Systematic Review of the Effectiveness of Occupational Therapy–Related Interventions for People With Amyotrophic Lateral Sclerosis

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MeSH TERMS
- amyotrophic lateral sclerosis
- exercise therapy
- occupational therapy
- palliative care
- patient care team
- self-help devices
- treatment outcome

We describe the results of a systematic review of the literature on occupational therapy–related interventions for people with amyotrophic lateral sclerosis (ALS). The review included 14 studies. We found limited to moderate evidence that people involved in multidisciplinary programs have longer survival than those in general care and limited evidence that those in multidisciplinary programs have a higher percentage of appropriate assistive devices and higher quality of life in social functioning and mental health. Limited evidence indicates that people with ALS are satisfied with the comfort and ease of use of their power wheelchairs (PWCs). In addition, limited evidence is available that PWCs allow people to have increased interaction in the community. Evidence also is limited that some assistive devices are more helpful than others. Moderate evidence indicates that a home exercise program of daily stretching and resistance exercise results in improved function. The implications for practice, education, and research are discussed.

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The objectives of this review were to identify the research literature and critically appraise and synthesize the applicable findings to address the following focused question: What is the evidence for the effectiveness of interventions within the scope of occupational therapy practice for people with amyotrophic lateral sclerosis (ALS)?

Background and Statement of the Problem

ALS is a late-onset, rapidly deteriorating neurological disorder characterized by the selective death of motor neurons in the brain and spinal cord that innervate skeletal muscles. ALS is the most common motor neuron disease (McGuire & Nelson, 2006), with an incidence of 2 per 100,000 people (Amyotrophic Lateral Sclerosis Association, 2010). Clinical symptoms of ALS are progressive weakness, muscle wasting, spasticity, and fatigue (Jackson & Bryan, 1998; Williams & Windebank, 1991). Findings from a national database of people with ALS over a 2-yr period indicated that 36% self-reported being primarily independent and 20% being primarily dependent in performing activities of daily living (ADLs; Miller et al., 2000).

People with ALS have a median survival of 3 yr, and the immediate cause of death is respiratory failure, pneumonia, or cardiac arrhythmias (McGuire & Nelson, 2006). Approximately 10% of people with ALS survive 10 yr or more (Rowland & Shneider, 2001). Evidence indicates that progression in ALS is not linear, but curvilinear, with the early and late phases of the disease showing the most rapid rate of decline (Gordon et al., 2010). Although clinically significant dementia is not commonly seen in people with ALS, up to 50% have deficits in

frontal executive function (Lomen-Hoerth et al., 2003). People with ALS and frontotemporal lobar dementia have shorter survival and are less compliant with recommended treatments than those with normal executive and behavioral function (Olney et al., 2005).

The cause of ALS is unknown. According to Simmons (2005), although no medications are available to halt or reverse the progression of ALS, several management strategies, including occupational therapy, can optimize quality of life. Findings from the national database indicate that 30% of people with ALS reported participating in occupational therapy (Miller et al., 2000). Occupational therapy practitioners work with people throughout the course of ALS to develop client-centered and occupation-based interventions in all areas of occupation, including ADLs, instrumental ADLs (IADLs), work, education, leisure, social participation, and rest and sleep (American Occupational Therapy Association, 2008). Practitioners may provide services to people with ALS and their caregivers in any treatment setting in which occupational therapy services are provided, including acute care hospitals, skilled nursing facilities, and multidisciplinary ALS clinics or through home health or hospice services. In addition, occupational therapy practitioners use their expertise when working with people with ALS and their caregivers to address end-of-life issues such as positioning for comfort, prevention of contractures, and family and caregiver training.

It is critical that practitioners use the best available research to guide current practice when providing services to people with ALS. This systematic review provides evidence-based information on interventions that occupational therapy practitioners can use in work with adults with ALS.

Method

This systematic review is an update of an earlier systematic review that covered the period from 1985 to 2002. It includes articles from the earlier review as well as articles that were found through database searches on articles published from 2003 through March 2011. In addition, we reviewed bibliographies of selected articles for potentially relevant articles, and other articles recommended by experts in the field were included in this review. Search terms for the review included *activities, activities of daily living, assistive devices, assistive equipment, caregiver support, cognition, disease management, education, emotional regulation, exercise, extended care, fall prevention, fatigue, health maintenance, instrumental activities of daily living, intervention, leisure, mobility, mobility equipment, neurorehabilitation, occupational therapy, quality of life, rehabilitation, self-management, sleep, social engagement, walkers, wheelchairs,* and *work.*

Articles selected for the review included studies in which the focus was on people with ALS. In addition, the interventions studied were within the scope of practice of occupational therapy. Detailed information about the methodology and a complete list of search terms used in this review can be found in the article "Method for the Systematic Reviews on Occupational Therapy and Neurodegenerative Diseases" in this issue (Arbesman, Lieberman, & Berlanstein, 2014).

