

Original Article

Medical and Supportive Care Among People with ALS in the Months Before Death or Tracheostomy

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Abstract

People with amyotrophic lateral sclerosis (ALS) who choose tracheostomy demonstrate a strong and mostly consistent attachment to life from the point of diagnosis. It is unclear if these patients also use medical and health services to a greater degree than patients who decide against tracheostomy. In this research, patients with a high likelihood of dying over six months (forced vital capacity < 50% predicted) were followed monthly until death or tracheostomy with long-term mechanical ventilation (LTMV). Patient service use was measured by caregiver reports of 1) ALS-specific prosthetic devices, 2) allied health or medical services, 3) legal preparation for medical care or the end of life, and 4) medical care episodes. Caregivers also reported all patient prescription medications. At follow-up, 57 patients died and 14 elected to have tracheostomy and LTMV. Patients who opted for LTMV were younger and had higher household incomes. They were significantly more likely to use nasal ventilation, paid home care, and family or personal counseling over follow-up, and they were also more likely to remain on medications. The proactive orientation to health and desire to live despite severe disability reported for people choosing LTMV thus extends as well to more intensive use of medical and supportive care in the months before tracheostomy. A challenging task for clinicians is to acknowledge this strong desire to live while providing appropriate expectations for life after tracheostomy. J Pain Symptom Manage 2009;38:546–553. © 2009 U.S. Cancer Pain Relief Committee. Published by Elsevier Inc. All rights reserved.

Key Words

Amyotrophic lateral sclerosis, palliative care, mechanical ventilation, service use, tracheostomy, mental health, decision making

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Introduction

The choice of tracheostomy and long-term care in people with amyotrophic lateral sclerosis (ALS) remains relatively rare but shows wide variation across treatment sites (1.4%–15% in the United States)¹ and countries (45% in Japan).² It also may be increasing. In a cohort followed across the last six months of life at one multidisciplinary ALS clinic, 18% opted for tracheostomy and long-term mechanical ventilation (LTMV).³ Prior research has examined psychosocial and demographic correlates of this choice. Patients choosing tracheostomy are younger, more likely to have children in the home, less likely to report depressive symptoms, and more likely to demonstrate strong attachment to life at the point of diagnosis.^{4–6}

In this research, we asked if patients choosing tracheostomy also demonstrate greater medical and health service use relative to patients who decide against tracheostomy. Evidence of increased service use in the period before tracheostomy would indicate a consistent, continuing preference for treatment and prolongation of life in this subset of patients. Their orientation to disease management may accordingly differ from that of patients unwilling to consider tracheostomy. Patients who opt for tracheostomy may be more likely to seek alternative or experimental therapies, for example. They may also be more likely to report frustration with standard patient education and clinical management. In our experience, these patients seek more engagement by clinicians and report great frustration when they are told “go home, get your things in order, there is nothing you can do,” as one patient recently complained. This broad orientation may also lead to more strain within families, who must cope with disease progression and the prospect of post-tracheostomy care. For all these reasons, we sought to investigate whether patients opting for tracheostomy differ systematically in disease management over the course of disease progression.

Method

Sample

Over 90% of patients were enrolled from the Eleanor & Lou Gehrig MDA/ALS Research Center at Columbia University. Other patients

were referred from hospices, ALS support groups, or two other ALS multidisciplinary clinics. The clinic coordinator identified potential participants whose forced vital capacity was less than 50%, a value related to the risk of hospice admission and death or need for mechanical ventilation within six months.⁷ Eligible patients did not meet criteria for dementia, spoke English, had an unpaid caregiver who was available for interview, were not using mechanical ventilation at baseline, were able to communicate at least “yes” and “no,” and lived within a three-hour drive from the medical center. Study enrollment began in January 2000 and ended in June 2004. The Institutional Review Boards of Columbia-Presbyterian Medical Center and the New York State Psychiatric Institute approved the research protocol.

Procedures

After the clinic coordinator identified potential participants, she described the study to patients and caregivers and obtained consent for the research team to make contact. The principal investigator then called for further explanation, answered questions about the study, and obtained verbal consent; informed consent from patients and caregivers was obtained during a home visit, when interviews were conducted. Interviews were scheduled at one-month intervals until patients met a study endpoint of tracheostomy or death. Patients and caregivers completed a separate interview on the same schedule. Caregivers also completed an interview after the death of patients.

