Measuring the effect assistive technology has on the quality of life of individuals with amyotrophic lateral sclerosis

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INTRODUCTION

Amyotrophic Lateral Sclerosis (ALS) is an incurable, progressive, neurological disease that affects nerves located in the brain and spinal cord.¹ As upper and lower motor nerves die, an individual loses the ability to control their muscles, experiences paralysis, and requires ventilator support for breathing assistance.² There is often a gradual onset of the disease, with early symptoms presenting as muscle atrophy, fasciculations, and slurred speech.³ Most patients with ALS live for two to five years after diagnosis; however, 10% of patients can live for ten years or longer.⁴ At this time, there is no cure for the disease. Medications including Riluzole and Radicava may be prescribed to slow the progression of the disease, with intent to prevent complications and maintain independence for as long as possible.⁵ Occupational therapists treat patients with ALS across the spectrum of care, providing evaluation and treatment as part of an interprofessional team. These services work to increase a client’s quality of life (QOL) and prevent disease complications.⁵ As medication and therapy services will not stop disease progression, it is vital to provide patients with ALS with the tools to be able to maintain the ability to communicate and interact with the environment throughout the course of the disease.

The Boston Children’s Hospital Amyotrophic Lateral Sclerosis Augmentative Communication Program (BCH ALS ACP) in Waltham, MA provides patients with ALS with assistive technology (AT) solutions to support patient communication, participation in functional daily tasks, and maintain QOL.⁶ Occupational and speech therapists work interprofessionally to evaluate and treat patients, recommending high, low, and no technology solutions to enable participation in activities of daily living and provide patient and family support throughout their disease. Recommending adaptations and AT is based on patient physical ability, cognition, current technology used, and support available. To address their goal of proactive, client-centered care, the program began to collect data on the patients seen within the program to better recognize trends in the assistive devices and the outcomes of strategies recommended.

RESEARCH QUESTION

The results of the collected data, and the desire to provide clients with assistive devices that increase levels of satisfaction with independence and communication, raised the question: In the Boston Children’s Hospital Amyotrophic Lateral Sclerosis Augmentative Communication Program, which standardized assessment will best measure the effect assistive technology has on the quality of life of patients with Amyotrophic Lateral Sclerosis?

METHODS

Needs Assessment

To begin answering the research question, a needs assessment was completed within the department to determine how to best define and measure QOL. QOL is a broad multi-dimensional concept that is defined by the positive and negative experiences in life that includes subjective evaluations.⁷ This construct is difficult to measure due to the fact that individuals define QOL differently based on their own personal experiences.⁷ When determining the values that occupational therapists and speech language pathologists considered a vital part of patient QOL, common themes arose including the importance of autonomy, satisfaction, and independence. The ability for a patient to express their needs or wants, interact with loved ones, and communicate his or her care needs, affects patient QOL. Through interaction with patients and his or her family members, it was clear QOL was affected by how well AT increases user independence and ability to communicate. Common themes arose across the questionnaire including the importance of autonomy, satisfaction, and independence. The ability for a patient to express their needs or wants, interact with loved ones, and communicate his or her care, affects patient QOL. The use of AT should enable users to increase independence and overall well-being. Data collection tools were also assessed, including methods of assessment administration, assessment length, location of administration, and when to introduce the assessment into patient care. Unanimously, the department voted to administer the assessment on multiple occasions with the same patient to track changes in QOL based on their stage of disease progression and the AT recommended. Therapists did not have a preferred assessment length, as long as it did not affect patient care or cause duress. The results of the needs assessment were beneficial to determine the required criteria of the reviewed standardized assessments to accurately measure the effect AT has on QOL.
Literature Review

A review of the literature was conducted to determine the supporting evidence of the importance of AT use within the ALS population, how AT affects QOL, and standardized assessments that address and measure QOL in both the general population and in individuals with ALS. This systematic review of QOL tools was completed to assess the beneficial effect AT has on the quality of life of individuals with ALS, and standardized assessments that can measure change. Online research databases available through the Treadwell Library, MGH Institute of Health Professions, The State University of New York at Buffalo, and Google Scholar were reviewed. Commonly searched databases included AOTA, CINAHL, EBISCO, MEDLINE, OVID, PsychINFO, and PubMed. Studies reviewed were published after 2000, peer-reviewed, and established the validity and reliability of QOL assessments. Study participants were from the general population, individuals with acquired injuries, chronic illness, or terminal diagnoses. From a total of 110 papers reviewed based on the determined research criteria, a total of 81 studies were used to address the proposed research question.