Results

We reviewed 872 abstracts for the updated search, and 14 articles were included in the final results of the review. Three articles were Level I randomized controlled trials (RCTs), systematic reviews, or meta-analyses. Three articles were Level II nonrandomized controlled trials, 4 articles were Level III cross-sectional surveys, 3 articles were Level IV single-subject design studies, and 1 article was a Level V case study. Supplemental Table 1 (available online at http://ajot.aotapress.net; navigate to this article, and click on "Supplemental Materials") summarizes all the studies included in the systematic review and provides the objectives and design, a description of the interventions and outcome measures, and summaries of the results and limitations of each study. Five programmatic themes resulted from the review of the literature: exercise, assistive devices and wheelchairs, multidisciplinary programs, palliative care, and preparatory methods.

Exercise

Moderate evidence from a Level I study indicates that participants in a home exercise program of daily stretching and resistance exercise improved their scores on the Amyotrophic Lateral Sclerosis Functional Rating Scale (ALSFRS; ALS CNTF Treatment Study (ACTS) Phase I–II Study Group, 1996) and the Medical Outcomes Survey 36-Item Short Form Survey (Ware, Snow, Kosinski, & Gandek, 1993) physical functional scales with no adverse effects (Dal Bello Haas et al., 2007). A Level I study provided limited evidence that people in an individual exercise program carried out twice daily to improve muscle endurance had better functional ability and lower spasticity than those in a no-exercise condition. However, both intervention and control groups deteriorated over time (Drory, Goltsman, Reznik, Mosek, & Korczyn, 2001). A Level II nonrandomized controlled trial (Aksu, Karaduman, Yakut, & Tan, 2002) provided limited evidence that a supervised exercise program maintains a person's functional capacity better than a home exercise program. A Level V study found that a participant had increased energy and

required less assistance for transfers after participating in aquatic therapy (Johnson, 1988).

Assistive Devices and Wheelchairs

Limited evidence was found from 2 Level III studies regarding the use of and satisfaction with wheelchairs. In a survey of power wheelchair (PWC) users, 77% of respondents were satisfied with the overall comfort of their PWC, and 72% were satisfied with the ease of use (Ward et al., 2010). Features frequently used with the PWC included tilt, recline, power elevating leg rests, ability to run power features through a joystick with upgraded electronics, air or gel cushion, height-adjustable armrests (flat, gel, or contoured), soft headrest, and seatbelts. Limited evidence from a survey of power and manual wheelchair users indicates that wheelchairs, particularly PWCs, allow for increased interaction in the community (Trail, Nelson, Van, Appel, & Lai, 2001). Participants using PWCs reported more satisfaction with their ability to participate in activities than those using manual wheelchairs; participants reported greater satisfaction with manual wheelchairs with respect to portability. No difference was identified in comfort or ease of maneuvering. Participants reported that head, neck, trunk, and extremity supports were helpful but that sling backs and sling seats were not.

One Level III study provided limited evidence that some assistive devices are more helpful than others (Gruis, Wren, & Huggins, 2011). Participants reported that the following devices were used often or always and had high usefulness and satisfaction: elevated toilet seat, rails by toilet, shower seat, shower bars, slip-on shoes, ankle brace, and transfer board. The following devices were used infrequently but with high satisfaction: sound- or voice-activated environmental controls and communication boards. Participants reported that the following were not useful and had low satisfaction: buttonhooks, dressing stick, and long-handled reaching tools.

Limited evidence from a Level I systematic review (Foley, Timonen, & Hardiman, 2012) indicates that the use of telemedicine may be associated with high levels of satisfaction. Even though participants were satisfied with telemedicine, they also reported that face-to-face contact was more highly valued for discussion of psychological and emotional concerns.

One Level IV study examined the effectiveness of a computer program for writing messages and choosing songs and videos through a virtual keyboard and microswitch (Lancioni et al., 2012). After training with the computer program, the participant wrote two messages per 20-min session and chose songs and videos independently.

