

**Examining the impact of rehabilitation interventions on quality of life (QoL) in people with  
amyotrophic lateral sclerosis (ALS)**

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amyotrophic lateral sclerosis (ALS)**

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## **Contributions**

The thesis consists of two separate papers, each formatted according to the requirements of the intended Journals to which they will be submitted. Although the papers are coauthored by more than one person, Ammarah Y. Soofi was responsible for all aspects of the two studies which include, database literature searches, analyses and writing of the manuscript. The co-authors on the papers had various contributions ranging from support with methodology, database searching, analyses, and reviewing the manuscripts.

## Abstract

The purpose of this thesis is to examine how rehabilitation interventions, specifically physiotherapy (PT), occupational therapy (OT), and speech and language pathology (SLP) or a combination of these interventions affect quality of life (QoL) in people with amyotrophic lateral sclerosis (ALS).

The purpose of the first study (Chapter 2) was to synthesize qualitative research through a qualitative meta-synthesis on the potential of rehabilitation interventions to maintain and/or improve QoL from the perspective of people with ALS. The literature search for this study was conducted using the SPIDER strategy and five articles were included. Four themes emerged: 1) the concept of control; 2) adapting interventions to disease stage; 3) struggles with interventions; and 4) barriers between healthcare providers and patients. The evidence suggests that from the perspective of people with ALS, PT, OT, and SLP interventions, or a combination of these interventions have the potential to be beneficial in the management of people with ALS and to optimize QoL.

The systematic review (Chapter 3) aimed to determine the effectiveness of rehabilitation interventions, in particular PT, OT and SLP interventions or a combination of these interventions, on QoL in people with ALS. The PICO search strategy was used and six studies were included: three RCTs, two cohort studies, and one cross-sectional study. A narrative synthesis of interventions was conducted as the included studies were not sufficiently similar thus data extracted were not adequate for conducting meta-analyses. Need to briefly discuss interventions, which outcomes you included and inconsistency in results across studies. Due to the limited evidence, it was difficult to determine the exact effects of the interventions from each rehabilitation field to affect QoL for people with ALS. The evidence suggests that more research

is required; currently therapists need to rely on their clinical expertise, expert opinions, and theoretical models to select the most effective interventions to sustain or improve QoL in people with ALS. Future research needs to take into consideration the needs of people with ALS to evaluate the impact of rehabilitation interventions on QoL.

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This thesis is dedicated to my mother (K.Y. Soofi) who could not be here today, but her death to ALS was and still is my motivation to increase my knowledge and understanding of the impact rehabilitation has for individuals with ALS to improve their quality of life while there is still no available cure. This thesis allowed me to gain a detailed understanding of the disease, the roles of healthcare professionals to help manage ALS, and the impact multidisciplinary care has in this population. Additionally, I was able to explore and relate to the experiences and struggles faced by these individuals and their family caregivers. I would also like to say a sincere thank you to my father, sisters, significant other, and friends, who continue to believe in me and have endless support for me in everything I do. Their encouragement and enthusiasm was always evident and they always ensured that I had plenty of time to work, while ensuring a balance in my life.

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## **Chapter One: Introduction**

### 1 Background

#### 1.1 Amyotrophic Lateral Sclerosis

Amyotrophic lateral sclerosis (ALS) is a progressive, terminal disease that causes an extensive and ever-challenging range of needs across the disease stages. The condition is characterized by the loss of motor neurons in the cortex, brain stem, and spinal cord, and is manifested by upper and lower motor neuron signs and symptoms affecting bulbar, limb, and respiratory muscles (Ng, Khan, & Mathers, 2009). Death usually results from respiratory failure and follows, on average, two to four years after diagnosis, yet some people survive for a decade or more (ALS Society of Canada, 2012; Gordon, 2013). ALS is a relatively rare neurodegenerative disease of the motor system in adults, with a reported population incidence in Canada of 2 per every 100,000 people per year. Between 2,500 and 3,000 Canadians currently live with this fatal disease (ALS Society of Canada, 2012; Habib, Mohammed, Vaculik, Wong, & Rotella, 2015).

ALS is a devastating disease with an unknown etiology and varying symptoms including weakness, spasticity, limited mobility and difficulty carrying out daily activities, communication deficits, respiratory problems, fatigue and sleep disorders, pain, and psychosocial distress (Ng et al., 2009). According to the International Classification of Functioning, Disability and Health (ICF) framework (World Health Organization, 2001), ALS-related impairments limit activities and participation in a variety of areas e.g., driving, employment, interactions with family, and social integration. While ALS is irreversible and incurable, rehabilitation is designed to help

patients and their caregiving family members adapt, manage and cope with the effects of the disease to maintain or optimize their quality of life (Mitsumoto & Del Bene, 2000).

