

### **Critical Review:**

## **Does early acceptance and subsequent decision-making regarding augmentative and alternative communication (AAC) for persons with amyotrophic lateral sclerosis (ALS) positively affect the patient's communication in the end-stages of the disease?**

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This critical review examines the effects of AAC use for persons with ALS on communication in the end-stages of their lives. The following aspects are of interest: a) early acceptance of the loss of speech accompanying the disease, b) the timeliness of referral for an AAC assessment and c) subsequent decision-making regarding the use of AAC devices. Research studies were all of a Level IV qualitative research design. Overall, the evidence suggests that persons with ALS who have a timely referral for an AAC assessment and begin to use their AAC device soon after their diagnosis experience a prolonged ability to communicate in the end-stages of their lives. More research is necessary to understand the evidence on the advantages of prolonged communication in the end-stages of the patients' life. This critical review includes recommendations for future research as well as implications for clinical practice in the field of speech-language pathology.

### ***Introduction***

Amyotrophic lateral sclerosis (ALS) is a rapidly progressive degenerative motor neuron disease (Ball, Beukelman, & Pattee, 2003), which results in a motor speech disorder (Beukelman, Fager, & Nordness, 2011). As the disease evolves, approximately 80 to 95% of people with ALS (PALS) will be unable to meet their daily communication needs using natural speech. Communication is important not only to share basic wants and needs, but also to maintain social relationships. Therefore, it is essential that with this loss of natural speech, the person with ALS does not also lose the ability to communicate in the end-stages of his or her life. In order to provide the possibility of communication, augmentative and alternative communication (AAC) options are available for persons with ALS.

With more recent developments in ventilation procedures for this population, life expectancy for persons with ALS has been extended, and thus so is the use of AAC devices (Beukelman et al., 2011). Early acceptance of the loss of speech that accompanies ALS and subsequent decision-making regarding AAC is becoming increasingly important with this extension of life expectancy, allowing PALS to communicate longer. Persons with ALS must be presented with AAC options and assessed for AAC immediately following their initial diagnosis. Then PALS are able to start using the AAC device and become comfortable using it and communication partners are given time to learn the device. With this early decision-making regarding AAC, the person with ALS will then be able to use the device in order to communicate his or her basic needs and wants.

The person with ALS will also be able to maintain social relationships in the end-stages of his or her life when natural speech is no longer a viable or functional means of communication. Thus, the person with ALS and his or her communication partners will have the potential for prolonged communication in the end-stages of his or her life.

### ***Objectives***

The primary objectives of this paper are to critically evaluate existing literature regarding persons with ALS, their acceptance of the loss of speech accompanying their disease, the timeliness of the referral for an AAC assessment and how subsequent decision-making regarding the use of AAC devices affects the person's communication in the end-stages of the disease. The secondary objective is to propose evidence-based practice recommendations to speech-language pathologists and other professionals who are providing AAC assessment and intervention to PALS.

### ***Methods***

#### Search Strategy

Studies selected for inclusion in this critical review were chosen from the computerized databases: ScholarsPortal, PubMed, PsychINFO, CINAHL, Scopus, Web of Science and Google Scholar; using the following search strategy:

[(AAC) OR (alternative communication) OR (augmentative communication) OR (augmentative and alternative communication)] AND [(ALS) OR (amyotrophic lateral sclerosis) OR (Lou Gehrig's

disease) OR (motor speech disorders)] AND [dysarthria]

Reference lists of previously searched articles and related systematic and literature reviews were also used to obtain other relevant studies. The search was limited to articles written in English.

#### Selection Criteria

Studies included in this critical review paper were required to discuss the acceptance of ALS by the patient, the timeliness of their AAC assessment, the subsequent decision-making regarding AAC, and the outcomes of AAC use on communication. Research subjects included patients with ALS and/or their communication partners. No limits were set on the demographics of the research participants, type or stage of ALS, or study designs; however, only articles dated after 2000 were chosen due to changes in AAC availability and technology in the current century. Literature reviews, position papers, and meta-analyses were excluded from this critical review, except to gain information.

#### Data Collection

Results of the literature search yielded eight Level IV qualitative research studies that met the above selection criteria. Levels of evidence were based on the Oxford Centre for Evidence-based Medicine Levels of Evidence (OCEBM, 2009). These included interviews and document reviews.

### **Results**

Although Level IV qualitative research is a lower level of evidence for study design (OCEBM, 2009), it is the most appropriate method of conducting research for this population of individuals, since ALS is a very progressive disease and many PALS only live for two to five years after diagnosis (Ball et al., 2010). All of the studies in this critical review are Level IV qualitative research.