Measures

Patient service use was measured by caregiver reports of 1) ALS-specific prosthetic devices (augmentative communication, feeding gastrostomy [percutaneous endoscopic gastrostomy (PEG) placement], nasal ventilation, suction device); 2) allied health or medical services (paid home care, personal or family counseling, alternative/complementary medicine, clinical trial participation, hospice); 3) legal preparation for medical care or the end of life (health care proxy, living will, power of attorney, autopsy consent); and 4) medical care episodes (emergency department, hospital, or skilled nursing facility admissions). This

set of services captures the major options for disease management available to patients in later stages of ALS.⁸ Caregivers reported whether patients were using each service at monthly assessments.

Caregivers also reported patient prescription medications. They could report up to nine medications. Medications were categorized by drug indication, which yielded 26 medication categories. These were further reduced to four broad categories: 1) ALS-specific therapies (medications specifically designed to slow the progression of the disease: riluzole, high-dose vitamins, gabapentin, antioxidants, creatine, and clinical trial medications); 2) palliative medications (pharmacological therapies aimed at treating common symptoms of ALS, such as pain, sialorrhea, respiratory insufficiency, constipation, muscle spasms, cramping, and insomnia); 3) mood medications (medications aimed at treating depression, anxiety, agitation, and emotional lability, such as alprazolam, buspirone, bupropion, paroxetine, and sertraline); and 4) "other" (medications to treat non-ALS chronic conditions and comorbidities).

Use of each service and medication type was noted at study baseline and at the last assessment before LTMV or death. In addition, we calculated the proportion of patients who used each service or medication type across all assessments.

To assess the severity of ALS, patients completed the ALS Functional Rating Scale (ALSFRS-R)⁹ at each assessment. The ALSFRS consists of 12 items covering four domains: bulbar functions (speech, salivation, swallowing), fine motor skills (handwriting, handling utensils, dressing and hygiene), gross motor skills (turning in bed, walking, climbing stairs), and respiratory status (dyspnea, orthopnea, use of ventilation support). Each item is rated on a 5-point scale and a sum computed.

To assess the depressive symptoms, interviewers administered the Patient Health Questionnaire (PHQ).¹⁰ The PHQ assesses Diagnostic and Statistical Manual of Mental Disorders (DSM)-IV criteria for major and minor depressive disorder. Although the structured clinical interview for DSM-IV is the gold standard diagnostic evaluation in psychiatry, it requires interactive queries and follow-up probes. A full diagnostic interview was not feasible in this sample of patients with end-stage ALS, where 40%

were unable to speak at baseline.⁶ A clinical psychologist reviewed all PHQ assessments to assign DSM-IV depression diagnoses. Caregivers reported on patient cognition (memory, concentration, ability to follow a plot on television) and lability. Caregiver reports of patient cognitive status were validated against patient performance on the Mini-Mental State Exam (MMSE).¹¹ Patients whose caregivers reported memory problems performed more poorly on the MMSE, especially in word registration and recall.

Caregivers also reported the level of burden and satisfaction associated with care. We used five items from the Zarit Burden Scale.¹² These capture stress, not having time for oneself, and restriction in social and family life because of caregiving responsibilities. Satisfaction with caregiving commitment was assessed with an additional five items from Folkman's measure of caregiving satisfaction.¹³

Analyses

Patients choosing tracheostomy and those who died over follow-up were compared to determine if service use differed at baseline and over follow-up. Similar analyses were conducted for medication use. Proportions were compared using χ^2 to test for differences. Because the length of follow-up differed in the two patient groups, we used Mantel-Haenszel tests to stratify patients by the number of study assessments (1, 2–4, 5+) and also developed logistic regression models to assess group differences adjusting for demographic indicators, study duration, and disease status. For assessing change between baseline and last assessments, we developed analysis of variance models using repeated measures.

