Top Lang Disorders
Vol. 32, No. 2, pp. 168–185

Methods of Communication at End of Life for the Person With Amyotrophic Lateral Sclerosis

Alisa Brownlee and Lisa M. Bruening

Amyotrophic lateral sclerosis (ALS) is a neurodegenerative disease that results in loss of most motor functions by the time of death. Most persons with ALS experience a dysarthria that eventually renders oral/vocal communication unintelligible. This article reviews the communication needs of persons with ALS and the range of communication strategies used, including most forms of augmentative and alternative communication (AAC). Survey data are presented concerning perceived presence and severity of communication deficits, common communication topics, and communication strategies used with different communication partners and at 3 time intervals (2–6 months, 4 weeks, and 1–2 days) before death. Survey data were collected from 625 family members/caregivers in 8 states in the United States, reporting on their experiences with persons with ALS who were deceased at the time of the survey. The analysis focused on patterns of communication used in the last 6 months as end-of-life approaches. Most common communication topics were physical needs, caregiving issues, and family issues. Least common topics were spiritual and death and dying issues. Communication strategies did change as end of life approached, with a decrease in all modes of communication including natural speech, writing, gestures, and electronic AAC. Unaided and low-tech strategies did not increase during the same time interval. Health care providers must be knowledgeable about the communication options available and factors influencing communication choices. Speech–language pathologists play an important role in monitoring speech changes, providing assistance in making choices about communication options, and educating clients, health care providers, and family members. **Key words:** *AAC*, *ALS*, *communication*, *end of life*, *Lou Gehrig's disease*, *persons with ALS*

Author Affiliations: *Assistive Technology Services, ALS (Lou Gehrig's Disease) Association National Office and Greater Philadelphia Chapter (Ms Brownlee), Hatfield, Pennsylvania; and ALS Association, Northern Ohio Chapter, Cleveland and University of Akron, Ohio (Ms Bruening).*

The authors have disclosed that they have no significant relationships with, or financial interest in, any commercial companies pertaining to this article.

Supplemental digital content is available for this article. Direct URL citations appear in the printed text and are provide in the HTML and PDF versions of this article on the journal.s Web site (www.topicsinlanguagedisorders.com).

Corresponding Author: *Alisa Brownlee, BS, BA, ATP, Assistive Technology Services, ALS (Lou Gehrig's Disease) Association National Office, 34 Market St Hatfield, PA 19440 (abrownlee@alsa-national.org).*

DOI: 10.1097/TLD.0b013e31825616ef

TERMINAL ILLNESS forces people to face end-of-life issues. The ability to communicate while approaching the end of life is critical to being able to participate in important medical or financial decisions, maintain interactions with family and friends, and sustain a level of independence. Not surprisingly, there is a rapidly growing body of literature discussing aspects of end-of-life communication content and adequacy (e.g., Gauthier, 2008; Levin, Moreno, Sylvester, & Kissane, 2010; Prince-Paul, 2008).

Many diseases that limit life expectancy are not expected to alter one's ability to communicate through speech. People with a diagnosis of amyotrophic lateral sclerosis (ALS), also known as Lou Gehrig's disease, face a different reality. It has been reported that between 75% and 95% of those with a diagnosis of

ALS will lose the ability to use natural speech before death (Beukelman, Fager, & Nordness, 2011; Saunders, Walsh, & Smith, 1981). Although the course of ALS and the symptoms vary considerably, a speech-related problem may be the reason people with ALS first seek medical help. By the time a formal diagnosis is received, the person may be experiencing significant difficulties communicating.

As a group, people with ALS demonstrate a wide range of needs and preferences for communication strategies, including some of the more technologically sophisticated speech-generating devices (SGDs). These needs and preferences may change as individuals approach the end of life. This article provides an overview of the changes in communication experienced by persons with ALS and factors influencing their use of augmentative and alternative communication (AAC) strategies. Results of a study exploring caregiver reports of persons with ALS communication topics and use of communicative strategies with different communication partners and across the last 6 months of life are presented. Implications for speech–language pathologists (SLPs) and other health care professionals are discussed.

AMYOTROPHIC LATERAL SCLEROSIS

ALS is a progressive, degenerative disorder of the voluntary motor system affecting both the corticospinal and corticobulbar tracts of the central nervous system (upper motor neurons) and the motoric cranial and spinal nerves of the peripheral nervous system (lower motor neurons). The autonomic motor system is not affected, and eye movement is usually spared. ALS can be diagnosed in adults of any age, although age of onset is typically between 40 and 60 years (National Institute of Neurological Disorders and Stroke, 2010). The cause is not known, although a familial variant has been identified in 5%–10% of all ALS cases (ALS Association, n.d.).

Typically, ALS begins with signs of weakness in some part of the body. The initial symptoms are used to classify patients as having either spinal ALS or bulbar ALS. The spinal form is more common than the bulbar, with estimates indicating 75% initially showing spinal manifestations and 25% demonstrating the bulbar form (National Institute of Neurological Disorders and Stroke, 2010). In the spinal form, spinal nerves are affected first, with the legs, arms, or hands becoming weak, and associated difficulties in activities such as walking, lifting, or writing. In the bulbar form, the cranial nerves are affected first, with disorders of speech and swallowing noted early. Dysphonia may be the first symptom experienced, with the person being evaluated by an otolaryngologist prior to referral to a neurologist (Hillel et al., 1999). The person with ALS may compensate for early signs of weakness, so considerable impairment may be present before the diagnosis of ALS is confirmed.

As the disease progresses, weakness continues and advances to affect additional body parts. Eventually, the motor neurons lose function and die. Signs indicating loss of the lower motor neuron include muscle fasciculation and cramping before the muscles atrophy. Upper motor neuron damage is indicated by spasticity and hyperreflexia. A mix of symptoms is common, resulting in a mixed spastic-flaccid dysarthria. If the initial symptoms are respiratory failure, the person typically requires artificial ventilation early in the course of the disease or has a limited survival time.

Muscle fatigue is common in ALS; even sitting up or wearing a heavy coat can cause fatigue in the postural muscles. Although strengthening exercises may exacerbate the condition, a lack of movement to avoid fatigue is not recommended. It is important that the person maintain a range of movement as long as possible, through passive movement if necessary. If movement is not maintained, contractures can develop rapidly.

The progression of the disease is variable, but death usually occurs within 2–5 years of diagnosis (ALS Association, n.d.). Life expectancy for those with bulbar onset is considerably less than for those with spinal onset. A number of factors, including adequate nutrition and use of invasive and noninvasive

positive pressure ventilation, influence life expectancy.

It is difficult to determine what constitutes the end-of-life time interval for persons with ALS. Despite differing initial symptoms and early disease trajectories, persons with ALS and their families are usually informed upon initial diagnosis that ALS is fatal. Thus, most may be experiencing psychological transitions associated with this knowledge. Quality of life is of primary concern during these years of living with a diagnosis of ALS. For many, quality of life is linked to communicative effectiveness, and AAC strategies and devices offer potential communicative support.

