Advance Care Planning for Patients with Amyotrophic Lateral Sclerosis

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Abstract

Purpose—To determine whether an advance care planning (ACP) decision aid for could improve communication about end-of-life treatment wishes between patients with amyotrophic lateral sclerosis (ALS) and their clinicians.

Methods—Fourty-four patients with ALS (21, English-speaking, without dementia) engaged in ACP using an interactive computer-based decision aid. Before participants completed the intervention, and again 3 months after, their clinicians reviewed 3 clinical vignettes, and made treatment decisions (n=18) for patients. After patients indicated their agreement with the team's decisions, concordance was calculated.

Results—The mean concordance between patient wishes and the clinical team decisions was significantly higher post-intervention (post=91.9%, 95% CI=87.8, 96.1, vs. pre=52.4%, 95% CI=41.9, 62.9; p <.001). Clinical team members reported greater confidence that their decisions accurately represented each patient's wishes post-intervention (mean=6.5) compared to pre-intervention (mean=3.3, 1=*low*, 10=*high*, p<.001). Patients reported high satisfaction (mean=26.4, SD=3.2; 6=*low*, 30=*high*) and low decisional conflict (mean=28.8, SD=8.2; 20=*low*, 80=*high*) with decisions about end-of-life care, and high satisfaction with the decision aid (mean=52.7,

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SD=5.7, 20=*low*, 60=*high*,). Patient knowledge regarding ACP increased post-intervention (pre=47.8% correct responses vs. post=66.3%; p <.001) without adversely effecting patient anxiety or self-determination.

Conclusion—A computer-based ACP decision aid can significantly improve clinicians' understanding of ALS patients' wishes regarding end-of-life medical care.

Keywords

Advance Care Planning; Advance Directive; Living Will; Decision Aid; Surrogate Decision Making

Introduction

Amyotrophic lateral sclerosis (ALS) is a progressive, degenerative neurological disorder that inevitably results in functional decline and death, typically preceded by some form of surrogate decision-making at the end of life. Patients with ALS survive an average of 27–43 months after the onset of symptoms (1, 2), and though only a minority of these patients undergo tracheostomy and mechanical ventilation, such interventions commonly occur under emergency conditions in the absence of advance directives (3). As with other medical conditions, when patients with ALS fail to communicate their wishes for future medical treatment, the consequence is often sub-optimal medical decision-making during a crisis (4, 5), unwanted treatment and associated financial burden to patients, families, and society (6, 7), as well as moral distress for those who must make surrogate healthcare decisions (8–10).

Though patients most often rely on their families and physicians to speak on their behalf during a crisis, studies have shown that neither group accurately predict patients' wishes for life-sustaining treatments (11–15). Furthermore, fewer than 30% of American adults create advance directives to document their wishes (16–20); and even among patients with ALS fewer than 40% have recorded their attitudes and wishes regarding life-sustaining ventilation, resuscitation, artificial nutrition, and hydration (21, 22). In response to the need for better communication, the American Academy of Neurology and the European Federation of Neurological Societies have published guidelines emphasizing the importance of having planning discussions about end-of-life issues (23).

Improving communication between patients with ALS and their healthcare providers is an important way to help respect ALS patients' wishes. Individuals with ALS want to be involved in end-of-life decisions, particularly those involving tracheostomy and mechanical ventilation (24, 25). Importantly, ALS patients who receive such interventions often become unable to communicate effectively (26), and studies have shown that most would want these life-sustaining measures discontinued under certain conditions (27). Organizations such as the ALS Association (28) and the Muscular Dystrophy Association (29) have developed educational materials to facilitate decisions about living with ALS. And it has been proposed that an ALS-specific advance care planning (ACP) instrument could help facilitate discussion of ACP for these patients (30).

Despite these important steps, a central challenge to the goal of effective ACP is being able to ensure that people have ready access to high quality materials and/or facilitation. The intervention used in the present study (*Making Your Wishes Known: Planning Your Medical Future, or MYWK*) is an online decision aid created in large part to address this need. It provides a standardized, facilitated process that prepares patients for end-of-life decision-making, and generates an advance directive document that can then be shared and discussed with family members and healthcare providers.