Ball, Beukelman, Anderson, Bilyeu, Robertson and Pattee (2007) documented the duration of AAC technology use by 45 PALS and identified the factors that influenced the duration of AAC use, through retrospective document review and interviews of surviving family and caregivers. These researchers concluded that AAC technology plays a sustained role in supporting PALS and their caregivers in functional communication; however, there is considerable variability in the use of AAC devices. They also reported that the decision to choose invasive mechanical ventilation by the person with ALS appears to have a substantial impact on the

duration of AAC use, meaning that PALS who use mechanical ventilation in the end-stages of their life will use their AAC device longer. Additionally, these authors reported that the timeliness of the referral to receive an AAC assessment has an important impact on the duration of AAC use, indicating that those who use AAC for less than ten months had a late referral for an AAC assessment. All of the participants used AAC until within one month of death, but toward the end of life, an increased use of lower-technology AAC strategies were used.

Ball et al. (2007) included persons with ALS who had accepted invasive ventilation and continued to live at the time of publication. These participants continued to use their AAC technology as well; thus, reported duration of AAC use was not finalized. This could be considered a weakness in this particular research study because it makes it difficult to compare the length of AAC use. When deciding the methodology of the study, the researchers established an endpoint date for data collection as the date when the PALS started the use of full-time ventilation. Therefore, only PALS who were not using mechanical ventilation regularly were included as research participants. However, the researchers then decided to include those patients who were on long-term mechanical ventilation and continued to use their AAC equipment at the time the data was collected. This change in study methods after the research had begun may have altered the results of the research. It does however demonstrate that the decision to choose invasive ventilation appears to have a substantial impact on the duration of AAC use.

In the second study, Nordness, Ball, Fager, Beukelman and Pattee (2010) identified people with ALS for whom the AAC assessment was delayed and documented the factors that resulted in this late AAC assessment. Nordness et al. (2010) stated that the use of a speech-generative device (SGD) can improve individuals' quality of life by fostering independence, improving self-esteem, helping to maintain relationships, reducing anxiety and frustrations, thus allowing for discussions about medical procedures and financial concerns and participating in their own decision-making process. Using a questionnaire they developed for the purpose of this study, the authors reviewed data found in the Nebraska ALS Database for 28 persons with ALS (representing 12% of PALS who had been entered into the database in the past ten years) who had a late referral for an AAC assessment. Of this 12%, 93% received their referral for an AAC assessment late due to difficulty in

obtaining the referral (e.g. doctor would not refer them). The authors found that there is a great need for “AAC finders”, who are usually medical personnel with an awareness of the limitations in communication associated with ALS and the intervention options available. These “AAC finders” will work with PALS and their caregivers to access communication services in a timely manner. These authors also found that the most common reason for the delayed referral was a delay in referral by the medical personnel. Persons with ALS need to make decisions regarding AAC while they are still able to speak, thus there needs to be a proactive decision-making strategy. Medical personnel also need to use the standard criteria provided in order to make a timely AAC referral.

One limitation in the study by Nordness et al. (2010) study is that the demographics of the participants were not stated explicitly, such as age, ALS type, and socio-economic status, thus it is unknown how or if patient demographics may have affected the results. The authors also stated a need to investigate AAC use and the timeliness of referrals for women with ALS vs. men with ALS, as they found a discrepancy in these results. The authors were also unable to compare their results to other studies or geographic regions, thus limiting their conclusions. It is important that this research be continued, especially around the theme of improving patient’s quality of life.

In a related study, Ball, Beukelman and Pattee (2002) examined the timing of speech deterioration in 101 PALS as it related to the time since their ALS diagnosis, the type of ALS, and their speaking rate. The results of this study indicated that speaking rate may be an important predictor of subsequent speech performance for PALS, regardless of ALS type. These results support previous conclusions that the speaking rate of 100 words per minute is an important milestone in the decline of speech intelligibility.

Ball and her colleagues (2002) discussed future research which would investigate two differing hypotheses in order to understand the relationship between speaking rate and intelligibility. There were no limitations found in this research.

Next, Ball, Beukelman and Pattee (2004) reviewed the use of AAC technology by 50 PALS over a 4-year time period and reported how the patients accepted this AAC technology. The results of the interviews revealed that only 4% of the participants rejected AAC technology altogether, while 96% of

the participants accepted AAC technology, either immediately (90%) or after some delay (6%). There was also no pattern of discontinuation of AAC use found by any of the participants. The authors concluded that PALS chose to use AAC because of a desire to communicate with others, a desire for community involvement, and a desire to continue employment or to volunteer.

Ball et al. (2004) effectively stated the areas that required further research, such as comparing varied clinical practices and intervention models, as well as acceptance patterns by PALS who are underrepresented minorities. These authors were the only researchers to acknowledge the influence and potential biases of the clinician (or first author), which included clinical experience, academic credentials, and type of intervention program selected. They recognized the need for additional scrutiny of all potential biases of the researchers and the participants in order to justify clinical outcomes.