Overall, use of AT was shown to have a direct effect on the QOL of individuals with ALS. Measuring changes in QOL using a standardized assessment is a challenging process due to the varying definitions of QOL. Standardized assessments have been designed to measure different aspects QOL based on the needs of a variety of populations. The factors of QOL that are important to the general population can greatly differ than those of individuals diagnosed with chronic or terminal diseases. A large majority of QOL assessments place a high focus on physical function when assessing QOL, which limits the total score an individual with ALS can receive, and meaningful change over time. AT recommendations within the BCH ALS ACP primarily focus on communication and access methods, not mobility devices. Changes in total QOL scores would be limited for patients with ALS assessed with measures that give heavy weight to general health, physical function, and mobility items.

Outcome Measure Rating Forms

The primary focus of this project was to evaluate the clinical utility of ten standardized QOL assessments for use within the BCH ALS ACP based off of a systematic review of the literature. An adoption of the CanChild Center for Childhood Disability Research outcome measures rating form guidelines to evaluate assessment clinical utility.[8] The purpose of completing outcome measure rating forms was to provide the BCH ALS ACP with systematic, standardized reviews of ten commonly used assessments that assess QOL. Use of a template ensured comparisons could be made because each assessment was evaluated using the same methodology. Information included within the template addressed the general focus of the assessment, methods of administration, clarity of instructions, cost, and methods of item selection. Overall clinical utility of each measure was evaluated based on numerous factors including assessment psychometric properties, length of assessment administration, QOL domains measured, and weighing of items. The following QOL standardized assessments were evaluated for use within the BCH ALS ACP: WHOQOL-BREF, SF-36, PIADS, MQOL-R, SCS-R, SF-12, ALSAQ-40, ALSAQ-5, ALSSQOL-R, and the SIP/ALS-19. Completed outcome measurement rating forms for each of these assessments were provided to the site. The results from completed rating forms were used to determine which assessments would be recommended for use within the BCH ALS ACP.

DISCUSSION AND RESULTS

QOL is difficult to define because within research, healthcare, and personal opinion, there is a lack of consensus about the variables that affect QOL. Through a review of the literature and completion of outcome measurement rating forms, it became clear that each assessment measured different aspects of QOL. Determining the clinical utility of a QOL assessment within the BCH ALS ACP was completed based on analyzing how similar the assessment domains matched the department needs assessment. A majority of standardized assessments designed for the general population were not tested with the ALS population and did not measure variables that affect QOL within the ALS population. Unfortunately, a higher amount of published research was available for general population assessments with established psychometric properties. This is likely due to the researchers’ ability to recruit a larger sample size with more common diagnoses, as it is estimated that there are only 20,000 individuals diagnosed with ALS living within the United States at any given time.[9]

Based on a review of the literature, and the completed outcome measure rating forms, the WHOQOL-BREF, ALSSQOL-R, and PIADS QOL assessments have the strongest clinical utility for implementation within the BCH ALS ACP. Recommendations are described below for future implementation of the chosen assessment to answer the proposed research question.

Assessment Recommendations
Although the WHOQOL-BREF is a QOL assessment designed for use with the general population, it has been widely used and has well-established psychometric properties. The World Health Organization defines QOL as "an individual's perception of their position in life, in the context of the culture and value systems in which they live and in relation to their goals, expectations, standards and concerns, is a broad ranging concept affected in a complex way by the person's physical health, psychological state, personal beliefs, social relationships and their relationship to salient features of their environment".[10] This definition best aligns with the themes discovered in the completed department needs assessment. There are numerous methods for assessment administration, as it can be administered through staff interview within the clinical setting or completed by a patient and his or her caregiver at home. Although it is 26 items and this is longer than the desired length addressed within the needs assessment, the WHOQOL-BREF completion is estimated to be 10 minutes. The assessment is available for free use with permission from the World Health Organization, and item phrasing has a positive connotation. This reduces the amount of distress a patient may feel as they complete this assessment.