## 1.2 Quality of Life

Quality of life (QoL) is an expansive multidimensional concept that typically includes subjective evaluations of both positive and negative aspects of life (Potter, Cantarero, & Wood., 2012). Due to this, measuring QoL becomes challenging as the term “quality of life” has its own meaning among individuals and nearly every academic discipline defines it differently (World Health Organization., 2005). Although health is one of the important domains of overall QoL, other key domains include employment, social activities, and independence. (Ng & Tallman, 2011). Additionally, aspects of one’s culture, values, and spirituality are also recognized as contributors of overall QoL (Potter et al., 2012). Thus, addressing all such factors add to the complexity of defining and measuring QoL. Nevertheless, researchers have developed several useful techniques that have helped conceptualize and measure these various domains and how they are related in one’s meaning of QoL. Therefore, QoL as assessed by patients with ALS, is not limited to changes in their abilities associated with the condition, but rather can be considered as “the extent to which our hopes and ambitions are matched by experience” (Mitsumoto & Del Bene, 2000, p.329).

QoL describes a person’s satisfaction with the way his/her life is progressing and possibly with the care measures diagnosis with a disease such as ALS (Mitsumoto & Del Bene, 2002). QoL describes complex aspects of a person’s physical and mental health but it is not limited to measureable indicators such as life expectancy or biomedical data, and therefore should include a subjective evaluation of life through indicators such as health status and

external life situations etc. (Mendlowicz & Stein, 2000). Although there are various factors that can affect a person's outlook on his/her life post-diagnosis, this study focuses specifically on those related to the care process.

The rapid progression and severity of muscle weakness in ALS greatly affects a person's feelings of hope and self-worth (Mitsumoto & Del Bene, 2000). Participation in activities (e.g., walking a dog), fulfillment of life goals (e.g., climbing a mountain), and continuing employment are also affected by the disease with respect to social interactions, participation in leisure activities and sexual relationships are affected as well (Mitsumoto & Del Bene, 2000). Due to disease-related symptoms, changes in self-esteem, loss of independence, and psychosocial stress all play important roles in the perception of QoL for the individual living with ALS throughout the disease process (Francis, Bach, & DeLisa, 1999).

Some studies have revealed that QoL in people with ALS is not necessarily associated with measures of strength and function, nor does QoL inevitably decrease due to the progressive nature of ALS (Lewis & Rushanan, 2007; Simmons, 2005). Consequently, the concept of QoL broadly encompasses how individuals measure the 'goodness' of multiple aspects of their life (Diener, Suh, Lucas, & Smith, 1999). These self-measures of 'goodness' include one's emotional reactions to life occurrences, disposition, sense of life fulfillment and satisfaction, and satisfaction with work and personal relationships (Diener et al., 1999).

Since QoL is being defined in this thesis as the matching of experiences with hopes and ambitions, it is important to consider the effect of the disease on one's expectations. Patients with any condition, including ALS, who do not have high hopes of recovering may not be eager to undertake any treatment as they have low expectations of its usefulness (Diener et al.,



1999). This in turn decreases QoL as disability has been linked with overall QoL and physical health status (Motl & Snook, 2008). With the multiple physical, social, and emotional strains impacting individuals diagnosed with ALS, the goal of healthcare professionals must be to continually improve QoL through medical, rehabilitative and psychosocial interventions, thus working to reduce the gap between the patients' hopes and their actual experiences.

### 1.3 Rehabilitation in ALS

Rehabilitation is a process aimed at enabling people with disabilities to attain and preserve their ideal level of physical, intellectual, psychological and social function by providing the tools necessary to manage independence and control of their life (World Health Organization, 2005). In people with ALS, rehabilitation aims to assist with independence and can be used to help individuals reach their fullest potential, regardless of the presence of the disabling disease (Ng et al., 2009; ALS Association, 2015). During the early ALS stages, rehabilitation aims to prolong independence with carrying out daily activities (Ng et al., 2009) and as the disease progresses, rehabilitation specialists can modify their care efforts to include the education of families and caregivers on techniques involved with carrying out exercises and safe transfer of patients (Simmons, 2005). Overall, the main purpose of rehabilitation for people with ALS is to reduce symptoms and limitations associated with inactivity and to promote participation through wide-ranging interventions, which incorporate personal and environmental factors (Ng et al., 2009).

The variety of symptoms in people with ALS requires careful assessment and management. Therefore, an ideal multidisciplinary rehabilitation team comprises of a group of clinical professionals with full understanding or knowledge of ALS (Hardiman, 2007). In their

systematic review, Ng and colleagues (2009) provide support for the role of multidisciplinary care for individuals with ALS, although their reviewed focused on care that included rehabilitation, medical, pharmaceutical, dietary supports etc., without isolating rehabilitation. The authors suggested that rehabilitation could lead to improvements in QoL without increasing healthcare costs. Additionally, hospitalization and disability were reduced (Ng et al., 2009). Therefore, if a person were to spend less time as a patient in a hospital setting and participated in rehabilitation in an encouraging environment, QoL can potentially be increased.