The present study assesses the impact of MYWK on communication between patients with ALS and clinicians who treat them with regard to end-of-life treatment wishes. We hypothesized that the ALS team's understanding of patients' wishes would significantly improve after patients used this decision aid.

Materials and Methods

The broad study design involved asking the ALS team to consider several clinical vignettes, and then make hypothetical end-of-life decisions about them. This was done at baseline, and again after patients completed MYWK and shared with the team the advance directive document generated by this decision aid. Patients were then shown the ALS team's decisions, and asked to indicate whether they agreed or disagreed with each decision. A concordance score was then calculated as described below.

Patient Recruitment, Screening, and Eligibility

Following approval from the institutional Human Subjects Protection Office, patients were recruited from an interdisciplinary ALS clinic at Penn State Hershey Medical Center. Over an 18 month period, letters were sent to all patients scheduled for appointments, and follow-up phone calls were made to all patients who did not return an opt-out form. Interested patients 21 years or older then met with the study coordinator at their next scheduled visit and were asked to provide informed consent. Consenting individuals were screened to ensure they 1) were English speaking; 2) could read at 8th grade level (greater than 25 on the WRAT-3 (31)); 3) did not have evidence of dementia (greater than 23 on Folstein Mini-Mental Status Examination (32)); 4) had at least one previous visit to the ALS Clinic; and (as determined by the patient's ALS physician) 5) could communicate effectively either verbally or through the use of an alternative/augmentative communication device; 6) had sufficient physical stamina to tolerate the study protocol; and 7) had an estimated life expectancy at enrollment of at least 3 months. Patients were sequentially recruited until the target number of 50 enrollees was reached. Patients who were ineligible for the study but still wished to use the decision-aid were permitted to do so, but no data were collected. Eligible patients who declined participation (n=33) were asked to complete a brief non-participant questionnaire.

ALS Health Care Team Recruitment, Screening and Eligibility

The ALS healthcare team was recruited from the interdisciplinary ALS clinic at Penn State Hershey Medical Center. This 15 member team consisted of physicians, nurses, physical and occupational therapists, as well as a speech therapist, dietician, mental health counselor,

social worker, and pastoral counselor. All ALS team members provided informed consent for their participation in this study, and no member of the ALS team declined to participate.

Decision Aid Intervention

Making Your Wishes Known (MYWK) was developed to educate individuals about advance care planning, help them clarify and articulate their values, and then (using an algorithm based on multi-attribute utility theory) (33, 34) translate their wishes and goals into an actionable medical plan (35, 36). MYWK is intended to facilitate, not replace, discussions between patients and healthcare providers, and previous studies with non-ALS patients demonstrated that people who used the program reported high levels of satisfaction, demonstrated improved knowledge about ACP, rated the advance directive generated by MWYK as an accurate and reliable representation of their wishes, and experienced no adverse effect on either hope or anxiety (37–39). A prior pilot study of 17 patients with ALS yielded similar results (40).

MYWK uses an interactive, multi-media format to: 1) provide tailored education about common medical conditions that can result in decisional incapacity, as well as medical treatments often introduced in life-or-death situations; 2) guide individuals to choose a spokesperson, prioritize values and goals, and match treatment options with priorities; and 3) translate individual values and preferences into a detailed advance directive document. Participants used a laptop computer to access MYWK. Individuals with insufficient upper extremity strength and/or coordination communicated their responses to the research assistant (or loved one, if such a person had accompanied them), who operated the computer mouse for them. Upon completing the program, participants received a printed copy of their personalized advance directive.

Patient Measures

Baseline measures included: 1) *Demographics and Health Assessment* (22 items); 2) *Self-Determination* (8 items, adapted from Pellino (41), assessing individuals' sense of control over future treatments); 3) *Advance Care Planning Knowledge* (27 items, previously described (42), assessing knowledge of the advance care planning process and the clinical decisions patients often face at the end of life); 4) *Anxiety* (State Trait Anxiety Inventory (43)); 5) *Functional Status* (ALS Functional Rating Scale-Revised ALSFRS-R (44)); and 6) *Quality of Life* (ALS-Specific QoL Scale (45)).

