a "yes" or "no" characterization of their symptoms. Patients already on invasive mechanical ventilation were excluded from the study. Invasive ventilation was defined only as intubation or tracheostomy with mechanical ventilation, not as bi-level or continuous positive airway pressure. All statistical analyses were performed with analysis of variance single-factor and Fisher's exact test.

### Results

Between June 1999 and September 2004, a total of 43 patients were treated. The patients' age range was 39–94 years, there were 19 males and 24 females, 28 patients were white, 10 were African American, 21 were married, and they had a wide range of baseline lung function, as measured by FVC. End-of-life discussions were initiated at the first pulmonary visit with 40 patients (95.2%). With 2 patients, end-of-life decisions were discussed at the second office visit. One patient was on a ventilator at the initial visit and was therefore excluded from the study.

Twenty-five patients (59.5%) decided to be DNR/DNI if their lung function deteriorated after the initial end-of-life discussion with the pulmonologist. Five other patients (11.9%) chose DNR/DNI status during subsequent clinic visits. Four patients (9.5%) were still undecided at their last clinic visit. Six patients (14.3%) were lost to follow-up before a decision was made. Two patients (4.8%) requested full ventilatory support.

Of the 30 patients who decided to be DNR/DNI, 20 (66.7%) had bulbar dysfunction. Both patients who requested invasive ventilatory support had bulbar dysfunction, as did 2 of 4 undecided patients (difference nonsignificant). The median percent-of-predicted FVC of the 30 patients who decided against invasive ventilatory support was 50% (range 10–118%), whereas the 4 patients who were undecided had a median percent-of-predicted FVC of 40% (range 26–55%) (difference nonsignificant). The 2 patients who requested invasive ventilation had a median percent-of-predicted FVC of 56% (range 35–77%).

Of the patients who had DNR/DNI orders, the median time between diagnosis and code status decision was 6.0 months (range 0–96 months). However, 83% made this decision at the initial visit. Those who were still undecided at the end of the study period had had ALS for a median of 5 months (range 0–72 months).

### Discussion

End-of-life decision making in patients with ALS is a critical topic for physicians to address. Research has shown that patients with ALS generally welcome the opportunity to discuss end-of-life issues with their physicians.<sup>2,4</sup> Our study supported these findings. Patients were largely receptive to our pulmonologist discussing end-of-life choices at the first clinic visit. Silverstein et al found that, though 81% of patients felt that their physician wanted to know their preferences about life-sustaining treatment, only 8 patients in their cohort of 38 had discussed this topic with their physician.<sup>2</sup> Thus, there remains a discord between patient wishes and physician practices regarding end-of-life issues in patients with ALS.

Although the median time from diagnosis to the DNR/DNI decision was 6 months, 83% of our patients made the decision against invasive ventilatory support at their initial visit. This suggests that patients with ALS have considered their end-of-life wishes before the topic is broached by their physicians.

We believe that code status should be discussed early in the disease course, once the diagnosis is made. Our opinion is that patients are not only responsive to this, but that the issue is crucial in a disease with such unpredictable month-to-month progress. Obviously, no end-of-life discussion should take place before the diagnosis of ALS is confirmed by a neurologist. Once confirmed, however, the worst-case scenario is for a patient with respiratory failure to undergo emergency intubation against his or her wishes because there was never any discussion of what this decision entails.

Our policy about early discussion of end-of-life care has been corroborated by other investigators.<sup>5</sup> A patient's preferences may change over the course of the disease, so, as proposed by Albert and colleagues,<sup>6</sup> we recommend continued clinical education and open discussion with patients during office visits.

Pulmonary and critical care physicians can serve an important role in patients stricken with ALS. Most often the terminal event in these patients involves progressive respiratory failure, as documented by a worsening FVC. This worsening restrictive lung defect necessitates close collaboration between the neurologist, who typically is the primary physician, and the pulmonologist, who is able to quantify the extent of the patient's respiratory compromise. With this in mind, this retrospective analysis seems to point toward another crucial role of the pulmonologist in the care of patients with ALS. Though we believe that early discussions with any health care professional can benefit patients with ALS, because the pulmonologist/intensivist has extensive experience in dealing with terminally ill patients, the pulmonologist is perhaps the most appropriate physician to address end-of-life issues with these patients. However, most neuromuscular centers do not have a pulmonologist on staff to see all ALS patients, and this difference may be reflected in our results. Physicians of all types can address these issues, with proper training. Carver et al<sup>7</sup> described a gap between legal, medical, and ethical guidelines about end-of-life issues and the practices of many neurologists. That study suggested the need for graduate and postgraduate education programs for neurologists in the practices and principles of palliative care medicine.<sup>7</sup> In ALS, however, because of its intimate relation with lung function, the pulmonary physician's expertise allows him or her to give the patient and family insight into what to expect if the patient chooses invasive life-prolonging measures.