Over the last few years, there is growing evidence supporting the importance of multidisciplinary care for the management of ALS. Other studies demonstrate that multidisciplinary care for patients with ALS improved QoL, reduced hospitalizations and length of hospital stays, increased longevity and decreased disability (Chio, Bottacchi, Buffa, Mutani, & Mora, 2006; Ng et al., 2009; Traynor, Alexander, Corr, Frost, & Hardiman, 2003). Although the health benefits are well documented, there are individuals who cannot attend specialist multidisciplinary clinics due to physical and geographical limitations; thus, focused single discipline service delivery in the community environment is necessary (O’Callaghan, Murray, & Vance, 2014). In people with ALS, rehabilitation goals should actively involve the input of the person with ALS and his/her primary caregivers, focus on activity and participation, and consider short and long term objectives, adjusted as the disease progresses (ALS Society of Canada, 2012). This thesis specifically looks at interventions designed or delivered by physiotherapy, occupational therapy and speech and language therapy professionals or a combination thereof. The interventions from these disciplines of rehabilitation can overlap, Table 1 provides an overview of the goals and types of interventions provided by the fields of physiotherapy, occupational therapy and speech and language pathology. In addition,

multidisciplinary interventions that incorporate interventions from combinations of the above mentioned professions are discussed.

### 1.3.1 Physiotherapy

Physiotherapy plays an important role in the treatment of injury and disease as well as health promotion for people with ALS. Physiotherapists combine their knowledge of the human body and specialized clinical hands-on skills to evaluate and treat patients with symptoms or diagnoses of illness, injury or disability (Canadian Physiotherapy Association, 2012). Over the course of ALS, physiotherapists play a significant role in supporting these individuals in order for them “to maximize functional independence, reduce the effects of disability, and enhance their QoL” (Dal Bello-Haas & Montes, 2009, p. 99).

Dal Bello-Hass (2002) and O’Callaghan (2014) describe each stage of ALS and the respective physiotherapy interventions that are potentially effective. In the early stage, there may be weakness in certain limb muscles as well as other muscles of the upper and lower body that causes minor limitations in terms of performance, endurance and the ability to carry out tasks (Dal Bello-Haas, 2014; O’Callaghan et al., 2014). In the middle stage, the individual may still be ambulatory; however, it is likely that there is severe weakness in specific muscle groups for example, in upper limbs (biceps, triceps and deltoids), and respiratory insufficiency may start to develop (O’Callaghan et al., 2014). Therefore, physiotherapists are encouraged to teach the individual techniques of energy conservation, supporting lung expansion, and secretion management through airway clearance strategies (O’Callaghan et al., 2014; Dal Bello-Haas & Montes, 2009). In the late stage, progressive weakness and deterioration in mobility, dexterity and endurance occurs (Dal Bello-Haas, 2014; O’Callaghan et al., 2014). As a result of this

deterioration, individuals in this stage of ALS are generally wheelchair dependent and/or may have some respiratory compromise (Dal Bello-Haas, 2014).

To preserve and/or achieve the above mentioned goals, common recommendations include therapeutic exercise and prescription of assistive and/or adaptive devices (Dal Bello-Hass & Montes, 2009). Therapeutic exercise is a physical activity program designed and prescribed for specific therapeutic goals (e.g., maintain mobility and muscle strength, flexibility, reduce pain etc. (Blair, Sallis, & Archer, 2012; Dal Bello-Haas & Montes, 2009)). The purpose of therapeutic exercise is to work towards the continuance of musculoskeletal function through the re-education of neurological pathways, gait training and therapeutic activities (Blair, et al, 2012). In individuals with ALS, the challenge is identifying when participation in exercise therapy is beneficial versus harmful. Historically, it was believed that muscle over-exertion may accelerate disease progression (Brouwer, Padberg, van der Ploeg, Ruys, & Brand, 1992; Fowler, 1984; Fowler & Taylor, 1982; Johnson & Braddom, 1971). However, in recent years research has found that exercise has beneficial effects related to muscle strength, aerobic capacity, pulmonary function, and QoL (Dal Bello-Haas & Florence, 2013; Drory, Goltsman, Reznik, Mosek, & Korczyn, 2001; Sanjak, Bravver, Bockenek, Norton, & Brooks, 2010).

In addition to exercise therapy, prescribing assistive and/or adaptive aids are also recommended by PTs to improve mobility and for support. These aids include canes, walkers, crutches, orthotics, mechanical lifts and beds, and braces (Dal Bello-Hass & Montes, 2009). These interventions are introduced to help people with ALS adapt to the effects of degeneration that are to come. Therefore, PT interventions frequently change across the ALS disease continuum (Dal Bello-Haas, 2002; O'Callaghan et al., 2014).