Post-intervention patient measures included: 1) *Self-Determination* and *ACP Knowledge* (same as pre-intervention); 2) *Satisfaction with Advance Care Planning* (15 items, assessing comprehensibility, length, pace, amount of information, balance, and overall satisfaction); 3) *Decisional Conflict* (6 items, assessing discomfort/uncertainty about decisions (46)); 4) *Satisfaction with Decision* (6 items (47)); and 5) *Time and Effort* (3 items).

ALS Team Measures

Every 3 months ALS patients undergo a half-day clinical evaluation, after which the ALS team convenes an interdisciplinary meeting. At these meetings ALS team members were asked to discuss three clinical vignettes describing events that would prompt the need for

surrogate decision making. For each patient, the ALS team discussed the three scenarios until consensus was reached about what hypothetical medical decisions would be appropriate for that patient (see Figure 1). This was done twice: prior to the patient having used MYWK, and at the patient's next clinic visit after they had completed MYWK and a copy of their advance directive had been shared with the ALS team. Following each discussion, the ALS team rated their confidence that their decisions accurately reflected the patient's wishes (10-point Likert-style scale, where 1=*low*, and 10=*high* confidence).

Patient-ALS Team Concordance Calculation

In the weeks following their post-intervention ALS clinic appointment, patient participants were sent the 3 clinical vignettes that had been shared with the ALS team. The patient and physician versions were worded slightly differently. For example, patients were asked "… w*hich of the following would you want and not want? Mechanical ventilation if your breathing worsens (Yes or No)*, while physicians were asked *"Based on your knowledge of this patient, would you initiate mechanical ventilation?"* In subsequent phone interviews, the ALS team's post-intervention decisions were shared with the patients, who were then asked whether they agreed with each decision. Each question-item counted for one point – however, to avoid falsely inflating the concordance score, decisions about *who should serve as the patient's surrogate decision-maker* were counted just once (rather than once for each vignette). A concordance score was calculated (using patients' responses during these telephone interviews) by summing the number of treatment decisions for which patient and ALS team responses were identical, dividing by the total number of treatment decisions, and multiplying by 100 (score range = 0–100%).. The rationale for asking patients whether they agreed or disagreed with the ALS team's responses (rather than asking them to respond to the vignettes by themselves, i.e., *without* knowing the clinical team's opinion) was that this was felt to more closely resemble typical patient involvement in clinical decision making.

Statistical analysis

All analyses were performed using SAS version 9.4 (SAS Institute, Cary, NC), and statistical significance was set at $p < 0.05$. All variables were summarized with frequencies and percentages or means, medians, standard deviations, and quartiles prior to comparisons being made. The distribution of continuous variables was assessed using histograms and box plots. A comparison of pre-intervention to post-intervention mean knowledge was made using a paired t-test. Mean concordance percentage for total and for individual vignettes as well as for procedures, average confidence overall and for the individual vignettes, self-determination, and anxiety were compared using a Wilcoxon signed-rank test due to their distributions being skewed or ordinal in nature.

Results

Completion Rates and Time Spent

Of the 130 eligible patients followed by the ALS team, 17 returned "opt-out" cards, and 33 declined in person. The remaining individuals were approached sequentially until we met our enrollment goal of 50 participants. For these, *demographics* were calculated for 44 participants (because 6 patients were too ill to attend the research study visit), *team*

confidence was calculated for 43 participants (because 1 patient completed the intervention but never returned for a subsequent ALS clinic appointment), and *concordance* was calculated for 42 participants (because 1 patient could not be reached after their subsequent clinic visit).

Patient Demographics and Experience with Advance Care Planning

Participants' mean age was 57.7 years (range=34–78) and the majority were white, married, and had at least a high school education, and moderate physical impairment (See Table 1 for *Demographics*). The only difference identified among patients who opted out or declined participation (n=50) was that non-participants were more likely to be female (56%) compared to study participants (34%).