AAC SUPPORT FOR ALS

For persons with ALS who experience speech impairment or loss, there are a number of AAC approaches available, often described in three broad categories. Unaided AAC can include gestures, signs, and/or facial expressions. Aided low-technology (low-tech) systems include letter-or picture boards as well as devices with a limited number of voice output messages (usually digital) that can be accessed by selecting a picture or word. High-technology (high-tech) options encompass all of the SGDs, which are computer-based systems that allow individuals to communicate and to control their environments through multiple access methods despite profound motor impairments. In recent years, the greatest advances in the high-tech arena have been made in reliable eye-tracking control of computer-based systems for communication and other technology applications, such as for environmental control, e-mail, and so forth (Ball et al., 2010).

Ideally, use of one or more of the aforementioned approaches should provide any person with ALS with a means of communicating even if the communication has limits. Efforts have been made to link stages in speech deterioration (Yorkston, Miller, & Strand, 1995) or clinical subgroups (Mathy, Yorkston, & Gutmann, 2000) with specific AAC interventions. Changes in physical status and personal needs and preferences of persons with ALS and their significant others can influence communicative choices. The best communicative option can be determined through a clinical process that considers all relevant factors in a timely fashion. On the basis of the combined professional experience of the authors of this article working with hundreds of person with ALS, however, one thing is clear. It can be difficult for many patients with ALS and their families to consider the need for AAC when the client is still using natural speech. Successful communication, particularly acceptance and use of AAC, is influenced by many variables. One of the most important is early referral to an SLP when any change in speech- or swallowing-related functions is noted. The SLP evaluates current communicative and swallowing status, identifies useful communication strategies that can be taught and used before AAC technology is needed, and monitors the need for timely and ongoing AAC evaluation.

Unfortunately, clinical experience and patient/family report suggest that many persons with ALS do not receive speech- and communication-related services early in the course of the disease due to a variety of reasons; some are referred (and assessed and treated) primarily for swallowing difficulties; others remain in denial or have difficulty accepting the diagnosis. Often, communication is not evaluated until speech intelligibility is markedly impaired. Obviously, clinical practice guidelines recommend early assessment and planning to accommodate reductions in function before they occur.

The timing of referral to an SLP often dictates the nature of AAC assessment and intervention activities. Referral timing is influenced in part by the actual motor speech decline and by accompanying motor impairments in other parts of the body, particularly initial symptoms and course of the disease for bulbar versus spinal onset. People who first demonstrate bulbar symptoms are often referred to SLPs relatively early in the course of the disease. At this point, some may already have adapted to their dysarthria by writing their messages and using gestures to

sustain communicative effectiveness. Although some retain their ability to write throughout the course of their disease, the ability to write probably will be affected as the disease advances to the spinal nerves. The sequence of AAC use in bulbar ALS may go from use of simple strategies (e.g., writing) to increasingly sophisticated technology. Conversely, those with a diagnosis of spinal ALS may have difficulty writing before experiencing significant difficulty speaking. Again, speaking may be possible throughout disease, but the majority will experience a gradual loss of the ability to speak. By the time some persons with spinal-onset ALS are seen by an SLP, they may require a fairly sophisticated AAC method, such as an SGD, because of device access issues.

If AAC assessment is postponed until intelligibility is impaired, acceptance and successful use of AAC can be adversely affected (Beukelman et al., 2011). Thus, other predictors of future speech impairment are needed. Ball, Willis, Beukelman, and Pattee (2001) created a protocol to identify early signs of bulbar involvement, given its importance in diagnosis and tracking of the disease. On the basis of a series of clinic visits during which speech samples were videotaped, they found that voice quality (laryngeal control), speaking rate, and impaired communicative effectiveness were the most important predictors of bulbar speech impairment in early stages. The importance of speaking rate as a predictor of speech decline was confirmed in another study (Ball, Beukelman, & Pattee, 2002). In addition, Ball, Beukelman, Ullman, Maassen, and Pattee (2005) determined that speaking rate could be determined reliably over the telephone. Ball, Beukelman, and Bardach (2007) used this research to develop guidelines for referral for AAC evaluation; the primary criterion being speaking rate dropping to or below 125 words per minute on the Sentence Subtest of the Speech Intelligibility Test (Beukelman, Yorkston, Hakel, & Dorsey, 2007). Clearly, SLPs need to be involved in monitoring patients, particularly those with bulbar onset, early in the disease progression.

On the basis of a review of medical records of 24 patients with ALS, Zeitlin, Abrams, and Shah (1995) suggested that successful AAC use was best predicted by high levels of patient motivation and by having specific communication goals. However, the AAC options available more than 15 years ago were quite limited compared with today's alternatives. Other factors associated with lower motivation for AAC include cost and fears of more rapid loss of speech if a device is introduced (Ball et al., 2007; Mathy et al., 2000). *Self-image*, defined as perception of self as a speaking individual and desire to be seen as such, has been identified as a factor limiting AAC acceptance and use (Brownlee & Palovcak, 2007; Mathy et al., 2000; Shadden, Hagstrom, & Koski, 2009).

Brownlee and Palovcak (2007) noted that any communication intervention must consider whether the person with ALS has recognition/awareness of declines in speech intelligibility. Sometimes people with dysarthria secondary to ALS do not realize that their speech is becoming difficult to understand. Family, friends, and even health care providers may not wish to indicate that they are having trouble understanding because they do not want to distress the person with ALS or call attention to another ALS-related problem. In some instances, failure to recognize reduced intelligibility can lead to communication partner frustration. Thus, it may be necessary to make the person with ALS aware of the increasing listener burden caused by worsening dysarthria. Without awareness, there is less motivation to explore other communicative strategies. Even with awareness, some persons with ALS persist in wishing to try speech first despite access to an AAC system (Murphy, 2004).

Communication partner acceptance plays a major role in AAC use as well. Richter, Ball, Beukelman, Lasker, and Ullman (2003) reported that high-tech SGDs were preferred to hard-to-understand natural speech and to low-tech augmented communication. Listeners were individuals with ALS, strangers, and caregivers; speech was videotaped narratives.

Fried-Oken et al. (2006) linked positive caregiver attitudes about AAC use with a sense of greater reward as a caregiver, more perceived closeness to patient, and less difficulty (less sense of burden) in the actual provision of care. Whether or not this is actually satisfactory to all communication partners is unique to each family unit.

An important factor in AAC use may be the differing communication needs or activities along the path toward adjustment to the diagnosis (Fried-Oken, Fox, Rau, Tullman, & Lou, 2003). Doyle and Phillips (2001) compared potential stages in adjustment with stages in AAC use, illustrating their premises with case studies. Although their work is more than a decade old, some of the core issues remain current. As with other terminal illnesses, the individual, family, and friends may struggle early on with understanding and accepting the reality of ALS, particularly the knowledge that the disease is progressive and there is no known cure. At this stage, many persons with ALS retain some intelligible speech. Their communication needs and partners may be diverse. Some may be actively seeking information; others may be experiencing degrees of denial. Not surprisingly, during this phase, most rely on unaided communication or low-tech AAC approaches (Doyle & Phillips, 2001).

As the disease progresses, some persons with ALS may wish to engage in end-of-life planning. The focus may be on making certain that legal and/or financial affairs are in order, that arrangements for future family needs are made, or that appropriate people are contacted. Communication needs can become more complex at this point, requiring more sophisticated language messages consistent with the high-tech systems described by Doyle and Phillips (2001) for middle-stage ALS.