O’Callaghan et al. (2014) recommend that physiotherapists educate caregivers on handling the patient during assisted exercises, daily mobility, and safe transfers to reduce the risk of caregiver and patient injury. As previously mentioned, part of the ALS disease process involves changes in physical function or balance. Although physiotherapy interventions cannot change the disease itself, the interventions can assist patients with maximizing their balance and safety, minimize risk of falling, and help with different aspects of daily function through mobility strategies and respiratory management (Dal Bello-Haas, 2002; O’Callaghan et al., 2014).

### 1.3.2 Occupational Therapy

Occupational therapists use assessment and treatment to enable engagement in daily life activities through occupation (Canadian Association of Occupational Therapists, 2015). The term, “occupation” refers to all activities in which individuals engage and that are essential to one’s health and well-being, therefore, occupations help to describe individuals and how they feel about themselves (Canadian Association of Occupational Therapists, 2015). In ALS, the level of disability progresses as the disease progresses. Occupational therapists prescribe home and personal equipment for patients, demonstrate their proper use and make adjustments when necessary. Like physiotherapy, implementation of hoists or manual handling belts can reduce caregiver burden and prevent secondary musculoskeletal injuries (ALS Society of Canada, 2012). Multidisciplinary care particularly between physiotherapy and occupational therapy is significant for achieving these goals.

Occupational therapists can construct or recommend adaptive and assistive devices such as upper limb orthoses and wheelchairs to assist with motor function in patients with ALS (Ng et

al., 2009). Occupational therapy interventions also include other adaptive equipment such as, customized eating utensils, Velcro ties for dressing, bathroom equipment (ex. rails, toilet frames, shower chairs) etc. (Ng et al., 2009). Due to the degenerative course of ALS, wheelchairs are generally required as ambulation declines. Occupational therapists who anticipate the future needs of the person may consider those necessities when prescribing a powered wheelchair, which can potentially lead to better QoL, as assistive devices can contribute to optimizing the patient's independence and social interaction (Carter et al., 1997; Mayadev, Weiss, Distad, Krivickas, & Carter, 2008). Assistive devices can help prevent contractures, compression nerve palsies, skin breakdown and sores (Francis et al., 1999; Ng et al., 2009). Other useful equipment such as a hospital bed with a pressure relieving mattress, hoists for lifting, and rolling walkers with seats may also be required. Occupational therapists also provide caregivers with the required training prior to the use of certain equipment (e.g. hoists) in order to prevent patient injury.

Occupational therapy interventions can be helpful for individuals with ALS in regards to employment, as engaging in activities that are essential to one's health and well-being falls under the scope of Occupational Therapy. Ng, Talman and Khan (2011) performed a study with 44 participants and found a small yet significant gap from the individuals with ALS in regards to advice and assistance related to continued employment and driving. Healthcare professionals may underestimate the importance of continuing employment as a priority for individuals with ALS (The ALS Association, 2015). This underestimation may lead to insufficient efforts in managing the disease so that continuation of employment is possible and QoL can be maintained for these individuals. Thus, the use of assistive technology may be particularly valuable early on in the disease process (Ng et al., 2011). At present, technology is fast advancing and there are

available selections of different wheelchair types, vehicle assistive devices, keyboard types, and integrated communication access packages that can be used in order to maintain employment.

Overall, it is important for health professionals not to undermine the value patients place on their autonomy within the workplace, and to recognize that individuals with ALS can benefit from advancing technology to continue their career and maintain QoL (Lewis & Rushanan, 2007).

Another aspect of OT interventions is psychological well-being. Wellings and Unsworth (1997) found that environmental control units (ECU) may contribute significantly to the facilitation of function and may decrease dependency on caregivers, improve family dynamics and a patient's self-esteem. Briefly describe what an ECU is here. Because assistive devices are beneficial in maintaining control, it is important that the patients, the patients' family/caregivers and the therapists work together closely before prescribing and using assistive technology (Carter et al., 1997). This collaborative effort helps ensure the correct, safe and optimal use of such devices and equipment, and to predict the patients' future needs. Engaging patients in making decisions maintains their sense of control over their life, thus allowing them to feel more confident and accepting to the new adjustments in their life (Carter et al., 1997). Additionally, it is important for therapists to frequently check the devices as certain technologies and equipment may be expensive and unaffordable if frequently changed or replaced. Ideally, occupational therapists prescribing assistive technological devices closely collaborate with specialized service providers that offer standby technical support, thus reducing the stress for individuals when support is required.