Results

Of 80 patients enrolled into the living with ALS cohort, three patients did not have unpaid family or friend caregivers and were excluded from analyses. An additional six patients were excluded because they were lost to follow-up ($n = 2$), did not die or elected to have LTMV during the course of the study ($n = 2$), or had caregivers who participated in a postdeath interview alone ($n = 2$). Excluding these nine left 71 patients with caregivers who reported on service and medication use in the

Table 1
Baseline Status: Patients Dying Without Mechanical Ventilation Compared with Patients Who Chose Tracheostomy/Long-Term Mechanical Ventilation

Baseline Status	Patients Dying Over Follow-Up (n = 57)	Patients Choosing Tracheostomy/LTMV (n = 14)
<i>Patient sociodemographic status</i>		
Age	64.5 ± 13.6	51.4 ± 12.4 ^a
Male, %	61.4	42.9
College, %	49.1	71.4
Nonwhite, %	8.8	14.3
Private insurance, %	75.0	84.6
Medicaid, %	17.5	8.3
Income > \$80,000/year	18.8	58.3 ^a
<i>Patient clinical status</i>		
ALSFRS-R	22.3 ± 8.1	21.3 ± 7.0
Any cognitive symptom, %	20.0	0.0
Lability, %	43.6	23.1
Major or minor depression, %	20.3	0.0+
Years since diagnosis	1.6 ± 1.2	2.2 ± 1.7
Years since first symptoms	2.5 ± 1.4	2.9 ± 1.6
<i>Caregiver status</i>		
Age	57.6 ± 15.3	56.1 ± 12.1
Female, %	77.2	64.3
College, %	56.1	85.7 ^b
Working, %	40.4	78.6 ^b
Spouse, %	60.7	71.4

^a $P < 0.01$.

^b $P < 0.05$, by t -test (continuous measures), χ^2 (proportions), or Kruskal-Wallis test (medians).

months before death or tracheostomy. Fifty-seven died and 14 elected to have LTMV. Two additional patients had unplanned emergency tracheostomies (one against her wishes) and died shortly after the procedure. These patients were not considered to have chosen tracheostomy.

Patients who died over follow-up had fewer days on study than patients who opted for tracheostomy and LTMV (163 vs. 253 days, $P = 0.13$; median of 110 vs. 123 days). The median number of monthly assessments was three among patients who died and four among patients who opted for mechanical ventilation. The mean time from first assessment to tracheostomy/LTMV was 364 days and to death 207 days ($P = 0.11$). Sixty of the 77 patients completed more than one monthly assessment. Based on caregiver reports of the date of diagnosis, our first assessment took place an average of 20 months (median: 17) after diagnosis.

Differences at Baseline Between Patients Dying without Mechanical Ventilation and Patients Opting for LTMV

At baseline, patients who died or chose mechanical ventilation differed in age (Table 1). Patients who later opted for LTMV were

younger (51.4 vs. 64.5, $P < 0.01$). The two groups did not differ in other sociodemographic features, including gender, race, education, insurance, or presence of spousal caregiver. However, patients choosing LTMV had significantly higher income, and their caregivers were more likely to be employed (78.6% vs. 40.4%, $P < 0.05$) and to have completed college (85.7% vs. 56.1%, $P < 0.05$). Patients in the two groups were similar in the level of disability, presence of cognitive symptoms, and time since diagnosis and symptom onset. Patients who later opted for LTMV were less likely to report symptoms consistent with major depression, but this difference did not achieve statistical significance.

Service Use in the Final Months of Life

Patients who later opted for LTMV were more likely to use medical and supportive care (Table 2). At baseline, 78.6% of the LTMV group used nasal ventilation, which increased to 92.9% across the follow-up period. In patients who died without LTMV, 51.8% used nasal ventilation at baseline and 62.5% over follow-up ($P < 0.05$). The LTMV group also was more likely to use supportive care, including personal or family counseling

Table 2
Service Use and Planning at Baseline and Follow-Up: Patients Dying Without LTMV and Patients Choosing LTMV

