Critical Review:
Impact of augmentative and alternative communication on the quality-of-life of individuals with amyotrophic lateral sclerosis

Emma Carruthers
M.Cl.Sc (SLP) Candidate
University of Western Ontario: School of Communication Sciences and Disorders

This critical review examines whether the use of alternative and augmentative communication (AAC) has an impact on the quality of life of individuals with amyotrophic lateral sclerosis (ALS). A literature search of computerized databases resulted in six articles meeting inclusion criteria. Critical appraisal of the articles was completed by taking into account study design, methodology, outcome measures, and statistical evidence. Overall, current research has found that AAC interventions improve or at least mitigate the changes in quality-of-life of individuals with ALS. In addition, evidence has been found that quality-of-life may be impacted by both early implementation of low-tech AAC and later use of high-tech AAC options such as eye tracking communication devices.

Introduction

Amyotrophic lateral sclerosis (ALS) is a rapidly progressing disease that causes loss of voluntary muscle strength. Patients with ALS have an average survival rate of 3-5 years after diagnosis. By the end of life, in addition to severe motor, respiratory, and swallowing impairments, approximately 80 to 95% of individuals with ALS become unable to meet their communication needs through natural speech (Beukelman, Fager, & Nordess, 2011).

As verbal communication abilities deteriorate, augmentative and alternative communication (AAC) strategies become more important. AAC strategies can range from no-tech (e.g. gestures, facial expressions) to low-tech (e.g. alphabet boards, partner-assisted scanning) to high-tech devices (e.g. tablets, eye-tracking devices, brain-computer interface). AAC enables individuals to communicate personal and medical decisions, maintain social interaction and closeness, and reduce caregiver burden near the end of life (Linse, et al., 2018).

Given that there is no curative therapy for ALS available, the main therapeutic focus is to improve the quality of life of individuals with ALS and reduce caregiver burden. In this population, level of verbal communication is one of the many variables strongly associated with patient's quality-of-life (Felgoise, et al., 2015). Therefore, this review will evaluate whether or not AAC use can improve quality-of-life in individuals with ALS, as well as evaluate what other factors, such as timing and type of AAC, have been found to impact this relationship.

Objectives

The primary objective of this paper is to critically evaluate the existing literature regarding the impact of AAC on the quality-of-life of individuals with ALS.

The secondary objective is to examine the impact of type and timing of AAC referrals on quality of life in individuals with ALS.

Methods

Search Strategy

The computerized databases UWO Library and PubMed were searched using the following search strategy: [(amyotrophic lateral sclerosis) OR (ALS) AND (augmentative OR alternative communication) OR (AAC) OR (communication device) OR (eye-tracking device) AND (quality of life)].

Selection Criteria

Papers selected for review investigated the impact of any form of assistive communication on quality-of-life of individuals with ALS. Articles selected had to include some form of instrument that specifically measured quality-of-life, regardless of their specific definition of quality-of-life.

Data Collection

The results of the literature search yielded six papers, with various levels of evidence, meeting the selection criteria: one mixed randomized clinical trial (level 1), one case control study (level 2b), one cohort study (level 2b), two single group studies (level 3), and one survey study (level 4).

Results

Londral et al. (2015) conducted a mixed-design randomized clinical trial to measure the effect of early intervention with an assistive communication device on the quality-of-life of ALS patients and their caregivers. Participants in the study included twenty-seven patients with bulbar-onset ALS (a form of the disease that impacts speech and swallowing first) and seventeen of their caregivers. Participants were randomized into two groups. The early intervention group received a tablet AAC device after baseline assessment, whereas the late intervention group received an AAC device according to protocols set by the center (when dysarthria scores reached 0 or 1 on the ALS Functional Rating Scale-revised (ALSFRS-R)). Patient outcomes were measured using the McGill Quality of Life questionnaire (MQoL), the ALSFRS-R, and the modified Communication Effectiveness Index (CETI-m). Caregiver outcomes were measured using the MQoL and the World Health Organization Quality of Life (WHOQOL-BREF).