### 1.3.3 Speech and language pathology

Speech and language pathologists are skilled professionals with specialized knowledge, skills, and clinical training in the assessment and management of communication and swallowing disorders (The Ontario Association of Speech-Language Pathologists and Audiologists, 2015). Therefore, speech and language pathology interventions for people with ALS focus on maintaining functional communication as opposed to reducing speech impairment (Pollens, 2012). Dysarthria is common in people with ALS where the bulbar pathways affected. Dysarthria is a motor speech disorder resulting from declined movement of the muscles used for speech production, including the lips, tongue, vocal folds, and/or diaphragm; and it is a major cause of frustration for patients with ALS (ALS Association, 2015). Pollens (2012) identified that in some types of ALS, during the early stages dysarthria consist of nasality, reduced vocal volume and changes in the amount of oral movement and speech. With disease progression, weakness and spasticity of the oral and laryngeal muscles increase, affecting articulateness and leading to imprecise consonant production (e.g. slurred speech), hyper-nasality, poor vocal quality, slow speech rate and breath (Hillel & Miller, 1989). Speech and language pathologists can teach the patient to slow down their speech rate, over-emphasize pronunciation and improve respiratory efficiency through phrasing (Francis et al., 1999).

Due to the progressive decrease in speech intelligibility as ALS worsens, use of augmentative and alternative communication (AAC) is often necessary. Brownlee and Palovack (2007) conclude that the use of AAC strategies may be helpful in optimizing function and assisting with decision-making. AAC devices range from no or low technology (e.g., using gestures) to high technology devices, allowing the patient to have voice output (Brownlee & Palovcak, 2007). One example of a commonly used speech-generating device is a



LightWRITER. This device is dependent on the user's voluntary motor movement, but specific access depends on the abilities of the patient. Examples include pointing with a body part or pointer, using an adapted mouse or joystick or switches and also using scanning technology. For individuals who have no voluntary motor control for communication, the use of a brain-computer interface (BCI) system has been reported to be beneficial and appears promising (Sellers, Vaughan, & Wolpaw, 2010).

Although AAC devices are shown to be helpful among individuals with ALS, there is an emotional aspect that can affect one's QoL as patients resist the use of the device and have difficulty accepting it into their everyday lives (Ng et al., 2009; Pinker & Jackendoff, 2005). The ability to speak and use language is what distinguishes humans from all other species (Brownlee & Palovcak, 2007). Losing this communicative ability and relying on the use of an assisted device instead can lead to a negative emotional effect (Brownlee & Palovcak, 2007). Therefore, the length of time for accepting the use of an AAC in individuals with ALS may range from weeks to months, and some may never accept it at all add reference here.

Individuals with ALS and their families face the challenge of adjusting to a landslide of losses, and one of the most devastating losses is losing the ability to speak. Although current interventions cannot prevent any of these losses, SLPs can assist individuals with ALS to maintain their ability to communicate using compensatory strategies and AAC devices and techniques in order to maintain human communication. SLP interventions offer techniques to improve respiratory efficiency. It can also recommend a variety of communicative devices for people with ALS to choose from and continue their ability to communicate.

Table 1: Overview of the Goals and Interventions from the PT, OT, and SLP Fields of Rehabilitation

Physiotherapy (PT) (O’Callaghan et al., 2014; Dal Bello-Hass & Montes, 2009)	Occupational Therapy (OT) (Canadian Association of Occupational Therapists, 2015)	Speech & Language Pathology (SLP) (Brownlee & Palovcak, 2007; Ng et al., 2009)
<p>- Goals of PT:</p> <ul style="list-style-type: none"> <li>• evaluation and treatment of conditions affecting the muscular, skeletal, and nervous system</li> <li>• regain and/or retain mobility</li> <li>• increase strength</li> <li>• exercise prescription programs</li> <li>• pain management</li> <li>• respiratory symptom management</li> <li>• patient energy conservation</li> </ul> <p>- Interventions include:</p> <ul style="list-style-type: none"> <li>• maximizing patient’s balance and safety</li> <li>• minimize risk of falling</li> <li>• different aspects of daily function through mobility strategies (exercise, stretching) and respiratory management</li> <li>• prevent re-injury during rehabilitation</li> <li>• recommending adaptive and assistive devices e.g. support braces, hoists, orthotics, walkers with seat, hospital bed with a pressure relieving mattress</li> </ul> <p>- Patient and caregiver education</p>	<p>- Goals of OT:</p> <ul style="list-style-type: none"> <li>• adapting to activities of daily living</li> <li>• maintaining employment</li> <li>• engaging in activities</li> <li>• psychological well-being</li> <li>• patient energy conservation</li> </ul> <p>- Interventions include:</p> <ul style="list-style-type: none"> <li>• assessing patient’s condition and home environment in order to prescribe home and personal equipment</li> <li>• alleviate the stress and emotional trauma of adjusting to these lifestyle changes</li> <li>• prescribe environmental control units (ECU), integrated communication access packages, adaptive keyboard types</li> <li>• demonstrating the proper usage and make adjustments when necessary</li> <li>• recommending adaptive and assistive devices e.g. walkers, wheelchairs, hospital bed, built-up cutlery, long-handled aids, and bathroom equipment (rails, over-the-toilet frames, bath boards, shower chairs, commodes)</li> </ul> <p>- Patient and caregiver education</p>	<p>- Goals of SLP:</p> <ul style="list-style-type: none"> <li>• diagnose and treat communication and swallowing disorders</li> <li>• patient energy conservation</li> <li>• determining the extent of a communication or swallowing problem</li> </ul> <p>- Interventions include:</p> <ul style="list-style-type: none"> <li>• providing augmentative and alternative communication (AAC) devices, ranging from no or low technology (e.g. using gestures) to high technology communication devices</li> <li>• provide methods for safe swallowing, slowing speech rate for preservation of energy, over-emphasize pronunciation, and improve respiratory efficiency through phrasing</li> <li>• promotion, prevention, counselling, treatment, consultation, and management</li> </ul> <p>-Patient and caregiver education</p>