In the final stage, closer to death, there may be a primary focus on saying goodbye and on meeting basic physical needs, often with a return to unaided or low-tech communication strategies, as described by Doyle and Phillips (2001). More recent research about changes in communication needs and strategies during the ALS disease progression suggests that newer technologies may make it possible for people with ALS to communicate effectively further into the disease (Ball et al., 2010; Beukelman et al., 2011), but additional research is needed.

Another factor with potential to affect overall communicative effectiveness and use of AAC is cognitive functioning. In the past, patients with ALS were considered ideal candidates for AAC because cognition was thought to remain unaffected. In fact, the absence of cognitive impairment has been a diagnostic criterion (Zago, Poletti, Morelli, Doretti, & Silani, 2011), and this fact reassured patients and families as they faced motor decline (Elman & Grossman, 2007). Over the past decade, however, evidence has accumulated that some patients with ALS have measurable cognitive and behavioral deficits that can have practical consequences (Elman, McCluskey, & Grossman, 2008; Zago et al., 2011). Recent studies suggest that anywhere between 15% and 41% of patients with ALS (or more) may develop deficits consistent with frontotemporal dementia (Miller et al., 2009; Zago et al., 2011). As many as half of those with ALS may have mild-to-moderate cognitive or behavioral abnormalities (ALS Association, 2005).

The impact of cognitive deficits on communication and AAC use is not yet well understood (Beukelman et al., 2011), although it is possible that limited communication in the late stages of ALS can be partly a function of cognitive deficits. Elman and Grossman (2007) suggest the most common executive function deficit is verbal fluency, which could affect communication. Executive function skills are involved when persons with ALS are asked to make decisions (e.g., whether to acquire an AAC device or which device to select) or to learn and use new communication strategies and/or complex devices. It is critical, however, to distinguish between cognitive impairment as measured on standardized tests and functional cognitive limitations in daily living. Because persons with

ALS typically experience a steady disintegration in their ability to speak, swallow, move, and perform activities of daily living, in addition to coping with a terminal illness, it is easy to overlook the presence of some common signs of mild cognitive or behavioral dysfunction, such as poor insight, lack of empathy, deficits in planning, agitation, or euphoria. SLPs should monitor clients with ALS for signs of cognitive decline. It is also important to recognize that some cognitive changes may result from nocturnal hypoxia (Thesen et al., 2012), which can signal impending respiratory failure and death. If not identified and managed, reversible cognitive change secondary to hypercapnia (excessive carbon dioxide in the blood) can lead to permanent cognitive deficits (Ogawa, Tanaka, & Hirata, 2009). Speech–language pathologists can play a role in monitoring respiratory functions and alerting the medical team to such problems as they arise.

Clearly, providing communication strategies and support for persons with ALS can be challenging for all parties. In addition to the physiological and cognitive factors already noted, it is important to recognize that person with ALS and his or her families are dealing simultaneously with current and impending loss.

PSYCHOLOGICAL IMPACT OF LOSS OF COMMUNICATION

ALS is often described as a disease of losses, most obviously loss of independence. Some are losses of function—of the ability to walk, eat, and breathe. These losses can occur on a daily basis and demand constant change on the part of the patient and the caregiver. A person with a diagnosis of ALS is also going through catastrophic psychological losses—of dreams, of the expected future, of life itself. These can lead to deep grieving (Luterman, 2001). Dealing with multiple losses can interfere with making decisions or accepting changes.

Among the many losses of ALS is the loss of communication. Under any circumstances, loss or impairment of communication is deeply personal. An acquired communication disorder can devastate the family unit as family roles change and the emotional balance of the family unit is upset (Hinckley, 2008). When communication loss occurs as death approaches, a critical tool for relationships and for conveying needs and preferences is disrupted. The artificiality of electronic AAC devices may be seen as a barrier to needed social closeness (Murphy, 2004). Emphasizing this point, some family members interviewed by Murphy indicated that the person with ALS did not need to "talk" to be understood.

Communication is associated with personhood and identity (Shadden et al., 2009). The loss or anticipated loss of speech and communication can be like the loss of humanity, and having to face this loss when confronted with a terminal illness is even more challenging. How each individual with ALS reacts to the loss of communication will be unique and may change over time. However, those reactions may influence the person's decisions about AAC specifically.

Because ALS is progressive, people with ALS may need assistance to look into the future and consider what equipment they may need in a few weeks or months because of another lost function. This is difficult enough when, for example, the person is still walking but may eventually need a wheelchair. At least wheelchairs are fairly common in society. When a person is asked to imagine being unable to talk except through a computer, however, it may be too much to accept until the loss of speech renders such forms of AAC absolutely necessary. Persons with ALS may avoid considering electronic devices throughout much of their illness because they look "different" or might call attention to themselves. Even if communication devices are accepted at early and middle stages of the disease, communication using devices at the end of life still may be difficult.

COMMUNICATION USING AAC AT END OF LIFE

Evidence that most persons with ALS cannot communicate with natural speech at end

of life underscores the importance of services supporting the acquisition of appropriate AAC systems and strategies. Early and effective service delivery may improve acceptance and use of AAC. Nordness, Ball, Fager, Beukelman, and Pattee (2010) reported that 88% of the 300 ALS cases they reviewed had received timely AAC evaluations (using the guidelines of Ball et al., 2007). This is encouraging because Ball et al. (2002) suggested that timely referral and appropriate preparation for AAC decisions resulted in 96% acceptance and use of AAC in their study. These high levels of acceptance and use can be contrasted with those reported by Murphy (2004), where only one of 10 persons with ALS offered an AAC device actually used it, with inadequate training identified as a contributing factor.

AAC service delivery for people with ALS is variable across regions (Beukelman et al., 2011). Much of the existing literature comes from the Nebraska database, in which Beukelman et al. (2011) describe services as coming " . . . from a highly integrated intervention system" (p. 4). The Nebraska system shows what is possible when a system includes three regional AAC clinics, an AAC interventionist, routine screenings and education, specific triggers for AAC referral, and ongoing training and support once the AAC system has been obtained. Beukelman et al. suggest that lower levels of AAC acceptance/use reported elsewhere may be the product of differing approaches to service delivery; however, further research is recommended to learn whether the Nebraska experience can be replicated.

The authors of the current article, an SLP and an assistive technology specialist with different ALS Association chapters, have extensive experience with AAC service delivery to persons with ALS as well. In our clinical work, we have observed less technology use toward end of life and changes in communication strategies across the progression of the disease, particularly as death approaches. These changes need to be better understood in order to facilitate end-of-life communication. Ideal informants about communication strategies and changes would be those family members/caregivers who were present as the end of life approached.

We began gathering data relevant to questions about end-of-life communication starting in 2006 as part of a 4-year survey of family/caregivers of person with ALS. This survey was completed under the auspices of the ALS Association in 10 regions with the goal to identify (a) perceived extent of communication difficulties with caregiver and with others; (b) common communication topics; (c) communication strategies used with different partners; and (d) and communication strategies at three time intervals approaching end of life: 2–6 months before death, 4 weeks before death, and the last 1–2 days of a person's life.