	In Place at Baseline (%)		Used Any Time at Follow-Up (%)	
	Death	LTMV	Death	LTMV
Augmentative communication	19.3	35.7	29.8	50.0
Counseling	24.6	42.9	45.1	78.6 ^a
Alternative/complementary medicine	21.1	28.6	28.1	57.1
PEG placement	33.9	57.1	39.3	57.1
Nasal ventilation	51.8	78.6	62.5	92.9 ^a
Clinical trial participation	21.4	42.9	21.4	50.0
Hospice	43.9	7.1 ^a	70.2	21.4 ^b
Paid home care	35.1	71.4 ^a	49.1	85.7 ^a
Suction device	41.8	50.0	58.2	57.1
Health care proxy	62.5	64.3	78.6	78.6
Living will	66.1	64.3	82.1	85.7
Power of attorney	71.4	78.6	83.3	81.3
Autopsy consent	7.1	0.0	16.7	0.0
Emergency department admission	30.4	30.8	39.3	38.5
Hospital admission	17.9	21.4	30.4	21.4
Skilled nursing home admission	5.4	7.1	8.9	7.1

^a $P < 0.05$.

^b $P < 0.01$ by χ^2 (baseline) and Mantel-Haenszel (ever use, adjusted for time on study) tests.

and paid home care. Use of feeding gastrostomies, augmentative communication devices, or alternative/complementary medicine did not significantly differ between the groups, nor did the groups differ in legal preparation for medical care or the end of life, as indicated by the proportion with a health care proxy, living will, power of attorney, or autopsy consent.

Patients who opted for LTMV were less likely to be certified for hospice. At baseline, only one (7.1%) of the 14 patients who opted for LTMV used hospice, and another two patients used hospice at some point over follow-up, yielding a cumulative total of 21.4%. In contrast, among patients who did not choose LTMV, 43.9% were on hospice at baseline and 70.2% used hospice at some point over follow-up ($P < 0.01$). The two groups did not differ in the use of emergency department admissions, hospitalization, or skilled nursing home care.

Because the follow-up period was longer among LTMV patients, tests of differences in proportions between the two groups were adjusted for the length of follow-up using Mantel-Haenszel tests. Differences remained significant in logistic regression models with duration of follow-up included as a covariate.

Looking only at the subset of people with more than one assessment over follow-up (47 deaths, 13 LTMV) confirmed these differences. In addition to the greater use of nasal ventilation, counseling, and paid home care,

patients opting for LTMV in this comparison were significantly more likely to report the use of complementary/alternative therapies (61.5% vs. 27.7%, $P = 0.02$) and participation in clinical trials (53.8% vs. 23.4%, $P = 0.03$).

Medication Use in the Final Months of Life

Patients choosing LTMV began follow-up with significantly less use of palliative medications (Table 3). They were also less likely to use "other" medications, that is, medications not defined as mood, palliative, or ALS-specific therapies, which may be related to their younger age. Differences in medication use attenuated over follow-up.

Larger differences emerge when comparisons are restricted to the 60 patients (47 deaths, 13 LTMV) who were seen more than once over follow-up (Table 4). Patients who did not choose LTMV were more likely to stop medication use by their last study assessment before death. Significant time and group effects ($P < 0.01$) were confirmed in repeated-measures analysis of variance models.

Impact of Service Use and LTMV on Caregivers

In this cohort, caregivers of patients who ultimately opted for tracheostomy reported greater depressive symptoms and higher burden at baseline than caregivers of patients who declined tracheostomy; however, burden

Table 3
Medication Use at Baseline and Follow-Up:
Patients Dying Without LTMV and Patients
Choosing LTMV

Medication Class	Used at Baseline (%)		Used Any Time at Follow-Up (%)	
	Death	LTMV	Death	LTMV
ALS therapy	33.3	21.4	35.1	35.7
Palliative	57.9	14.3 ^a	66.7	57.1
Mood	36.8	35.7	54.4	42.9
Other	50.9	14.3 ^a	56.1	28.6

^a $P < 0.05$.

and depressive symptoms declined in both groups over time.¹⁴ Service use in the two groups was not related to caregiver burden or satisfaction. Similarly, we did not observe a clear pattern of change in caregiver burden or satisfaction after tracheostomy. Of the 14 patients who began LTMV, we were able to interview eight before and after patient tracheostomy. We found increases in burden in five, stability in two, and decline in one. Caregiver satisfaction was stable across the transition.