#### 1.4 Thesis rationale

The rehabilitation interventions in this thesis include interventions offered by the professions of physiotherapy, occupational therapy, and speech and language pathology. Qualitative and quantitative evidence are both explored in regards to rehabilitation's impact on QoL in people with ALS, as considering evidence from both research approaches may provide useful information to understand the impact, if any at all. While the existing literature on ALS research focuses on a variety of outcomes, QoL has received less direct attention (Stromberg & Weiss, 2006), and is therefore the focus of this thesis.

#### 1.5 Overview of thesis manuscripts

Study # 1: The impact of rehabilitative interventions on quality of life: A qualitative evidence synthesis of personal experiences of individuals with amyotrophic lateral sclerosis

There are many aspects of QoL, but an essential component is the perception of health, not only for the patient but also for caregivers and healthcare professionals. Despite the progressive nature of ALS, QoL does not correlate with measures of physical strength and function, nor does it necessarily decrease over time (Simmons, 2005; Lewis & Rushanan, 2007). Physiotherapy, occupational therapy, and speech and language therapy may enhance QoL in people with ALS by helping them engage in meaningful and functional activities (Traynor et al., 2003). Aiding individuals with ALS by prescribing equipment and techniques can allow them to access social supports and participate in leisure activities. Therefore, it is important to understand how rehabilitation interventions impact QoL in people living with an incurable progressive disease from the perspective of the recipients of these services. This first study answers the

question: How do patients with ALS perceive the potential of rehabilitation services to optimize QoL?

Study # 2: The impact of rehabilitation interventions on quality of life in individuals with amyotrophic lateral sclerosis: A systematic review

Upon symptom appearance and diagnosis of ALS, rehabilitation plays an important part in managing the disease. Specialists from various rehabilitation fields use their knowledge and expertise to understand disease progression, symptom control, managing expectations, issues relating to communication, addressing end of life issues, specialist interventions (i.e. ventilation, exercise), equipment needs, and counselling and support (Royal College of Physicians National Council for Palliative Care and British Society of Rehabilitation Medicine, 2008). The original research included in Chapter 3 includes all levels of quantitative evidence exploring the effectiveness of rehabilitation interventions for people with ALS (including randomized controlled trials, cross-sectional studies, and cohort studies). The study aims to answer the question: In individuals with amyotrophic lateral sclerosis (ALS), what interventions within the scope of rehabilitation are effective for improving quality of life (QoL)?

### 1.6 Thesis objectives:

The thesis was designed to address the following two objectives:

- To explore the impact of rehabilitation on interventions that maintain or improve QoL of individuals with ALS through a qualitative and a quantitative review of original research
- To identify the gaps in current knowledge for future research

## 1.7 Conclusion

Amyotrophic lateral sclerosis (ALS) is an unrelenting, degenerative neurological disease with unknown etiology in the majority of cases (Walling, 1999). Individuals with ALS struggle with carrying out daily life activities as they ultimately face problems with walking, controlling movement, balance, full or partial paralysis, breathing, swallowing and speaking etc.

Rehabilitation disciplines i.e. occupational therapy, physiotherapy, speech and language pathology, or any combination of these 3 are the focus of this thesis. The aim of rehabilitation is to have individuals with ALS self-manage, maintain function and stay independent for as long as possible with the intention to preserve or improve QoL (Gordon, 2013). Therefore, by exploring both qualitative and quantitative evidence, health professionals, people with ALS, their families and carers could increase their understanding of the influence rehabilitation services have on QoL.

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## CHAPTER 2: STUDY ONE

The impact of rehabilitative interventions on quality of life: A qualitative evidence synthesis of personal experiences of individuals with amyotrophic lateral sclerosis

The following paper has been formatted for submission in the Journal of Quality of Life Research.