METHODS

The complete survey was developed to gather data on the perceptions of family/caregivers of persons with ALS who were now deceased with respect to communication impairment associated with ALS, communication methods or strategies used at different times as end of life approached and with different partners, and their perceptions of their family members' acceptance/use of communication technology, along with their personal perceptions of service delivery experiences. The research was conducted in two waves. The first surveys were sent to family/caregivers receiving services from ALS Association chapters in two locations; the second wave, using a modified and expanded survey, was distributed to service recipients in eight more locations.

Survey tool

The original survey contained 24 questions; it was expanded to 38 questions for the second wave of data collection. Surveys included three demographic items (gender, relationship to the person with ALS who was now deceased, and city/state). The questions used in this analysis are presented in the Supplemental Digital Content (available at http://links.lww.com/TLD/A4). Both

versions of the survey contained items to probe basic problems with intelligibility and communication strategies at the three time intervals preceding death. The one item added for the second wave of data collection was a question about communication strategies with a variety of individuals within the social network.

Procedures

Potential participants were identified through records of cooperating ALS Association chapters across the United States. The only criteria for subject participation were documented affiliation with a chapter and death of the person with ALS in the year prior to completion of the survey (from 2006 to 2010). In the first wave of the project, surveys were sent to eligible persons who had been involved with the Northern Ohio and Greater Philadelphia ALS Association Chapters and whose loved one had died between January 1, 2006, and December 31, 2007. Of the 483 surveys mailed out, 215 were returned, for a response rate of 44.7%.

For the second wave, the authors contacted all other ALS Association Chapters in the United States, explained the project, and asked them to become involved. The chapters in Colorado, Greater New York, Upstate New York, North Carolina, Central and Southern Ohio, Indiana, Iowa, and Rocky Mountains agreed to participate. Surveys were mailed to the 1,104 persons who met study criteria from 2007 to 2010, and 410 surveys were returned, for a return rate of 37.1%. The composite return rate was 39.4% across the two waves of data collection.

In both waves of this study, the mailings to potential participants included a cover letter explaining the general purpose of the study, a print copy of the survey, and a stamped self-addressed envelope for survey return. The cover letter for the second wave also contained a link to SurveyMonkey where the survey could be completed online for those who preferred this option. Mailings were sent at the end of each calendar year.

Data analysis

Data analysis was descriptive in nature. In the following discussion of results, data from the two waves of surveys are combined when the questions are identical and reported separately when only available on the second survey.

RESULTS

Participant relationship to the person with ALS was as follows: 72% spouse; 24.4% other family member; 0.6% friend; and 0.5% other. In the second wave survey, the gender of the person with ALS was asked; 62.3% were male. No further demographic information about participant or person with ALS was collected to reduce possible participant concerns about being identified.

Overall communication intelligibility

Respondents were asked, "From your perspective, how difficult was it for *you* to understand what your loved one was communicating?" A similar question was asked with respect to the difficulty "others" had in understanding the person. These questions did not designate a specific time frame because the goal was elicitation of general impressions of problems understanding the person with ALS and differences between close partners and others. As shown in Figure 1, respondents clearly felt that others had more difficulty understanding the person with ALS than they did themselves. Forty-one percent of respondents reported that other communication partners found it very difficult to understand the person with ALS in contrast to only 22% of the respondents themselves. Similarly, 35% of persons with ALS reportedly had no trouble communicating with the respondent, but only 21% had no trouble communicating with other partners. If all categories of problems communicating (could not speak, very difficult, somewhat difficult) were combined, 79% of persons with ALS were described as having trouble communicating with others and 65% were reported to have trouble communicating with the family/caregiver respondent.

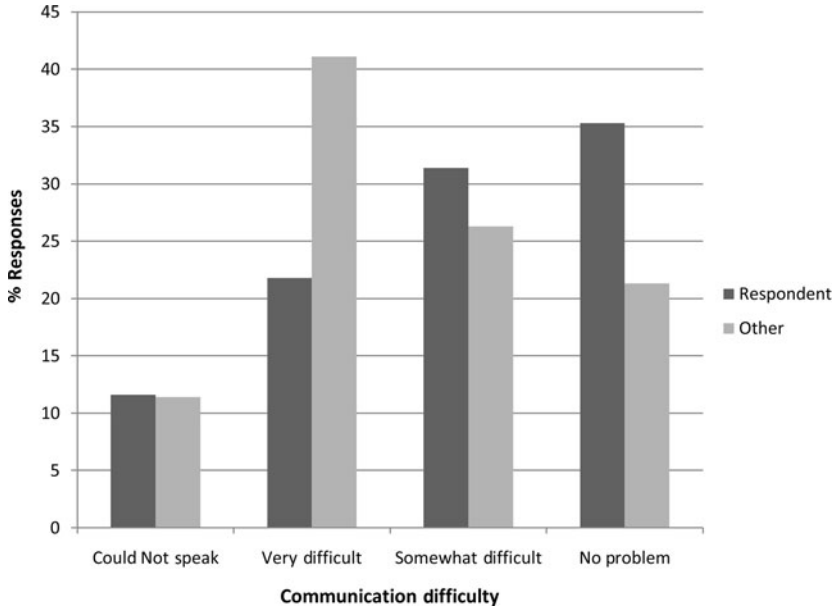

Figure 1. Levels of communication difficulty as reported by respondents for persons with ALS when communicating with respondents and with others during the last few months of life.

Communication topics

Respondents were asked to indicate what content they believed their loved one communicated about the most, from a set of seven topics drawn from the literature on end-of-life communication. More than one response could be selected. The most commonly cited topic of communication was physical needs (73.3%), followed by caregiving issues (42.1%), family issues (39.8%), "usual comments about the day" (33.1%), and pain (32.2%). The least commonly selected topics were spiritual issues (19.1%) and death and dying (16.3%).

Communication strategies

Communication technology

Although many aspects of communication technology referral, use, and resistance were probed in the full survey, the variable of interest here was the percentage of persons with ALS who reportedly obtained an AAC device at any point in the disease progression. On the first wave, 43.3% (93 of 215 respondents) reported the person with ALS obtained a device; on the second wave, 45.4% (186 of 410 respondents) indicated the person with ALS had a device. For both waves, 44.6% obtained devices.

With different partners

Many verbal and nonverbal modalities for communication exist. In the second wave of the survey, respondents were asked if their person with ALS had communication problems, how did they primarily communicate with different partners. Again, no specific time frame was designated for this sequence of questions because general impressions of differences in strategies were of primary interest. Respondents were asked specifically about communication with the family/caregiver respondent, children or grandchildren, friends, health care professionals, health aides or other caregivers, and strangers (see the Supplemental Digital Content [available at http://links.lww.com/TLD/A4)] for question format). Eleven response options were provided: talking, writing, gestures, letter board, communication device, eyeblinks, hand squeeze, sounds, partner-assisted

scanning, flash cards, and no means of communication.

Data are provided in Table 1. Percentages are based on total number of persons responding to each question (total number between 284 and 296) and number of times a particular communication method was selected. Across partners, talking was the most common communication strategy, reportedly used by more than 45% of persons with ALS. Talking was typically followed by writing, gestures, and electronic communication device (in slightly differing orders depending on the partner).