Results found that participants in the early intervention group had higher scores on the MQoL questionnaire when compared to the late intervention group, especially on the existential well-being and psychological symptoms domains. Caregivers in the early intervention group had higher scores on the support domain of the MQoL, whereas no significant differences on the WHOQOL-BREF were found between the two groups.

Strengths of this study include clearly stated inclusion criteria and methodology. Further, researchers ensured that no significant differences in bulbar decline were found between the early and late intervention groups. This is important because bulbar decline was found to be linked to decreases in quality-of-life. Appropriate statistical tests were employed.

However, no measurements were included in the study for the number of hours per day that devices were being used by participants. Further, the participants were 81.5% female, had low education levels, and poor technology training, which could effect the generalizability of the results.

Overall, this study shows compelling evidence that early intervention with an assistive communication device has a positive impact on quality-of-life in ALS patients.

Hwang et al. (2014) conducted a case-control design study to measure whether the use of an eye-tracking communication device improves quality of life and reduces caregiver burden for individuals with ALS and their caregivers. Participants included twenty Taiwanese patients with ALS who used a phonetic board for communication and their caregivers. Ten participants were arbitrarily selected to use an eye-tracking communication device, whereas the other ten participants continued to use their phonetic board. Assessment of quality-of-life, depression, and caregiver burden was conducted before and after the intervention period using the Taiwanese Depression Questionnaire (TDQ), the Revised ALS Specific Quality of Life Instrument (ALSSQOL-R), and the Caregiver Burden Scale (CBS).

Results found higher TDQ scores in the user group as compared to the non-user group, indicating that those who used an eye-tracking communication device were generally less depressed. Users of the eye-tracking communication device were also found to have higher ALSSQOL-R scores, indicating a better quality of life, as compared to those who simply used a phonetic board. Caregiver burden, as measured by CBS scores, was also significantly improved in the caregivers of users of the eye-tracking device.

Researchers in this study chose a culturally relevant questionnaire, the TDQ, to study their outcome measures in a specific population. Appropriate statistical tests were employed. Researchers also included the measurements for length of use per day of the eye-tracking device, however they provided no analyses as to whether the length of use had an impact on quality-of-life.

Although, the user and non-user groups were similar at baseline for measures of quality-of-life, depression, and caregiver burden, participants had some influence during grouping therefore the two groups were not completely randomized. Further, participant eligibility for the study was not clearly stated, which would make replication of the study difficult.

Overall, this study shows suggestive evidence that implementation of an eye-tracking communication device can help improve quality-of-life in patients with ALS.

Korner et al. (2013) conducted a cohort study to measure the impact of speech therapy and/or communication devices on the quality of life and mood of patients with ALS. Thirty-eight participants with dysarthria or anarthria participated in the survey, which included three standardized questionnaires: the Beck Depression Inventory-II (BDI-II), the 36-Item Short Form Survey Instrument (SF-36) and the ALSFRS-R; as well as a self-designed questionnaire targeting individual's use and benefit from speech therapy and/or

communication device as it relates to their quality-of-life and disability.

Participants self-rated the use of a communication device as having a stronger impact on their quality of life as compared to speech therapy. No differences were found in depression (BDI) and quality of life (SF-36) between those who used a communication device and those who received speech therapy only. However, through multiple regression analysis, researchers found that bulbar impairment, as measured by ALSFRS-R scores, had an independent effect on quality of life and mood. Further analysis is required to determine the impact of a communication device while controlling for bulbar impairment.

This study included a relatively large sample size of thirty-eight participants, however, the study did not describe how participants were recruited or the eligibility criteria for participating in the study. Researchers sought to understand whether communication devices can improve or maintain quality of life, however, due to the confounding variable of bulbar impairment, their study design did not fully address their question. Further statistical tests would need to be performed to determine whether a communication device has effect.

Overall, this study shows equivocal evidence that use of a communication device has a stronger effect on quality-of-life as compared to speech therapy alone.