**The impact of rehabilitative interventions on quality of life: A qualitative evidence synthesis of personal experiences of individuals with amyotrophic lateral sclerosis**

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**The impact of rehabilitative interventions on quality of life: A qualitative evidence synthesis of personal experiences of individuals with amyotrophic lateral sclerosis**

**Abstract**

**Background.** The nature of ALS is progressive and degenerative, thus influencing individuals physically, emotionally, and socially. A broad review of qualitative studies that describe the personal experiences of people with ALS with physiotherapy, occupational therapy and speech and language pathology interventions and how those affect QoL is warranted. **Purpose.** This study synthesizes qualitative research regarding the potential that rehabilitation interventions have to maintain and/or improve QoL from the perspective of people with ALS. **Methods.** The SPIDER search strategy was applied and five articles were selected that addressed the perceived impact rehabilitation has on QoL for individuals with ALS. **Results.** Four themes emerged: the concept of control; adapting interventions to disease stage; struggles with interventions; and barriers between healthcare providers and patients. **Conclusions.** Rehabilitation interventions were perceived to have potential to support QoL by people with ALS. Advantages and limitations of rehabilitation services within this population were identified.

**Key Words**

Amyotrophic lateral sclerosis (ALS), physiotherapy (PT), occupational therapy (OT), speech and language pathology (SLP), quality of life (QoL), rehabilitation

## Introduction

A major concern of rehabilitative interventions for people with amyotrophic lateral sclerosis (ALS) is improving quality of life (QoL) by reducing disability [1-2]. ALS primarily causes muscle weakness and atrophy, affecting activities and participation [1-4]. There is no cure for ALS and the average life expectancy from disease onset to death and/or ventilator dependency ranges between two to four years [1]. QoL has been defined in many ways, but in this manuscript it is conceptualized as “the extent to which our hopes and ambitions are matched by experience” [3]. In other words, it refers to a person’s satisfaction with the way his/her life is progressing, despite the diagnosis of a disease such as ALS, and with the care measures being taken [3]. Rehabilitation is a process aimed at enabling people with disabilities to reach and maintain their “ideal physical, intellectual, psychological and social functional levels” by providing the tools needed to manage independence and control of their life [18]. Physiotherapy (PT), occupational therapy (OT) and speech and language pathology (SLP) are key rehabilitation professions in the care of people of ALS and thus, can be critical to optimizing a person’s QoL. However, the implementation of interventions can overlap, and determining the effectiveness of interventions is an area that warrants much more attention [4]. There is an apparent lack of awareness within the healthcare community about the objectives and benefits of these rehabilitation professions specifically in relation to ALS [2-4].

With the physical, social, and psychological effects of ALS, the goal of healthcare professionals is to continually focus on improving QoL of clients with ALS through appropriate medical, rehabilitative and psychosocial interventions. Some studies have focused on examining the effect of PT, OT and SLP interventions for individuals with ALS [11-15]. However, to our knowledge, there have been no reviews of rehabilitation based on the personal experiences of patients with

ALS. Thus, the purpose of this study was to comprehensively review existing literature, in the form of a metasynthesis, to consider implications for rehabilitation professionals in the treatment of people with ALS. The research question answered by this metasynthesis was: how do people with ALS perceive the potential of rehabilitation services in optimizing QoL?

## **Methods**

A qualitative metasynthesis was chosen as the analytical approach to integrate the findings from studies that explored (directly or indirectly) the effect of rehabilitation on the QoL in individuals with ALS. There are several steps involved with conducting a metasynthesis [5], from search to synthesis.

### Step 1: Search Strategy

The search was conducted in March 2015 using the SPIDER approach (Appendix 1), which comprises the Sample, Phenomenon of Interest, Design, Evaluation, and Research type [5]. The primary researcher (AS) conducted the search for relevant studies using AMED (1985 to March 2015), CINAHL, Embase (1974 to 2015 March 7), MEDLINE (1946 to Present (March 7<sup>th</sup> 2015)), OVID Health Star (1966 to March 2015), PsycINFO (1806 to March Week 1 2015) and Global Health (1973 to 2015 Week 10). As this study aimed to identify personal experiences of ALS patients and the impact of rehabilitation on their QoL, upon consultation with a health research librarian, we developed search terms and keywords including exact terminology and variations of each term/word. For example, amyotrophic lateral sclerosis is also commonly known as motor neuron disease (MND), thus we included MND as a keyword for “Sample”. The same approach was implemented for the other components of the SPIDER search strategy. We the detailed search strategy, search terms, and keywords in Appendix 1.



## Step 2: Criteria for Studies to be Included in the Search

Studies (in English only) were searched from 1980 to March 2015. We selected the year 1980 following the publication by Mikulic, DeLisa, and Miller (1979), which first proposed the idea of using rehabilitation for people with ALS [19]. There were no restrictions on the geographic locations of where studies were conducted. Any study that used qualitative methods of data collection and analyses (i.e., either a stand-alone study or a part of a mixed-methods study) was eligible. The population was limited to adults (18 years and older) diagnosed with ALS at any stage of the disease, and participants should have been receiving or had previously received interventions provided by rehabilitation specialists through PT, SLP, OT and/or a combination of interventions from these professions. The included studies needed to address domains or themes that individuals considered important to optimizing QoL in ALS. For individuals with ALS, longevity is an unlikely opportunity. Therefore, there are certain occurrences which these individuals consider important through the course of their disease i.e. continuing employment, ability to communicate, sense of importance, and the highest level possible of independence.