The communication strategy profiles were quite similar across partner groups. Slight differences were found in comparisons of the communication of persons with ALS with strangers versus with other partners. Ten percent of the respondents indicated the person with ALS could not communicate with strangers (the next closest percentage was 3% with friends), and talking was used slightly less with strangers than with other groups. The greatest range of responses across communication partners was for use of writing (10.3% with children/grandchildren to 14.2% with health care providers) and gesture (8.5% with respondent to 13.5% with children/grandchildren and home health aides). Electronic devices were reportedly used least often with home health aides (10.3%), and the highest percentage of use for devices with any partner was 14.2%.

As end of life approaches

Respondents were asked to reflect on the last 6 months of the patient's life and to indicate all of the communication modalities used by the patient within three time frames: 2–6 months before passing (Time 1); 4 weeks before passing (Time 2); and the last 1–2 days of life (Time 3). In theory, these time frames might be distinguished by differences not only in the physical stage in progression of the disease but also in communicative strategy.

Figure 2 shows the percentage of times each communication modality was reported to be used during each of the three time intervals. Percentages are based on total number

Table 1. Communication Strategies Used With Different Communication Partners During the Last few Months of Life (% Total Responses)

Communication strategy	With respondent	With children/ grand children	With friends	With health care providers	With home health aides or caregivers	With strangers
Talking	47.2	46.6	47.3	46.6	46.4	44.6
Writing	13.1	10.2	11.5	14.2	11.3	11.4
Gestures	8.5	13.5	9.5	9.8	12.7	10
Letterboards	6.7	6.9	5.7	6.1	5.8	3.5
Electronic communication device	12.8	13.1	14.2	12.5	10.3	13.1
Eyeblink	7.1	4.4	5.4	5.4	7.6	4.5
Hand squeeze	0.4	0.4	.3	0	0	0
Sound	2.8	2.2	2.4	2	3.4	1.7
Listener-assisted scanning	0	0.4	.7	1	0.7	1
Flash cards	0	0	0	0	0	0
Person could not communicate	1.4	2.6	3	2.4	1.7	10

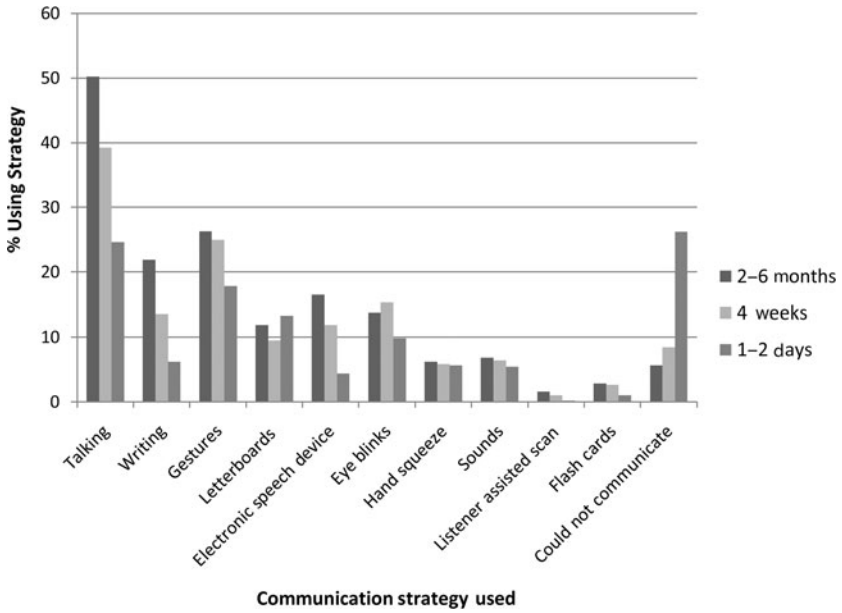

Figure 2. Percentage of persons reported by survey respondents to be using each communication strategy for the three time periods preceding death.

responding to each question. During Time 1, talking was the most common mode of communication (50%), followed by gestures (26.3%) and then writing (21.9%). Use of talking declined consistently across the three targeted time intervals, with less than half of those persons with ALS originally talking at Time 1 still talking in the final day or two. However, this still represented one fourth (24.6%) of those responding. Writing declined even more abruptly than talking, with 6.2% of persons with ALS using writing at Time 3.

At Time 1, 16.5% of persons with ALS were using electronic AAC systems to communicate (roughly one third the number of those using speech). Use of AAC devices declined across the time intervals; at Time 3, less than 5% of persons with ALS (11.4% of the 44% who had access to them) reportedly used devices to communicate. At Time 1, 6% had no means of communication, but this number rose to 26.2% for the last 1–2 days. Use of gestures remained stable and comparatively high (around 25%, second only to talking) moving into the final month, with a decline seen only in the final days (down to 17.8%). Use of letter boards (range: 9.5%–13.2%) and eyeblinks (range: 9.8%–15.3%) stayed at about the same level throughout the three time periods. Sounds and hand squeezes were reportedly used by between 5% and 7% of person with ALS at all time intervals. Flash cards and listener-assisted scanning were used infrequently.

Relevant write-in comments

A number of respondents took advantage of the opportunity to write in additional comments at the end of the full survey. Although analysis of those comments is beyond the scope of this article, a few general observations underscore the importance of the topic to respondents.

Many respondents indicated that lack of communication was the number one obstacle in dealing with ALS. As one caregiver wrote:

> My Mom could deal with the loss of the ability to walk and use her hands, but she was not able to deal with the loss of communication—it would simply make her cry; this represented her biggest loss in life.

Caregivers commented about having to develop their own communication systems

through flash cards, poster boards, and other low-tech systems without professional input or assistance. Numerous respondents stated that people needed to get devices earlier in the diagnosis of ALS. They were frustrated by an insurance system that would not fund a device before the need arose. A number of respondents postulated that if people learned how to use a device early enough, they would become proficient in its use. Some acknowledged that their loved one waited too late and was unable or not interested in using a device in late-stage ALS.

The cognitive component in some cases of ALS seemed to be the most difficult to understand and deal with, based on survey write-in responses. A number of families seemed to struggle to understand the implications and ramifications of frontotemporal dementia in their loved ones. As one caregiver wrote, "ALS care teams should educate/consider the need of patients with frontotemporal dementia. We got the recommended electronic device but by the time we got it, his dementia prevented him from being able to use it." Clearly, helping persons with ALS communicate is not a simple matter.

DISCUSSION

Communication is typically impaired for persons with ALS at some point in the progression of the disorder, during a time when the ability to express needs, preferences, and feelings is critical for the person with the illness, their family and friends, and health care providers. This reality of communication impairment was confirmed in part by data from this study showing that almost two thirds of the respondents indicated that their person with ALS had trouble communicating with them; almost four fifths of the respondents reported the individual had problems communicating with others. In addition to questions about the communication impairment itself, caregivers answered questions about communication topics and strategies used by persons with ALS who were now deceased. Of particular interest was the use of different communication strategies as end of life approaches, given limited research on this topic and the presumed importance of communication at this time.

Surveys were distributed through 10 ALS Association chapters to caregivers. A low participation rate was anticipated, given the loss of a loved one and the experience of caregiver burden and stress. However, the 39% response rate across the two waves was reasonably high. On the basis of written comments at the end of the survey, it was apparent that some caregivers responded out of a sense of ongoing commitment to staff who had helped them. Others wanted someone to listen to them and to know about the person with ALS. A number of respondents felt their participation in the survey was a way for their loved one's experience to make a difference for others in the future.