In another study, Ball, Nordness, Fager, Kersch, Mohr, Pattee, and Beukelman (2010) described the acceptance, training and communication patterns of 15 people with ALS who used eye-gaze tracking access for communication with their AAC device. The participants in this study independently selected Eye-gaze Response Interface Computer Aid (ERICA) with different communication software after their AAC assessment. A questionnaire was used to examine overall effectiveness of this eye-gaze technology for functional communication. The results demonstrated that 14 of the 15 participants had success with the eye-gaze tracking access mode, indicating that PALS may maintain functional eye control that supports their ability to communicate. These results are important in showing the current trends in AAC use for persons with ALS, especially with the technological advances, including mechanical ventilation, for this population.

Ball and colleagues (2010) stated that the results and outcomes of their study of 15 participants were encouraging and highlighted the need for further research into eye-gaze tracking access for communication for persons with ALS. This research may benefit from longitudinal examination of the participants to see if ERICA remains a viable AAC option for PALS throughout the progression of the disease. One limitation of this study is that the authors only examined PALS who chose the ERICA eye-gaze tracking device, despite there being other eye-tracking systems available. Therefore, the population could be more representative of this area of research by including and comparing individuals

who chose different eye-tracking systems, such as the Eye-Gaze Edge by LC Technologies.

Doyle and Phillips (2001) used four case studies to describe the tendency of persons with ALS to use unaided and low-technology AAC systems early in the disease, and higher-technology devices later on in the disease process. These authors also demonstrated a trend to revert back to lower-technology AAC systems in the later (end-stages) of the disease, when reduced motor skills are present and there is a diminished number of communication topics and communication partners available. Doyle and Phillips also identified a need for continued research in the area of alternate access methods for PALS in the later stages of the disease and the development of switches. This research has subsequently been taken up by other researchers, such as Ball et al. (2010).

The study by Doyle and Phillips (2001) contains beneficial information; however, the small sample size is a weakness and limitation of this study. Drawing conclusions on the trends in the types of AAC approaches used by persons with ALS based on four case studies is a difficult inference to make. This research should be continued and more case studies should be reviewed in order to support the conclusions made by these authors.

Fried-Oken, Fox, Rau, Tullman, Baker, Hindal, Wile and Lou (2006) conducted survey research of 34 caregivers supporting 26 PALS in order to describe the purposes of AAC device use. Each caregiver evaluated the importance, mode and frequency of use of 17 purposes of communication using a checklist. The results indicated that the most important purposes for persons with ALS to communicate using their AAC device were to have their basic needs met, to give instructions to others, and to clarify their needs. The patients most frequently used the mode of face-to-face spontaneous conversation, even though it was slow, lacked permanence and had higher demands on the communication partners.

In this study by Fried-Oken and her colleagues (2006), the description of how the data were analyzed and what methods were used was vague, although the findings were consistent with and reflective of the data. They simply stated that they used descriptive statistics and correlational procedures using SPSS. The authors stated that a "rigorously designed survey tool" was used to collect responses; however, there is also no description of how this survey tool demonstrated a rigorous design. The authors also discussed quality of life for PALS and their

caregivers, but did not use a quality-of-life measure to infer this implication.

Finally, Murphy (2004) presented some of the findings from a three-year research project examining the communication of 15 people with ALS and their partners in their own homes. Multiple strategies and techniques used by patients with ALS and their families in order to communicate are identified, including low-tech AAC, non-verbal strategies, speaking strategies and conversation strategies. The development and maintenance of social closeness was found to be the most important purpose of communication. This study implied the need for speech and language intervention for not only the person with ALS but for their communication partners to help them learn strategies to facilitate an effective conversation.

Murphy (2004) implied that persons with ALS and their communication partners would benefit from speech and language therapy that does not only focus on swallowing difficulties. However, one limitation of this study was the lack of description on how to turn the focus of speech and language therapy for PALS from swallowing difficulties to communication. This research did not have many theoretical connections, and the findings did not contribute to theory development.