Approximately half of participants reported having read or heard "a lot" (25%) or "a fair amount" (27%) about advance care planning or living wills, while 16% reported having heard almost nothing about them. Half of participants had prepared some form of advance directive prior to enrolling in the study, and half had also formally assigned someone as their spokesperson. All but four participants reported owning a computer (average weekly use=20 hours, range 0–56), and the majority of participants (91%) reported feeling comfortable using a computer.

Participants spent an average of 16 minutes to complete pre-intervention questionnaires, 77 minutes to complete the computer intervention, and 21 minutes for post-intervention questionnaires, with none reporting that completion of the study activities was burdensome.

ALS Team Decisions

Pre-intervention ALS team decisions were recorded for all 50 patient-participants, and post-intervention decisions were recorded for 43 (6 were too ill to attend the research study visit, and 1 completed the intervention but never returned for a subsequent ALS clinic appointment).

Decision Concordance and Confidence

Pre-intervention, patients agreed with 52.4% of the ALS team's decisions for the clinical vignettes (18 total decisions). Post-intervention, concordance increased to 91.9% (p<.001) (Table 2). Additionally, the ALS team's confidence that their decisions accurately represented patients' wishes was significantly higher post-intervention, after they had reviewed the patients' MYWK-generated advance directive (pre-intervention mean confidence=3.3 vs. post-intervention=6.5, where 1=*low* and 7=*high confidence*, see Table 2).

Satisfaction with Advance Care Planning

Patients reported high overall satisfaction with the ACP decision aid (mean=9.1 ± 0.9, 1=*low* and 10=*high*), as well as for 12 specific tasks (mean=52.7 ± 5.7, *12=low* and *60=high*), such as providing information about medical conditions and treatments, clarifying values, choosing a spokesperson, and preparing to discuss one's wishes with others. Patients also reported that the decision aid-generated advance directive was highly accurate in reflecting their wishes (mean=9.4 ± 0.8, 1=*low* and 10=*high*).

Impact on Advance Care Planning Knowledge

Participants' *ACP knowledge* increased significantly (p<0.001) from a mean of 47.8% (CI=42.8, 52.9) correct answers at baseline to 66.3% (CI=62.8, 69.7) post-intervention.

Impact on Self-Determination, Anxiety, and Decisional Conflict

Participants had a strong sense of *self-determination* at baseline (mean=35.0, CI= 33.8, 36.2; 8=*low*, 40=*high*), which increased significantly (p=0.034), albeit modestly, following the intervention (mean=36.1, CI=34.9, 37.3). Participants' *anxiety* was low at baseline (mean=30.5, CI=27.8, 33.1;, 20=*low*, 80=*high*), and decreased slightly post-intervention, though not significantly (mean=28.7, CI=26.0, 31.4; p=0.075). Participants reported low levels of decisional conflict for the treatment decision made for the clinical vignettes (mean 28.8 ± 8.2, 16=*low*, 80=*high*).

Discussion

This study found that clinicians' decisions about future end-of-life care were better aligned with patients' actual wishes after patients completed an online decision aid and shared the advance directive document that it generated. After the intervention, the medical team also reported significantly greater confidence in the accuracy of their treatment decisions.

These findings are noteworthy given the inevitable clinical course faced by patients with ALS, and the observation that physicians are not accurate at predicting patients' preferences for end-of-life care (48). Hence, a tool that assists healthcare providers to better understand patients' wishes can help them both engage patients in ACP discussions, and make treatment decisions that are consistent with patients' wishes.

We do not have detailed information about what happened in the time interval between the intervention and the final discussion, so it is difficult to know with certainty what accounts for this large increase in patient-ALS team agreement. Some of the improvement in concordance may have resulted from direct discussions that improved communication between the patient and the clinical team, which itself is a central goal of MYWK. However, it does not appear that the increased concordance was simply a result of improvement over time (e.g. the clinical team becoming more sensitized and skilled as the study progressed, and hence interacting differently with patients recruited later in the study). Specifically, we found that the 52% concordance at baseline was no different at the end of the 18-month study, nor were there significant differences between early versus end-of-study post-intervention concordance scores.