Communication topics and strategies with different partners

The most common communication topics reflected the physical challenges of ALS itself (physical needs) and the centrality of others in the everyday life of the person with ALS (caregiving and family issues). These are consistent with previous research (Fried-Oken et al., 2003, 2006; Murphy, 2004). For example, Fried-Oken et al. (2003) described common communication purposes such as regulating the behavior of others to address basic wants and needs, giving instructions, clarifying needs, and staying connected. The least commonly selected topics were spiritual issues and death and dying, topics often mentioned in hospice literature as important during end-of-life communications. Because persons with ALS have known since the initial diagnosis that they are dying, they may have been dealing with such issues throughout the disease process. As they get closer to death, simply surviving the day is physically and emotionally challenging and requires considerable focus and energy.

It was anticipated that there would be more diversity in the use of communication strategies across familiar and unfamiliar

partners and that electronic communication devices might be used more often with unfamiliar partners. Surprisingly, communication methods remained relatively consistent across partners, with talking used most commonly, followed by writing, gestures, and then electronic communication devices. When there were small differences in strategy, explanations were easy to hypothesize. For example, writing was used slightly less often with children/grandchildren, who might not be able to read. More persons with ALS were described as unable to talk with strangers than any other group, as would be expected because a stranger would have no previous knowledge to support understanding of the person's speech or strategies. Because the survey did not ask respondents to pinpoint a particular point in the disease progression when answering this question, it is not possible to explore whether topics changed over time.

Communication strategies approaching end of life

Major questions posed in this study were as follows: "Did communication strategies change as end of life approached?" and "What communication strategies were used and when?" Although these data should be interpreted with caution, it seems that changes did occur in communication strategies used most frequently across the three time intervals probed on the survey. At 2–6 months, natural speech was used most often (by about half of the persons with ALS), followed by gesture and then writing. Natural speech use declined consistently to around 25% in the final few days. This figure is higher than that reported by Beukelman et al. (2011) but comparable with much earlier analysis of hospice patient data of Saunders et al. (1981). It should be noted that the use of speech does not necessarily mean that the user communicated successfully with speech output. Many persons with ALS attempt to use speech first as a communicative tool even when it is unintelligible (Richter et al., 2003).

Previous reports suggest that as many as 96% of persons with ALS use AAC devices if referred for evaluation in a timely fashion and supported appropriately (Ball et al., 2002). In contrast, in this survey, less than half of the individuals with ALS obtained an electronic device, a fact that undoubtedly influenced reports of device usage in the last weeks and months. One sixth of individuals with ALS reportedly used AAC devices at 2–6 months before death. By the last few days, less than 5% did so, although this percentage was influenced by the fact that less than half of individuals were reported to have had access to devices. Clearly, high-tech systems were not being used as primary communication tools at that time. Similarly, reported declines in the use of speech and writing toward the end of life are consistent with increasing loss of motor function and exhaustion. Use of gestures demonstrated less decline until the final days.

Logically, loss of motor functions and growing fatigue might lead to increased use of strategies such as eyeblinks, letterboards, and/or listener-assisted scanning. Instead, the use of letterboards remained relatively stable but low, as did eyeblinks; listener-assisted scanning was reported in only a few instances. This is of some concern, given the fact that more than one fourth of individuals had no means of communication in the final days. Although many physical factors, including respiratory deterioration, might be responsible for some breakdown in communication at the end, it is still important for individuals with ALS to be able to communicate wishes or needs to caregivers.

To summarize, as these individuals moved into the final stage of ALS, there was limited use of AAC technology and perhaps not enough use of simpler, unaided, or low-tech communication systems. Instead, speech remained a preference when the person could still talk, and other simpler or less effortful communication strategies (gestures, eyeblinks) played a limited role. A different sample of persons with ALS, one that represents the higher AAC technology usage levels reported in other studies, might yield different percentages but would probably not change the finding that communication

strategies change and this reality must be dealt with.

STUDY LIMITATIONS

There are a number of factors that might influence interpretation of survey outcomes. It was a retrospective survey completed by caregivers, so it can reflect only the perceptions of those individuals (actually their memories of perceptions) and not the actual feelings and perceptions of individuals with ALS or the respondents' perceptions at the time periods of interest. Because surveys were mailed out at the end of each calendar year, the time between death of the person with ALS and caregiver completion of the survey was variable. Given the distress and fatigue associated with coping with the end stages of ALS, willingness to participate as well as actual responses might be influenced by how much time passed between death and receipt of survey. There might also be differences in how well caregivers remembered the communication behaviors of the person with ALS if the survey was completed as much as 11–12 months after death of loved one, although most respondents reported vivid recollection of their time with the person with ALS.

Interpretation of the data presented in this article was affected by not knowing whether the persons with ALS being described by respondents had bulbar versus spinal-onset ALS and the extent of their motor deficits. More generally, additional information about the individual with ALS and the course of his or her illness would have been helpful, particularly over the 6-month time interval used to explore changes in communication strategies. More demographic data (type of residence, geographic setting, use of hospice) could have allowed consideration of the role of external factors in choices of communication strategies. For this article, data were collapsed for reporting purposes, so any differences in response profiles across geographic locations were not analyzed. It is possible that there might be regional variations in a number of measures, depending on the availability of services and support. Knowledge of the level of involvement of the respondent in the care of the person with ALS would have further validated the perceptions each provided.

The framing of some questions may have unintentionally limited response options. For example, the survey presented a fixed set of communication topics, with no "other" category available. Also, with respect to end-of-life changes in communicative strategies, the survey again presented a fixed (although comprehensive) set of response options.

RESEARCH DIRECTIONS

One strength of this study was the size of the respondent pool (both in terms of actual numbers and in response rate). Although survey data concerning high-tech AAC referral, acquisition, and use have not yet been reported in the literature, they will provide additional insight into AAC strategies used by persons with ALS and possible factors influencing utilization.

With respect to the questions explored in this article, future research could build on the study by addressing previously described limitations and extending the outreach of the survey nationally. Additional demographic data related both to the person with ALS and caregiver should be collected, along with more detailed information about the disease progression. Surveys could be mailed to caregivers at a specific time interval after the death of the person with ALS. More opportunities for open-ended responses should be provided to capture the more qualitative aspects of the experience and the nature of the service delivery. Questions about communication topics should be linked to specific time intervals. Because this study provides one of the first snapshots of changes in communicative strategy over time as end of life approaches, there is considerable value in making appropriate changes to the survey and disseminating it nationally.

In addition, both quantitative and qualitative data should be gathered while the person

with ALS is still alive. Quantitative data could be used to document service delivery steps and outcomes, including training to use an AAC device and actual use related to types and amounts of training. However, numbers do not necessarily capture the complex feelings and needs that may drive actual daily use of AAC devices or any other communicative strategy. Qualitative data (e.g., the study of Murphy, 2004, of 15 families dealing with ALS) provide a rich context for understanding those complex feelings and needs for both persons with ALS and their significant others.