An important factor in whether an ALS patient decides for or against invasive measures is whether the physician addresses the issue in a timely manner. The patient's functional status may decline rapidly and unpredictably from one office visit to another, and the patient may be unable to communicate his or her wishes at a subsequent visit, so discussing end-of-life decisions early in the disease course is advisable. The patient should be offered continued education about prognosis and palliative care.

Factors that seemed to have little influence in end-of-life decision making include time to definitive diagnosis, bulbar dysfunction at the time of the decision, and the severity of the restrictive lung condition. Silverstein et al also found that ALS patients' responses were independent of functional status.<sup>2</sup> Two patients in that retrospective analysis elected invasive ventilatory support, and one patient was already on a ventilator. Our findings here are similar to those of Albert et al: 6–12% of their patients were certain they wanted tracheostomies at the end of the study period.<sup>6</sup>

Possible limitations of the present study include the fact that it was a retrospective analysis. Additionally, since all of the end-of-life discussions were conducted by only one pulmonary physician, it is possible that this physician's personal feelings about invasive life support measures may have altered some patients' thinking to some extent, although efforts were taken to minimize the effect of personal bias. Moss and colleagues found, in 1993, that physician bias can affect end-of-life decision making in ALS patients.<sup>8</sup>

## Conclusions

End-of-life decision making in patients with ALS is often delayed. Final decisions seem to be independent of the patient's level of respiratory insufficiency or bulbar function. The most important factor in whether the ALS patient decides for or against invasive measures is whether the physician addresses these issues in a timely manner and provides continued education about prognosis and the available palliative measures. The pulmonologist can play a valuable role in end-of-life discussions in patients with ALS.

## REFERENCES

1. Strong M, Rosenfeld J. Amyotrophic lateral sclerosis: a review of current concepts. *Amyotroph Lateral Scler Other Motor Neuron Disord* 2003;4(3):136–143.
2. Silverstein MD, Stocking CB, Antel JP, Beckwith J, Roos RP, Siegler M. Amyotrophic lateral sclerosis and life-sustaining therapy: patients' desires for information, participation in decision making, and life-sustaining therapy. *Mayo Clin Proc* 1991;66(9):906–913.

## END-OF-LIFE DECISION MAKING IN 42 PATIENTS WITH AMYOTROPHIC LATERAL SCLEROSIS

3. Smyth A, Riedl M, Kimura R, Olick R, Siegler M. End of life decisions in amyotrophic lateral sclerosis: a cross-cultural perspective. *J Neurol Sci* 1997;152 Suppl 1:S93–S96.
4. Benditt JO, Smith TS, Tonelli MR. Empowering the individual with ALS at the end-of-life: disease-specific advance care planning. *Muscle Nerve* 2001;24(12):1706–1709.
5. Mandler RN, Anderson FA Jr, Miller RG, Clawson L, Cudkowicz M, Del Bene M; ALS C.A.R.E. Study Group. The ALS Patient Care Database: insights into end-of-life care in ALS. *Amyotroph Lateral Scler Other Motor Neuron Disord* 2001;2(4):203–208.
6. Albert SM, Murphy PL, Del Bene ML, Rowland LP. A prospective study of preferences and actual treatment choices in ALS. *Neurology* 1999;53(2):278–283.
7. Carver AC, Vickrey BG, Bernat JL, Keran C, Ringel SP, Foley KM. End-of-life care: a survey of US neurologists' attitudes, behavior, and knowledge. *Neurology* 1999;53(2):284–293.
8. Moss AH, Casey P, Stocking CB, Roos RP, Brooks BR, Siegler M. Home ventilation for amyotrophic lateral sclerosis patients: outcomes, costs, and patient, family, and physician attitudes. *Neurology* 1993;43(2):438–443.



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