Multidisciplinary Programs

Limited to moderate evidence from a Level II study indicated that people with ALS who participate in multidisciplinary programs have approximately 30% longer survival than those in general care (Traynor, Alexander, Corr, Frost, & Hardiman, 2003). Multidisciplinary programs may include disciplines such as neurology, nursing, occupational therapy, physical therapy, speech pathology, pulmonology, nutrition, psychology, and social work. Limited evidence was provided by a Level III study that people in multidisciplinary programs have a higher percentage of appropriate assistive devices and higher quality of life in social functioning and mental health (Van den Berg et al., 2005). No differences were found between groups, however, for physical functioning or quality of life for caregivers.

Palliative Care

A Level II study provided limited evidence that occupational therapy was part of the care team for people with ALS even immediately before death (Albert, Murphy, Del Bene, & Rowland, 1999). Limited evidence from a Level I systematic review (Foley et al., 2012) indicated that maintaining control in making decisions about end-of-life care was important for people with ALS. Although some participants preferred independent decision making, shared decision making with family, service providers, or both was also a preference.

Preparatory Methods

Minimal evidence was provided by a Level IV study that electrical stimulation improves bilateral hand function and knee extension over a period of 3 mo (Handa et al., 1995).

Discussion and Implications for Practice, Education, and Research

In reviewing the available research for occupational therapy practice with people with ALS, several themes emerged involving the role of multidisciplinary clinics, the significance of equipment choices, the importance of appropriate exercise, and the role of occupational therapy practitioners on the palliative care team. These components of occupational therapy practice are important for this population.

The results of the review indicate that multidisciplinary programs or clinics increase quality of life and length of survival of people with ALS. Occupational therapy practitioners bring a unique client-centered and occupation-based perspective to the multidisciplinary team. It is important to focus on helping the person with ALS participate in the activities and roles that are most

important to that individual and his or her family (Ward & Brooks, 2012). According to Phukan and Hardiman (2009), this client-centered approach focuses on optimizing the environment of the client and may include home modification and safety awareness, appropriate adaptive equipment, information on energy conservation, and modification of meaningful activities such as eating and driving.

The results also indicate that an individualized resistance exercise program may help people with ALS prolong functional independence. Although muscle strength cannot be regained in muscles weakened by ALS, some evidence has indicated that a mild resistance exercise program may help people with ALS maintain strength in the unaffected muscles and preserve functional independence for ADLs. In addition, a personalized range of motion program for weakened muscles is critical to preventing joint contractures.

In the area of assistive devices, bathroom durable medical equipment—specifically, grab bars, raised toilet seats, and shower seats—were rated among the most useful equipment in a self-reported satisfaction survey. People with ALS seem most interested in equipment that increases safety and independence for bathing and toileting. Although not included as a choice in the reviewed research surveys, many people with ALS find that a bidet increases dignity and independence in toileting.

PWC features must be considered carefully to ensure the greatest comfort and ease of use for the client and the greatest ability to be adapted to the client's changing needs throughout the disease process. People with ALS have reported that a PWC should have the ability to tilt as well as to recline. In fact, some people find that as their mobility declines and getting in and out of bed becomes more difficult, they are more comfortable sleeping in their PWC. Power elevating leg rests are also an important feature and can be very helpful for clients with dependent edema in the lower extremities. Armrests should be contoured and adjustable to provide comfort, particularly for clients whose arms are no longer functional.

Intervention for clients with ALS offers occupational therapy practitioners unique opportunities to expand their practice and embrace their role in both hospice and palliative care. According to Mitsumoto et al. (2005), who reported on the results of an ALS peer work group, the best time to begin palliative care is at the time of diagnosis. The work group indicated six triggers for initiating discussion about end-of-life issues: request by the family; presence of severe psychological, social, and or spiritual distress; levels of pain requiring high dosages of analgesic medications; dysphagia requiring a feeding tube; dyspnea or symptoms of hyperventilation, with a forced vital capacity of 50% or less; and loss of function in two body regions, such as arms, legs, and tongue.

Positioning and comfort are important palliative care needs of people with ALS. Occupational therapy practitioners can play a vital role in helping the client find positions of comfort in bed or in a wheelchair. Practitioners can also provide splinting to address hand contractures. When people with ALS enter hospice care, practitioners can provide education to hospice aides and family members on a passive range of motion program for the client. In addition, practitioners can educate hospice workers to focus on proper positioning so the person can participate in meaningful activities (e.g., visiting friends, carrying out ADLs and IADLs, using equipment to control the environment).

Research on the effectiveness of all intervention types for people with ALS is limited. To date, intervention effectiveness research has not been conducted at all on two topics that are important to examine in future occupational therapy research studies. The first topic is fatigue, a common complaint with complex qualities in people with ALS (Hallum, 2001). For example, research has shown that muscle fatigue and muscle weakness do not correlate with each other in people with ALS (Sanjak et al., 2001). Jackson and Bryan (1998) suggested that energy conservation may be valuable in reducing fatigue and improving performance in people with ALS.