Before deciding how best to respect a patient's wishes, clinicians must first determine which treatment options are medically appropriate. The treatment options in this study were limited to interventions that typically would qualify as "medically indicated." Accordingly our findings may not be generalizable to situations in which patients desire interventions that are medically inappropriate. To the extent that an ACP decision aid can help patients better appreciate which interventions are and are not medically warranted, this too is a benefit.

Consistent with previous studies involving other patient populations, we found that after using the decision aid, patients' knowledge of ACP increased, and patients expressed high levels of satisfaction with MYWK (37, 40, 42, 49, 50). So too, there were no adverse effects on anxiety, which should help allay concerns some clinicians have about discussing end-of-life issues with patients.

Taken together, these findings suggest that MYWK can be a valuable tool for busy clinicians whose multiple competing obligations make it is increasingly challenging to find sufficient time to educate patients about end-of-life decisions, help them clarify values, and explain the clinical implications of ACP decisions. Preparing patients to have more informed conversations with members of their healthcare team not only can help patients deal with this sensitive topic, but can help clinicians adhere to professional guidelines about ACP.

Limitations

Like all studies, this one has certain limitations. First, the study design did not include a control group, so it is difficult to know with certainty that the improvement in concordance is not attributed to factors other than the decision aid. Second, the main outcome was concordance with decisions made in response to clinical vignettes, not actual decisions. What people decide in real life may differ from decisions involving hypothetical vignettes. Third, the study was conducted at a single institution with low ethnic diversity, and this could affect generalization to other populations. However, the age, male predominance, and ALS-Specific QOL scores of the subjects reflect those in other, larger studies of ALS patients (45). Fourth, the concordance was based on patients' post-intervention views (rather than comparing patients' 'pre-intervention agreement with the ALS team's pre-intervention decisions' to their 'post-intervention agreement with the ALS team's post-intervention decisions'). The rationale for this approach was that introducing the vignettes prior to the intervention risked introducing bias insofar as the vignettes themselves could constitute an ACP intervention. Additionally, prior research (51) has shown stability in patients' overall preferences, suggesting that patients' wishes would not change dramatically over the 3 months of their involvement in the study. Finally, asking patients to agree or disagree with the clinical team's treatment decisions may have introduced social desirability bias. That said, the clinical reality is that patients make decisions in response to the options and recommendations given to them by clinicians. Additionally, the vignettes and accompanying questions actually sent to patients were framed for patients (*Which of the following medical treatments would you want in the circumstances described?*), not physicians; and the questions themselves were unanswered (i.e., the ALS team's responses were not recorded on the materials sent to patients).

Conclusions

The present study suggests that an online decision aid is an effective approach for helping patients with ALS engage in advance care planning, and for helping ALS team members have a better understanding of patients' wishes regarding end-of-life care. Tools that can help patients and clinicians agree on treatment decisions that are consistent with patients'

values and goals are particularly valuable for patients with progressive, degenerative diseases such as ALS.

Acknowledgments

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ALS Team version

For the following scenarios, please accept as given the prognoses provided. Though there will always be clinical uncertainty and reasons to question the accuracy of predicted outcomes, medical decisions still must be made based the evidence that is available.

Patient: _____

This patient has reached a stage in the progression of ALS where she is wheelchair-bound and has only minimal movement of her arms and hands. Her breathing capacity is only about 50% of normal, but she is not short of breath at rest or with performing routine activities. She has had <u>no</u> decline in her mental abilities. A few days after a family gathering, she began with a fever that evolved into a rapidly progressive pneumonia. Along with this, she developed significant mental status changes. She is confused and cannot carry on a meaningful conversation or express her views in a coherent manner. She has been brought to the local emergency room, and it is determined that she will need to be put on a mechanical ventilator if she is to survive. With aggressive medical care, it is highly likely that her pneumonia can be successfully treated and that her thinking will clear, but her strength will continue to worsen as a result of her ALS, and will likely do so quite quickly because of the period of forced bedrest required. There is a high chance that she will never be able to breathe on her own again, and thus would require mechanical ventilation for the rest of her life.