While the studies analyzed are valuable to obtain the experiences and opinions of persons with ALS, all of the research that was critically reviewed has some common limitations. Firstly, the authors did not use an explicit theoretical framework to present their information (e.g. phenomenology or grounded theory), thus it is difficult to know what perspective was taken for interpreting the data and results. Secondly, very few of the researchers acknowledged any potential biases that may have influenced their understanding and interpretation of the data collected. Thirdly, all of the study sample sizes are small. Finally, many of the authors drew their participants from the Nebraska ALS Database or a regional clinic sponsored by the Muscular Dystrophy Association in Nebraska; as a result, the research samples may not have been representative of a widespread geographical area. It is important to note that it is challenging to acquire a representative sample for PALS who use AAC. The researchers themselves present another limitation in the reviewed research. Most research in this area of study is conducted by the same team of researchers, and thus the same potential biases may have influenced their research in each study. A final common limitation in seven of the eight studies was a lack of description of the

statistical treatment of the data. Therefore, the research would be very difficult to replicate in that it is unknown how the researchers analyzed the data. Given these common limitations, the results of this review should be interpreted with caution.

### ***Discussion***

As stated earlier, the aim of this critical review was to examine whether early acceptance and subsequent decision-making regarding augmentative and alternative communication (AAC) for persons with amyotrophic lateral sclerosis (ALS) positively affect the patient's communication in the end-stages of the disease. Eight studies were reviewed, revealing compelling and suggestive evidence that early acceptance of AAC use for persons with ALS, including a timely referral for an AAC assessment, allowed a prolonged ability to communicate in the end-stages of the disease. Small sample sizes were one of the main methodological concerns for all of the studies with a range of 4 to 101 participants. These small sample sizes resulted in low statistical power, meaning that it is more difficult to detect a meaningful effect when it existed. Thus, with larger sample sizes, it would be easier to generalize how AAC use affects the communication of persons with ALS in the end-stages of their lives, and more specifically whether early acceptance of the imminent loss of speech, a timely AAC assessment and subsequent decision-making regarding AAC devices and AAC use does positively affect communication later in the disease progression. A second major limitation in this research was a potential unrepresentative population sample. Four of the studies drew participants from a regional clinic sponsored by the Muscular Dystrophy Association in Nebraska and one study obtained its participants from the Nebraska ALS Database. Therefore, it is unknown if these conclusions would be relevant in other geographical locations. Another common methodological constraint was a lack of description with regards to how the data were analyzed. Only one of the research groups described how they analyzed their results and the statistical treatment of the data and even this description was ambiguous. Consequently, it would be challenging to replicate these research studies. All these factors likely affected the reliability of the results and need to be taken into consideration when interpreting the findings.

Despite concerns with methodology, the consistency of the evidence from all the studies suggests positive effects of AAC use on communication for persons with ALS. Persons with ALS who accept the idea

that they will lose their ability to use natural speech as a functional mode of communication and thus obtain a timely referral for an AAC assessment and begin using AAC devices soon following diagnosis reported an extended capacity to communicate throughout the disease progression. This ability to communicate allowed them to share their wants and needs, make decisions regarding their own medical treatment, and maintain social relationships. Therefore, it can be assumed that these persons with ALS were more satisfied in the end-stages of their disease and had an improved quality of life before their death. In particular, with more recent developments in AAC access methods and the use of invasive mechanical ventilation that is prolonging the life of PALS, the acquisition and use of AAC devices early in the disease progression is important. Persons with ALS are now able to continue using their AAC device to communicate for longer.

More research needs to be completed in the area of AAC use with ALS in order to better understand the evidence on the advantages of prolonged communication in the end-stages of the patient's life, particularly research in improvements to quality of life because the person with ALS is able to preserve the ability to communicate his or her wants and needs, assist in medical decisions via communication and maintain social relationships.

### ***Conclusion***

AAC devices are commonly used during the progression of ALS and therefore associated steps and outcomes should be researched. Nevertheless, research is limited in the area of AAC and ALS. Eight studies were critically examined in this review, which explored the following: early acceptance of ALS diagnosis, timeliness of referral for AAC assessment, and decision-making regarding the device and use of the device. Some of the studies exhibited numerous limitations and therefore their results should be interpreted cautiously. Future research should focus on the advantages of prolonged communication in the end-stages of the patient's life, particularly research in improvements to quality of life for persons with ALS. Research studies mentioned in this critical review could be used as a starting point for future research.

### ***Clinical Implications***

The reviewed literature revealed several common themes which speech-language pathologists and other professionals should take into consideration, albeit with caution:

- a) PALS need to be referred for an AAC assessment before their speaking rate declines lower than 100 words per minute in order to ensure the patient has access to an AAC device prior to losing the ability to use natural speech as a viable means of communication.
- b) Clinicians and medical personnel need to be pro-active in providing a referral for an AAC assessment to all PALS immediately following diagnosis.
- c) PALS need to be provided with low-tech and high-tech AAC options and should learn how to use both of these options to enable them to change their device use with the decline in motor abilities and if they choose to have mechanical ventilation.

When clinicians know the positive effects AAC has on the ability to communicate when natural speech is not functional, they can ensure their clients have full access to AAC information and devices, thus providing a better quality of life.

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