Although energy conservation techniques have been evaluated in people with multiple sclerosis (Hugos et al., 2010; Mathiowetz, Finlayson, Matuska, Chen, & Luo, 2005; Mathiowetz, Matuska, Finlayson, Luo, & Chen, 2007; Sauter, Zebenholzer, Hisakawa, Zeitlhofer, & Vass, 2008; Vanage, Gilbertson, & Mathiowetz, 2003), Lewis and Rushanan (2007) indicated that it is difficult to generalize from this research to people with ALS. Lewis and Rushanan suggested, however, that adaptations such as sitting versus standing to complete activities, taking rest breaks, and using adaptive tools may improve function in people with ALS. These techniques can be applied in all environments in which people with ALS participate, such as home, workplace, and community.

The second area for which research is lacking at this time is the impact of occupational therapy services on quality of life for people with ALS and their caregivers. Most studies use outcome measures that assess more objective aspects of function, such as the ALSFRS–R (Cedarbaum et al., 1999), which examines daily activities and the progression of disability. Foley (2004) stressed the need for occupational therapy practitioners to incorporate quality-of-life measures into their practice to understand people with ALS from a client-centered perspective. Using quality-of-life measures in occupational

therapy research with this population will provide a more accurate picture of outcomes for people with ALS and their caregivers.

This client population provides unique educational opportunities for occupational therapy students and practitioners, and the evidence provided here should be incorporated into academic programs. Occupational therapy students and practitioners must understand positioning and pressure ulcer prevention and how to incorporate these concepts into ADL routines. In addition, practitioners and students should have a thorough understanding of splinting, splinting concepts, and the value and drawbacks of using various splints with people with ALS. Occupational therapy practitioners also need to be confident in their skills for helping clients functionally navigate the psychosocial issues surrounding end-of-life decisions, such as choosing a ventilator or considering the placement of a feeding tube, and students need to be made aware of these challenges during their training.

Strengths

The systematic review presented in this article has several strengths. The methodology included a large time frame (the initial review included research literature published since 1985) and incorporated several bibliographic databases, thus ensuring that relevant literature was captured. In addition, the articles in the review included a wide range of interventions.

Limitations

Some of the articles included in this systematic review had limitations, such as small sample sizes and high dropout rates. In addition, few Level I RCTs were included in the review. These limitations may reflect the challenges of conducting research with people with ALS, a disease for which the pool of potential participants is limited and few participants are in the same stage of disease progression. In addition, the swift decline in medical or health status because of the disease course may be a major cause of the high dropout rates reported in the trials. Given these challenges, it may be realistic to expect that surveys and study designs that can be conducted over short periods with fewer participants will be the norm in rehabilitation research with this population.

Implications for Occupational Therapy Practice

In summary, the findings of this review have the following implications for occupational therapy practice with people with ALS:

- Limited to moderate evidence is available that participants in multidisciplinary programs have longer survival rates than those in general care, and limited evidence indicates that they have a higher percentage of appropriate assistive devices and higher quality of life in social functioning and mental health. Occupational therapy practitioners are uniquely qualified to help people with ALS make wise choices of equipment and environmental modification with consideration of the disease progression. A multidisciplinary clinic setting allows occupational therapy practitioners to work with the team to assist the client and family in setting goals and making choices that are client centered and allow engagement in desired roles and activities.
- Limited evidence indicates that some assistive devices are more helpful than others. Bathroom equipment is especially helpful for people with ALS. The most helpful equipment includes raised toilet seat, shower seat or tub bench, grab bars, and bidet.
- Evidence is limited that people with ALS are satisfied with the comfort and ease of use of their PWCs. In addition, evidence is limited that PWCs allow the person to participate in community-based activities. PWC features must be selected with the progression of the disease in mind. Some important features for this client population include headrest, padded full-length armrests, recline, tilt in space, elevating leg rests, caregiver controls, and the ability to modify wheelchair controls.
- Moderate evidence indicates that a home exercise program of daily stretching and resistance exercise results in improved function. Daily range of motion—particularly for weakened muscles—is important for maintaining joint mobility and preventing contractures.
- Although the evidence is limited, research indicates that occupational therapy is part of the palliative care team for people with ALS, even immediately before death. The course of ALS provides distinct opportunities for occupational therapy practitioners to be involved in the nontraditional practice areas of palliative and hospice care. Practitioners can address positioning and seating needs for people with ALS and train family members and hospice aides to provide an individualized range of motion exercise program. ▲

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