The use of an ALS-specific questionnaire is recommended for the BCH ALS ACP as it has established norms and psychometric properties for patients diagnosed with ALS. The ALSSQOL-R assessment contains 46 scored items, which is also larger than the amount of items individuals in the department desired for assessment length; however, it is estimated to take 15 minutes to complete. This assessment is also available for free with permission from the Pennsylvania State University College of Medicine. When compared to the ALSAQ-5 and ALSAQ-40, the increased focus on psychological, existential, and spiritual domains measure more person-centered aspects of QOL and reduces the focus on physical function. Many of the assessments that are ALS-specific place too high of an emphasis on the domain of mobility and physical ability. Items have negative overtones, with a focus on activities of living the individual is unable to complete, negative emotions and feeling of being a burden, and asking if life is worth living. The ALSQOL-R does not include questions that may distress the client, and includes items about the intrinsic and extrinsic factors that affect QOL. A section on religiosity is included within the measure, although these items do not need to be scored if the site wants to measure QOL based on the themes established in the needs assessment and the conceptual model.

Last, the PIADS has strong clinical utility when assessing AT. If BCH ALS ACP determines they want to further specify the research question to evaluate the effect on specific communication devices or access methods on the QOL of ALS patients, this is an effective tool. Forms included in the assessment can be used with individuals using AAC devices and environmental control units. A script has also been included for administration for patients with a terminal illness, which is applicable to patients seen with ALS. Due to the complexity of the assessment administration and scoring, it is recommended that a staff member review the materials prior to using the assessment with a client.

Future Research

To trial the clinical utility of a recommended QOL assessment, a pilot study should be designed and conducted within the BCH ALS ACP using the WHOQOL-BREF, ALSSQOL-R, or PIADS assessment. A pilot study assesses the clinical utility of a specific assessment and identifies the resources needed to complete a planned study, possible barriers to collecting data, and logistical problems in assessment.[11] The BCH ALS ACP should modify the proposed research question to include the chosen assessment and adopt a conceptual framework to ensure the data collected are measuring the correct variables. Next, the site should establish methods of collecting preliminary data. The assessment should be administered by the same clinician, or with an identical script if completed within the clinical setting. If numerous staff members will be administering the assessment, completing training with the assessment and reviewing administration procedures in a group will address internal validity of the pilot study. This will reduce the risk of contaminating the data collected and create a more uniform method of administering the chosen assessment. Collected data should include the amount of time it took to administer the assessment, administration location, the client’s total assessment score, and any barriers the patient faced when completing the questionnaire. Data currently collected at BCH ALS ACP that will be beneficial to this study includes patient date of diagnosis, a history of AT recommended by clinician, and patient ALSFRS-R score. The BCH ALS ACP will establish a time period and sample size of patients to be included in the study. Although it can be time consuming to design and conduct a pilot study, it provides necessary information on the feasibility of use of a chosen assessment. Further studies can be completed using the other recommended assessments as needed.

CONCLUSION AND IMPLICATIONS FOR PRACTICE

AT is an essential resource to individuals with ALS. It enables users to maximize their independence, express their needs and wants, communicate with others, and participate in valued occupations. The recommendation of AT can include low, high, or no tech options, but must fit the client. Within the BCH ALS ACP, clinicians provide
patients with ALS with custom AT that increase ability and overall QOL. Integration of a standardized QOL assessment allows clinicians to make informed, patient-centered recommendations, and measure the distinct value of the services provided.

The results of this research have the following implications for practice within the BCH ALS ACP and future research for measuring the QOL of individuals with ALS:

- QOL is a multi-dimensional concept that is affected by internal and external influences. Selection of a shared definition of QOL for research purposes is difficult due to unique, personal experiences and values that affect how QOL is individually perceived.
- The recommendation of AT by occupational and speech therapists improves the QOL for patients diagnosed with ALS by providing the patient increased communication and independence.
- There is a need for further development of ALS-specific QOL assessments with decreased focus on physical ability. Further, established assessments should be tested with the ALS population to provide additional methods of assessing QOL within the clinical setting.
- In the BCH ALS ACP, selection and implementation of a QOL measure will benefit clients by producing measurable data to guide the recommendation of AT.

REFERENCES