Maresca et al. (2019) employed a single group design study to evaluate the impact of a low-tech communication support on the quality of life of patients and caregivers. Participants in this study included ten patients with ALS and their primary caregivers. This study composed of two phases: an AAC-intervention phase and an AAC-familiarization phase, resulting in three assessment points. At the end of the AAC-intervention phase, patients were given three paper communication tables: an alphanumeric table, a preformed sentences table based on the individual, and a symbol table used for identification of pain. Outcomes were measured using Edinburgh Cognitive and Behavioural ALS Screen (ECAS), the Hamilton Anxiety Rating Scale (HRS-A), the BDI-II, the Psychosocial Impact of Assistive Devices Scale (PIADS), the SF-36, the Brief Coping Orientation to Problems Experienced (COPE), the Caregiver Burden Inventory (CBI), the Participation Inventory, and the Communication Observation Board for Functional Communication Skills.

Significant improvements were found in patient's quality-of-life (SF-36), mood (BDI and HRS-A), cognitive performance (ECAS) and coping strategies (COPE) at the end of AAC training. Caregivers showed a reduction in psychological, emotional, and social burden (CBI).

This paper provided readers with detailed inclusion criteria, recruitment process, and training process, which would make replication of the study easier. Statistical tests were performed and well reported. However, the sample size (n=10) was rather small in this study. Further, researchers excluded participants who had co-occurring cognitive impairment. Given the high prevalence of cognitive impairment among ALS patients, this may limit the generalizability of the study.

Overall, this study shows suggestive evidence that a low-tech communication support can improve quality-of-life of ALS patients in the early stages of the disease.

Calvo et al. (2008) described a single group experimental design study aimed to evaluate the impact of an eye-tracking communication device on the quality of life of individuals with ALS. Sixteen participants in the advanced stages of ALS were lent an eye tracking communication system to use for one week at their home. Speech-language pathologists trained the participants and caregivers to use the device. Outcome measures were administered before and after the device training and evaluation period. The following questionnaires were used: the MQoL, the Satisfaction with Life Scale (SWLS), the Zung Self-Rating Depression Scale (SDS), and the Self-Perceived Burden Scale (SPBS).

Improvements were found in perceived quality of life of individuals after the trial period, as measured by the MQoL and SWLS. However, no significant improvements were found in the depression and perceived burden scores, as measured by SDS and SPBS. The majority of users reported high satisfaction with the device.

A large limitation of the study was the short duration of use. Due to this being a pilot study, participants were given access to an eye-tracking communication device for only one week. This is not enough time to truly be familiarized with the device, therefore this could explain why no differences were found in depression and burden scores. Further, statistical results and values were not well described. Participant demographics were also poorly described.

Overall, this study shows equivocal evidence that an eye-tracking communication device improves the quality-of-life of individuals with ALS.

Caligari et al. (2013) conducted a survey study to measure the effect of a high-tech eye-tracking device on the quality of life of individuals with ALS. Thirty-five individuals with ALS who used an eye-tracking communication device participated in the study via questionnaires. Three questionnaires were administered: 1) The Psychosocial Impact of Assistive Devices Scales (PIADS); 2) The Quebec User Evaluation of Satisfaction with Assistive Technology (QUEST 2.0); and 3) The Individually Prioritized Problem Assessment (IPPA) scale, which was administered under three conditions: with an eye-tracking device, with an eye-transfer board requiring partner assistance, and without a device. The IPPA scale measures the impact of assistive technology on alleviating problems in daily activities.

Results from the IPPA scale indicate that the eye tracking device had a significantly larger impact on reducing communication impairments, as compared to the eye transfer board, followed by no device, which was associated with the largest number of communication impairments. Use of an eye tracking communication device also had a positive impact on quality of life compared to without the device, as measured by PIADS.

Strengths of this study include a relatively large sample size, as well as clear inclusion criteria for the study. Statistical tests administered for the IPPA scale were well reported and described. However, there was no control group in this study, therefore results cannot be compared to individuals who did not use an eye-tracking device. Further, statistical analysis compared the impact of disability between conditions, however, no statistical analysis was performed on the PIADS scale due to lack of a control group.