Discussion

In this sample of ALS patients with late-stage disease, use of health services and medications was more likely in patients who ultimately opted for tracheostomy and LTMV. In the months before death or tracheostomy, patients who chose tracheostomy were more likely to use nasal ventilation, paid home care, and personal or family counseling. The two groups of patients did not differ in legal preparation for the end of life or medical care episodes. Not surprisingly, none of the patients who opted for LTMV gave consent for autopsy, whereas 16.7% of patients who died gave such consent. Also consistent with these outcomes, patients who died were more likely to use hospice. It is important to note, however, that three of the 14 patients

who opted for LTMV were certified for hospice at some point over follow-up, suggesting that for some patients, the choice of LTMV is less clear and comes late. The majority, however, appears to have a clear preference for tracheostomy early on, as also reported in a separate cohort of recently diagnosed patients, who were mostly enrolled within one year of diagnosis and followed for a median of 12 months.⁸ This research shows that patients choosing LTMV use medical and supportive care services consistent with this preference.

Patients choosing LTMV also had different medication profiles. They were less likely to use palliative medications. Also, unlike patients who were approaching death, patients opting for LTMV were likely to continue taking medication over the course of follow-up. Almost all patients who died stopped taking medications by the time of their last study assessment.

Comparing our late-stage ALS cohort with the more recently diagnosed cohort described earlier shows how service use increases with progression of the disease. The early-stage cohort had a mean forced vital capacity of 78.5%. At baseline, very few patients in the early-stage cohort used nasal ventilation (3.2%) or had PEG placement (1.1%).⁸ In contrast, two-thirds of patients in the late-stage cohort used nasal ventilation and half had PEG placement at their initial assessment. Similar differences were obtained for paid home care, counseling, and use of alternative/complementary medicine. The two cohorts are separated by five years, and changing practice patterns may explain some of these differences.¹⁵ Still, comparisons suggest that in the first 12 months after diagnosis, patients are unlikely to use medical and supportive care services, but by 18–24 months, half to two-thirds make use of advanced medical technologies (e.g., nasal ventilation and PEG placement). Usage continues to increase

Table 4
Medication Use at Baseline and Last Assessment Before Event: Patients with Multiple Assessments ($n = 60$)

Medication Class	Death over Follow-up ($n = 47$)		LTMV ($n = 13$)	
	Baseline	Last assessment	Baseline	Last assessment
ALS therapy	34.0	4.3	23.1	23.1
Palliative	57.5	6.4	15.4	23.1
Mood	40.4	4.3	38.5	46.2
Other	51.0	4.3	15.4	15.4

All time and group effects were significant in repeated-measures analysis of variance.

in patients opting for LTMV but levels off and then declines in patients who decide against LTMV and approach death. For example, all but one patient who chose LTMV (92.9%) used nasal ventilation beforehand. The cumulative proportion among patients who did not choose LTMV was 62.5%.

Thus, the proactive orientation to health and desire to live despite severe disability reported for people choosing LTMV^{3,4} extends as well to more intensive use of medical and supportive care in the months before tracheostomy. Consistent with the strong preference to live, patients choosing LTMV also report fewer depressive symptoms in this period.^{3,6} Yet, the outcomes of LTMV are poor. A recent Swedish series found that only 5% of people with ALS who choose LTMV survived five years.¹⁶ In an Italian series of LTMV ALS patients, 9% died in hospital and median survival among the people discharged was 37 months.¹⁷ In our cohort, four (29%) patients on LTMV died over two years of follow-up, and half also reported a waning commitment to life after tracheostomy.³

Limitations of this research include its reliance on patients mainly from a single tertiary care center who were willing to participate in a demanding interview schedule. This may limit generalizability of results and suggests a need for replication with other samples. Still, we have shown in prior research that our study cohort is similar to patients seen in the tertiary ALS care setting.^{3,4} In addition, hospice eligibility in the United States requires patients not to pursue tracheostomy; the dynamics of service use in ALS might be different in health systems that do not require such a choice.

These results suggest that patients seeking tracheostomy have a different orientation to disease management than patients unwilling to consider tracheostomy. A challenging task for clinicians is to acknowledge the strong desire to live among this subset of patients while providing appropriate expectations for life after tracheostomy.

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