Both persons with ALS and caregivers could be asked to report prospectively (rather than retrospectively) use of strategies and topics (with different partners) at predetermined time intervals after diagnosis, perhaps monthly. Patients and caregivers could then be given an opportunity to identify *preferred* modalities of communication, as possibly distinct from what is actually used, and to explain *why* they preferred certain strategies. Observational data also could be collected to document the actual use of different strategies with different partners; however, this would have to be done in a sensitive manner, if at all, because the collection of such data is extremely intrusive to those living with ALS as they approach the end of life.

Finally, it would be helpful to conduct a survey of SLPs to determine current practices and knowledge related to clients with ALS. Practice questions might probe how many SLPs actually work with clients with ALS and their families, what types of services they provide, and what expectations they have regarding AAC for persons with ALS. Knowledge questions could probe information about ALS in general, services for ALS in their geographical area, the range of communicative strategies available to persons with ALS, current best practice recommendations about timing of referral for AAC evaluation, and current research concerning barriers to AAC use. Data from such a study could be used to improve preservice training or design continuing education.

CLINICAL RECOMMENDATIONS

The study data have implications for AAC service delivery to persons with ALS, for end-of-life clinical practice, and for education of health care providers *and* SLPs. On the basis of study outcomes and our collective clinical experience, a series of broad recommendations are provided.

AAC service delivery to persons with ALS must be comprehensive

The process of choosing a communication system must be a fluid one that changes and adapts as the skills, abilities, and needs of the person change. AAC assessment protocols specific to ALS should be developed on the basis of an understanding of current and projected movement limitations of the person with ALS, followed by consideration of the many additional factors known to influence AAC acceptance and use (Brownlee & Palovcak, 2007).

Speech–language pathologists should be responsible for early assessment and client/family education, assistance in developing communication strategies, and monitoring for needed changes in strategies as skills or needs change. Service delivery must include support for persons with ALS and caregivers and appropriate counseling for communication options and barriers. Ideally, persons with ALS should be seen by SLPs who are knowledgeable about both ALS and AAC options.

SLPs can and should be more involved in end-of-life health care

One common misconception about end of life for persons with ALS is that they will already have an established AAC system going into the last 6 months of life (a time frequently associated with enrollment in hospice programs). If they do not have such a system, or they develop communication difficulties very late in the illness, they may not be referred automatically to an SLP. Instead, other health care providers may be the ones attempting to address

communication needs. Unfortunately, many health care professionals have limited understanding of the range of communication options available to persons with ALS. Some may have seen one or more persons with ALS using a high-tech AAC device and believe this is the only viable and preferred option for *all* persons with ALS at all times. Some SLPs also share this assumption, particularly if they have limited experience with individuals with ALS.

In this study, use of electronic AAC systems was reportedly limited even between 2 and 6 months before death and was close to nonexistent in the final days. Thus, it is important for an SLP to be a team member in end-of-life care for persons with ALS whenever appropriate. The clinical obligation does not stop with the AAC evaluation and perhaps initial training but should extend through the course of the illness in support of persons with ALS, their families, and other health care providers. Education is an important tool in ensuring that SLPs remain engaged with ALS clients across the disease progression.

Education is needed for health care providers, as well as for persons with ALS and their families, about communication options

All health care providers serving persons with ALS, including SLPs, need a better understanding of the range of communication options available and the factors influencing acceptance and use of AAC over time. Ideally, clinicians with ALS experience can take the lead in educating others.

SLPs should be proactive in reaching out to hospice and other health care agencies, as well as to professional groups working with patients with ALS and/or their caregivers, to develop collaborative programs and to educate service providers. Education of medical professionals plays a critical role in securing more timely referrals to SLPs for evaluation and ongoing monitoring of speech status and communication needs.

Education should underscore the importance of probing a patient's level of interest in using communication technology and the desirability of a referral to an SLP for assessment *before* presenting the client with a device and relatively early in the process. End-of-life health care providers in particular need to understand that learning a complex SGD when in the very late stages of ALS is like learning a new language, requiring time, training, and patience, all of which the person with ALS does not have. Practitioners do not always recognize that persons with ALS are often fatigued and unmotivated to learn new things in the last stages of the disease and that caregivers may be overwhelmed by the task of caring for their loved one.

Given the variety of communication options selected in this study, it is clear that health care providers must be particularly knowledgeable about factors that influence the use of communication strategies over the course of the illness. Communication needs and preferences are unique to individuals; having a diagnosis of ALS does not change that reality. Communication strategies must be ones the patient and significant others endorse and feel comfortable with, ones that allow needs to be met and interactions to succeed. Some of the newer socially acceptable handheld technologies (e.g., smartphones, iPads) with communication applications may reduce resistance to technology in the future.

Many communication strategies do not require the use of sophisticated technology. Education can highlight the range of communication strategies available, even in the last weeks and days when simple, unaided, or low-tech solutions may work best. This survey documents that these solutions are being used but perhaps not as frequently as possible or as effectively as desirable. SLPs must be proactive in educating others about communication options throughout the disease process.

One good example of a simple communication strategy with universal applications is the letterboard, used by few in the study, yet clinically observed to be a powerful

communication tool, particularly within the last days and weeks. A letterboard is any paper, frame, and so forth, that contains the letters of the alphabet in some configuration, usually rows. Some assume that letterboards can be used only by a person who can spell out a message by pointing physically to each letter, ruling out those with limited hand/finger movement. However, letterboards also can be accessed through partner-assisted scanning. With this approach, the partner points to a row and the person with ALS in some fashion (eyeblink etc.) indicates whether or not the letter is on that row. If the letter is on the row, the partner moves across the row, pointing to each letter and waiting for confirmation from the patient. Although slow, the approach works for most basic communication and it remains a good example of simple but effective communication strategies toward end of life.

Education of health care providers and those living with ALS also should include discussion of the potential cognitive component of ALS and its effects of communication and other aspects of everyday life. Patients and families who believe that ALS never affects cognition may not acknowledge any cognitive problems when faced with them. Although this is understandable, given the overwhelming motor deficits already being confronted, if the family is unwilling to accept the evidence of cognitive deficits, they may push the patient or health care team into an inappropriate communication system.

CONCLUSIONS

Most persons with ALS and their caregivers eventually face a breakdown in communication through natural speech. This study documented changes perceived retrospectively by caregivers in the use of a range of communication strategies across the last 6 months of life. The results showed that one fourth of persons with ALS were unable to communicate immediately before death, but a comparable percentage of persons with ALS were still using natural speech at that time.

Although many assume that electronic AAC devices are the solution to communication breakdown, less than half of the persons with ALS in this study had acquired an AAC device, and most did not use one at the end of life. A number of research and service delivery needs related to end-of-life communication strategies were identified, including the need for earlier and appropriate services focused on communication needs (not just swallowing), as well as questions about the preparation of SLPs to meet the communicative needs of persons with ALS and their loved ones. Finally, recommendations were made about how to support the communication of persons with ALS, particularly in the last days and weeks.

REFERENCES

ALS Association. (2005). *ALS, cognitive impairment (CI) and frontotemporal lobar dementia (FTLD): A professional's guide*. Retrieved from http://www.alsa.org/assets/pdfs/fyi/cognitive_impairment_prof.pdf

ALS Association (n.d.). *Forms of ALS*. Retrieved from http://www.alsa.org/about-als/forms-of-als.html

Ball, L. J., Beukelman, D. R., & Bardach, L. (2007). Amyotrophic lateral sclerosis. In D. R. Beukelman, K. L. Garrett, & K. M. Yorkston (Eds.), *Augmentative communication strategies for adults with acute or chronic medical conditions* (pp. 287–316). Baltimore, MD: Paul H. Brookes Publishing Co.