Overall, this study shows equivocal evidence that an eye-tracking communication device improves the quality-of-life of individuals with ALS.

Discussion

The studies analyzed in this review were in agreement that use of AAC has an effect on the quality-of-life of individuals with ALS. Further, there is suggestive to compelling evidence that early intervention with a low-tech AAC device, as opposed to late or no intervention, can improve quality of life in individuals with ALS (Londral et al., 2015; Maresca et al., 2019). There is equivocal to suggestive evidence that individuals with ALS who use an eye-tracking device, which is a more high-tech device, have improved quality-of-life (Caligari et al., 2013; Calvo et al., 2003; Hwang et al., 2014). There is equivocal evidence that use of an AAC device has stronger effect on quality-of-life of individuals with ALS, as opposed to speech therapy alone (Korner et al., 2013).

A secondary objective of this review was to evaluate the impact of type of AAC device on quality-of-life in individuals with ALS. However, the disease course of ALS largely affects the type of AAC referrals that patients receive. For this reason, only one study directly compared the difference between two AAC systems for their impact on quality-of-life. Hwang et al. (2014) compared the effect of a high-tech eye-tracking communication device and a more low-tech phonetic board that required partner-assisted scanning. The researchers found that the eye-tracking device had a greater impact on quality-of-life compared to the phonetic board. Both of these AAC supports may be used for an individual at a similar disease stage.

The studies in this review used a variety of quality-of-life instruments, therefore it is important to note that not all of these instruments measure the same concept. Instruments can measure either health-related quality-of-life or global quality-of-life. For example, the SF-36 is a health-related quality-of-life measure that focuses on physical and mental health status, whereas the MQoL is a global quality-of-life measure that includes many more factors. Quality-of-life measures can additionally be classified as being disease-specific. For example, a measure specific to the quality-of-life of patients with ALS is the ALSSQOL-R (Simmons, 2015). There is no gold standard for the type of instrument used, however given the variety of instruments used by the studies analyzed in this review, it is important to note each study varies in the type of quality of life it is measuring. For example, in the Londral et al. (2015) study, researchers found improvements in the existential well-being domain of the MQoL, and a more health-related QoL instrument would not have appreciated these changes.

As an additional variable, multiple studies included in this review measured levels of depression and caregiver burden before and after implementing AAC. However, depressive symptoms were measured using clinical depression measures (e.g. BDI, TDQ, SDS) when other depression measures might have been better appropriate to measure a depression that might be more situational and related to their diagnosis. For example, the ALS Depression Inventory (ADI-12) is a measure that has been designed and validated specifically for individuals with ALS (Atassi et al., 2011).

Several studies in this review excluded participants who had evidence of dementia or cognitive impairment with their ALS diagnosis (Londral et al., 2015; Maresca et

al., 2019). However, in individuals with ALS, 14% have been found to have co-morbid dementia and over 40% have been found to have associated cognitive impairment (Phukan et al., 2011). Given the high prevalence of cognitive issues, results from these studies cannot be generalized to individuals with ALS as a whole, only to individuals with ALS who do not show any cognitive impairment. Many patients with ALS who reject use of AAC also suffer from frontotemporal dementia (Linse et al., 2018). Therefore, these patients may not experience improved quality-of-life as a result of an AAC device.

Conclusion

Current research has found a positive relationship between use of AAC and quality-of-life. However, it should be noted that quality-of-life is a difficult outcome to measure because of the wide range of factors that may impact it. In individuals with ALS, quality-of-life is impacted by psychological symptoms, fatigue, pain, dysphagia, pseudobulbar affect, caregiver support, religion/spirituality, coping strategies, and gastronomy tools (Simmons, 2015). The use of AAC is therefore one of the many additional variables that must be included when finding ways to improve quality-of-life of ALS patients.

However, future directions for research should focus on additional variables that may impact this relationship, in order to best optimize AAC use. Information about the effect of cognitive impairment, amount of daily use, and patient preferences is lacking. This additional knowledge would help to guide policies to ensure that individuals who might benefit the most from an AAC device would receive them.