Based on your knowledge of this patient:

1. Would you initiate mechanical ventilation for this patient?   Yes / No

2. Would you initiate intravenous antibiotics for this patient?   Yes / No

3. Would you perform CPR if the patient's heart stopped?   Yes / No

4. Would you initiate renal dialysis if the patient's kidneys failed?   Yes / No

5. Would you order a feeding tube if the patient could not take food by mouth?   Yes / No

6. Who is the appropriate person to speak for this patient? _____

7. Based on your knowledge of this patient, how confident are you that you were able to appropriately translate the patient's goals and values into clinical decisions that accurately reflect his or her wishes? (circle a number)

1	2	3	4	5	6	7	8	9	10
Not at all Confident									Extremely Confident

Figure 1.

Table 1

Participant Demographic Characteristics (n=44)

Characteristic	Percent (unless otherwise noted)
Age in years, mean (range)	57.7 ± 11.3 (34–78)
Gender	
Female	34.1
Male	65.9
Race	
White	100.0
Duration of disease (months)	
Mean (SD)	28.9 ± 25.2
ALSFRS-R score [*]	
Mean (SD)	36.3 ± 7.9
ALSSQOL-R score [**]	
Mean, (SD)	7.5 ± 1.0
Highest education	
Some high school	6.8
High school graduate or GED	25.0
Some college or technical school	25.0
College graduate	25.0
Graduate or professional school	18.2
Religion	
Catholic	18.2
Protestant	59.1
Other	11.4
No formal religion	11.4
Marital status	
Never married	6.8
Married or Domestic partnership	81.8
Divorced or Legally separated	2.3
Widowed	9.1
Employment status	
Retired	50.0
Employed full-time	27.3
Not currently employed	13.6
Disabled	9.1

[*] ALS Functional Rating scale ranges from 0–48, with higher scores indicating greater function.

[**] ALS-Specific Quality of Life scale ranges from 1–10, with higher scores indicating better quality of life (n=39).

Table 2

Patient-Provider Decision Concordance and Provider Decisional Confidence

Concordance (%)[*]	N	Baseline Mean (95% CI)	Post-Intervention Mean (95% CI)	P-value
Total concordance	42	52.4 (41.9, 62.9)	91.9 (87.8, 96.1)	**<0.001**
Vignette #1	42	51.6 (40.8, 62.4)	96.4 (94.0, 98.9)	**<0.001**
Vignette #2	42	55.2 (43.6, 66.7)	94.8 (91.3, 98.4)	**<0.001**
Vignette #3	42	50.4 (38.1, 62.7)	89.3 (83.0, 95.6)	**<0.001**
Mechanical ventilation	42	54.8 (43.1, 66.5)	81.4 (74.4, 88.5)	**<0.001**
Dialysis	42	48.4 (34.6, 62.2)	79.4 (69.1, 89.6)	**<0.001**
Feeding Tube	42	53.2 (40.6, 65.8)	70.6 (60.3, 81.0)	**<0.001**
CPR	42	47.6 (33.1, 62.2)	88.9 (80.7, 97.1)	**<0.001**
Antibiotics	42	57.9 (44.4, 71.5)	71.4 (60.1, 82.8)	0.073
Confidence (1=*low*, 10=*high*) [*]				
Mean confidence	43	3.3 (2.5, 4.1)	6.5 (5.7, 7.2)	**<0.001**
Vignette #1 confidence	43	3.4 (2.6, 4.3)	6.6 (5.8, 7.4)	**<0.001**
Vignette #2 confidence	43	3.4 (2.5, 4.8)	6.5 (5.7, 7.3)	**<0.001**
Vignette #3 confidence	43	3.1 (2.3, 3.8)	6.3 (5.4, 7.1)	**<0.001**

[*] Wilcoxon Signed Rank test

Because 1 participant could not be reached after the ALS team made decisions, a discrepancy exists between the numbers used to calculate *Concordance* (n=42) vs. *Confidence* (n=43) scores.