Ball, L. J., Beukelman, D. R., & Pattee, G. L. (2002). Timing of speech deterioration in people with amyotrophic lateral sclerosis. *Journal of Medical Speech-Language Pathology*, *10*, 231–235.

Ball, L. J., Beukelman, D. R., Ullman, C., Maassen, K., & Pattee, G. L. (2005). Monitoring speaking rate by telephone for persons with amyotrophic lateral sclerosis. *Journal of Medical Speech-Language Pathology*, *13*, 233–240.

Ball, L. J., Nordness, A. S., Fager, S. K., Kersch, K., Mohr, B., Pattee, G. L., & Beukelman, D. R. (2010). Eye-gaze access to AAC technology for people with amyotrophic lateral sclerosis. *Journal of Medical Speech-Language Pathology*, *18*, 11–23.

Ball, L. J., Willis, A., Beukelman, D. R., & Pattee, G. L. (2001). A protocol for identification of early bulbar

signs in amyotrophic lateral sclerosis. *Journal of the Neurological Sciences*, *191*, 43–53.

Beukelman, D. R., Fager, S., & Nordness, A. (2011). Communication support for people with ALS. *Neurology Research International*. doi: 10.1155/2011/714693

Beukelman, D. R., Yorkston, K., Hakel, M., & Dorsey, M. (2007). *Speech Intelligibility Test (SIT)* [computer software]. Lincoln, NE: Madonna Rehabilitation Hospital.

Brownlee, A., & Palovcak, M. (2007). The role of augmentative communication devices in the medical management of ALS. *NeuroRehabilitation*, *22*, 445–450.

Doyle, M., & Phillips, B. (2001). Trends in augmentative and alternative communication use by individuals with amyotrophic lateral sclerosis. *Augmentative and Alternative Communication*, *17*, 167–178.

Elman, L. B., & Grossman, M. (2007). Neuropsychiatric features of amyotrophic lateral sclerosis. *NeuroRehabilitation*, *22*, 425–429.

Elman, L. B., McCluskey, L., & Grossman, M. (2008). Motor neuron disease and frontotemporal lobar degeneration: A tale of two disorders linked to TDP-43. *Neurosignals*, *16*, 85–90.

Fried-Oken, M., Fox, L., Rau, M. T., Tullman, J., Baker, G., Hindal, M., & Lou, J. (2006). Purposes of AAC device use for persons with ALS as reported by caregivers. *Augmentative and Alternative Communication*, *22*, 209–221.

Fried-Oken, M., Fox, L, Rau, M. T., Tullman, J., & Lou, J. S. (2003). Using augmentative communication technology to interact: Why caregivers and persons with ALS converse. *ALS and Other Motor Neuron Disorders*, *4*, 219.

Gauthier, D. M. (2008). Challenges and opportunities: Communication near the end of life. *Medsurgical Nursing*, *17*, 291–296.

Hillel, A., Dray, T., Miller, R., Yorkston, K., Konikow, N., Strand, E., & Browne, J. (1999). Presentation of ALS to the otolaryngologist/head and neck surgeon: Getting to the neurologist. *Neurology*, *53*(8), S22–S25.

Hinckley, J. J. (2008). *Narrative-based practice in speech-language pathology*. San Diego, CA: Plural.

Levin, T. T., Moreno, B., Sylvester, W., & Kissane, D. W. (2010). End-of-life communication in the intensive care unit. *General Hospital Psychiatry*, *32*, 433–442.

Luterman, D. M. (2001). *Counseling persons with communication disorders and their families* (4th ed.). Austin, TX: Pro-Ed.

Mathy, P., Yorkston, K. M., & Gutmann, M. L. (2000). AAC for individuals with amyotrophic lateral sclerosis. In D. R. Beukelman, K. M. Yorkston, & J. Reichle (Eds.), *Augmentative and alternative communication for adults with acquired neurologic disorders* (pp. 183–231). Baltimore, MD: Paul H. Brooks Publishing Co.

Miller, R. G., Jackson, C. E., Kasarskis, E. J., England, JD, Forshew, D, Johnston, W, & Woolley, SC. (2009). Practice parameter update: The care of the patient with amyotrophic lateral sclerosis: multidisciplinary care, symptom management, and cognitive/behavioral impairment (an evidence based review): Report of the Quality Standards Subcommittee of the American Academy of Neurology. *Neurology*, *73*, 1227–1233.

Murphy, J. (2004). "I prefer contact this close": Perceptions of AAC by people with motor neuron disease and their communication partners. *Augmentative and Alternative Communication*, *20*, 259–271.

National Institute of Neurological Disorders and Stroke. (2010, February). *ALS (amyotrophic lateral sclerosis) fact sheet* [NIH Publication No. 10-916]. Washington, DC: NINDS.

Nordness, A. S., Ball, L. J., Fager, S., Beukelman, D. R., & Pattee, G. L. (2010). Late AAC assessment for individuals with amyotrophic lateral sclerosis. *Journal of Medical Speech-Language Pathology 18*, 48–54.

Ogawa, T., Tanaka, H., & Hirata, K. (2009). Cognitive deficits in amyotrophic lateral sclerosis evaluated by event-related potentials. *Clinical Neurophysiology*, *120*, 659–664.

Prince-Paul, M. (2008). Relationships among communicative acts, social well-being, and spiritual well-being on the quality of life at the end of life in patients with cancer enrolled in hospice. *Journal of Palliative Medicine*, *11*, 20–25.

Richter, M., Ball, L. J., Beukelman, D. R., Lasker, J., & Ullman, C. (2003). Attitudes toward communication modes and message formulation techniques used for storytelling by people with amyotrophic lateral sclerosis. *Augmentative and Alternative Communication*, *19*, 170–186.

Saunders, C., Walsh, T., & Smith, M. (1981). *Hospice care in the motor neuron diseases.* London, England: Edward Arnold.

Shadden, B. B., Hagstrom, F., & Koski, P. (2009). *Neurogenic communication disorders: Life stories and the narrative self*. San Diego, CA: Plural. Retrieved from http://onlinelibrary.wiley.com/doi/10.1002/hbm.21242/full

Yorkston, K. M., Miller, R. M., & Strand, E. A. (1995). *Management of speech and swallowing in degenerative diseases*. Tucson, AZ: Communication Skill Builders.

Thesen, T., Leontiev, O., Song, T., Dehghani, N., Hagler, D. J., Huang, M., Buxton, R., & Halgren, E. (2012). Depression of cortical activity in humans by mild hypercapnia. *Human Brain Mapping*, *33*, 715–726.

Zago, S., Poletti, B., Morelli, C., Doretti, A., & Silani, V. (2011). Amyotrophic lateral sclerosis and frontotemporal dementia (ALS-FTD). *Archives Italiennes de Biologie*, *149*, 39–56.

Zeitlin, D. l., Abrams, G. M, & Shah, B. K. (199S). Use of augmentative/alternative communication in patients with amyotrophic lateral sclerosis. *Neurorehabil Neural Repair, 9*, 217-220.