Clinical Implications

The information provided in this review can support the recommendations that clinicians make for their ALS patients in multiple ways. For example:

- Knowledge of quality-of-life outcomes can help support clinicians while initiating discussions surrounding AAC options with their patients
- Knowledge about the impact of AAC can help guide policies surrounding funding of high-tech devices, such as eye-tracking technology.
- Knowledge about the impact of early AAC intervention can guide clinicians towards suggesting some low-tech options in the early stages of the disease.
- Knowledge of the importance of timing can guide policies towards earlier referrals.

References

Atassi, N., Cook, A., Pineda, C., Yerramilli-Rao, Pulley, D., & Cudwkowitz, M. (2011). Depression in amyotrophic lateral sclerosis. *Amyotrophic Lateral Sclerosis, 12*(2), 109-112.

Beukelman, D., Fager, S., & Nordness, A. (2011). Communication support for people with ALS. *Neurology Research International, 201:* Article ID 714693.

Calvo, A., Chio, A., Castellina, E., Corno, F., Farinetti, L., Ghiglione, P., Pasian, V., & Vignola, A. (2008). Eye tracking impact on quality-of-life of ALS patients. In Miesenberger, K., Klaus, J., Zagler, W., Karshmer, A. (eds) *Computers Helping People with Special Needs.* Paper presented at International Conference on Computers for Handicapped Persons (pp. 70-77). Berlin, Heidelberg: Springer.

Caligari, M., Godi, M., Guglielmetti, S., Franchignoni, F., & Nardone, A. (2013). Eye tracking communication devices in amyotrophic lateral sclerosis: Impact on disability and quality of life. *Amyotrophic Lateral Sclerosis and Frontotemporal Dementia, 14*, 546-552.

Felgoise, S.H., Zaccheo, V., Duff, J., & Simmons, Z. (2016). Verbal communication impacts quality of life in patients with amyotrophic lateral sclerosis. *Amyotrophic Lateral Sclerosis and Frontotemporal Degeneration, 17*(3-4),179-93.

Hwang, C., Weng, H., Wang, L., Tsai, C., & Chang, H. (2014). An eye-tracking assistive device improves the quality of life for ALS patients and reduces the caregivers' burden. *Journal of Motor Behaviour, 46*(4), 233-238.

Korner, S., Siniawski, M., Kollewe, K., Jan Rath, K., Krampfl, K., Zapf, A., Dengler, R., & Petri, S. (2013). Speech therapy and communication device: Impact on quality of life and mood in patients with amyotrophic lateral sclerosis. *Amyotrophic Lateral Sclerosis and Frontotemporal Degeneration, 14*, 20-25.

Linse, K., Aust, E., Joos, M., & Hermann, A. (2018). Communication matters: pitfalls and promise of high-tech communication devices in palliative care of severely physically disabled patients with amyotrophic lateral sclerosis. *Frontiers in Neurology, 9.*

Londral, A., Pinto, A., Pinto, S., Azevedo, L., & De Carvalho, M. (2015). Quality of life in amyotrophic lateral sclerosis patients and caregivers: impact of assistive communication from early stages. *Muscle & Nerve, 52*(6), 933-941.

Maresca, G., Pranio, F., Marco, A., De Luca, R., Grazia Maggie, M., Scarcella, I., De Domenico, C., Bramanti, P., Conti Nibali, V., Portaro, S., & Salvatore Calabro, R. (2019). Augmentative and alternative communication improves quality of life in the early stages of amyotrophic lateral sclerosis. *Functional Neurology, 34*(1), 35-43.

Phukan, J., Elamin, M., Bede, P., Jordan, N., Gallagher, L., Byrne, S., Lynch, C., Pender, N., & Hardiman, O. (2011). The syndrome of cognitive impairment in amyotrophic lateral sclerosis: a population-based study. *Journal of Neurology, Neurosurgery & Psychiatry, 83*(1).

Simmons, Z. (2015). Patient-perceived outcomes and quality of life in ALS. *Neurotherapeutics, 12*(2), 394-402.