Upon completion of the database search and removal of duplicates, two investigators (AYS and LL) independently implemented a two level screening process to identify relevant studies. In level 1, both investigators individually screened titles and abstracts and retrieved all potentially relevant articles identified by either investigator for full article review. Citations only available as conference abstracts were excluded (confirmation was obtained by contacting the authors that a full publication was not available). The same two investigators independently reviewed full articles for inclusion. Inter-rater agreement using Cohen's Kappa was calculated for the determination of strength of agreement. As Kappa takes into account the agreement occurring by

chance, it is considered to be a strong measure for calculating agreement [17]. Any areas of disagreement were resolved by discussion, and consensus on inclusion was then established.

### Step 3: Appraisal and Assigning the Level of Evidence of the Studies

The quality of the studies was assessed using McMaster University's Guidelines for Qualitative Review Version 2.0[6], and the Rosalind Franklin – Qualitative Research Appraisal (RF-QRA) instrument [7]. The McMaster guideline assesses study quality considering the purpose of the study, literature review, design, sampling methods, data collection and analysis, overall rigour, conclusions and implications [6]. The RF-QRA [7] assesses the quality of a study based on Guba's four aspects of trustworthiness (credibility, transferability, dependability, and confirmability) to determine the level of evidence [8]. Using both tools ensured a greater level of trustworthiness and relevance than would have been achieved using either tool alone.

### Step 4: Data Extraction and Synthesizing the Findings

Once the studies were identified, two authors (AYS& LL) used a data extraction form (Appendix 2) to describe the study characteristics, study participants, and conclusions. Data extracted provided rich information about attitudes, opinions, and observations reported by participants regarding rehabilitation services. All findings were then compiled in a single chart, which facilitated identification of common findings across studies. The descriptive findings across studies ultimately resulted in the emergence of themes, based on the significant statements regarding rehabilitation interventions that impacted QoL. Inductive reasoning was used by formulating generalizations through observing patterns and describing interpretations. The next step involved synthesising findings using methods described by Sandelowski and Barroso [9]. These methods included comparing and contrasting concepts, identifying themes across studies,

and synthesizing results. Once themes were identified, and data were clustered by theme, further detailed review resulted in identification of sub-themes or concepts. Furthermore, comparisons of similarities and differences were made, ultimately developing an interrelated explanation across all studies [10]. This resulted in formulation of final themes and appropriate sub-headings (concepts) for each of them. No software was used for the organization of themes. The identified themes were verified by another author (LL) to confirm that the information obtained during the process of data extraction and synthesis was deduced in a coherent manner and no major concepts were omitted.

### **Findings**

The obtained search results are presented in Figure 1. Of 506 citations, 5 articles including 87 people with ALS met inclusion criteria. Cohen's Kappa was calculated to be 0.318, and the proportion of observed agreement ( $p_o$ ) was 0.85.

Four of the five studies were ranked as Level I evidence and one study was ranked as Level II, as it did not meet the confirmability component (Table 1). From the five included studies, none were performed with the express or sole purpose of identifying if any relationship existed between rehabilitation and QoL; however, this topic was addressed through the conduct of these studies but not explicitly the topic of the study. Due to this, exploration of how rehabilitation influences QoL required interpretation of data, as described in the methods section.

The included studies consisted of four studies specific to people with ALS [11 – 14], and one regarding degenerative neurological conditions with specific identification of participants with ALS [15]. Of the included studies, not all contained information regarding the duration or type of ALS in all participants. The most recent study performed was published in 2014 [11] which

aimed to understand the meaning of loss in people with ALS and how they exert control in care decisions in response to loss. All 10 participants in this study [11] were receiving rehabilitation interventions i.e., Augmentative and alternative communication (AAC) devices to communicate and the duration of the disease ranged from 4 months to 13 years. Hughes et al. [12] studied the experiences and impacts of ALS and how services can be improved to meet people's needs. The study addressed the questions: "What are the lived experiences of people living with ALS? What are people's experiences of services? Can improvements to care be identified?" [12]. Two studies [13, 14], investigated the experiences of people with ALS using AAC devices for all communication. However, one [13] aimed to study communication of people with ALS with their environment, whereas the other [14] focused on the maintenance of employment. The final study [15] included 6 participants diagnosed with ALS and aimed to determine the opinions of individuals with neurological conditions on factors facilitating their physical activity participation. Despite variations in the primary purpose of the studies, QoL was incorporated in various ways, and enabled the meta-analysis to focus on themes related to rehabilitation and QoL.

### **Concept of Control**

As disease symptoms emerge, progress and influence abilities, individuals with ALS started to feel a loss of control over their lives due to the significant increase in dependency on others [11] and the decreased ability to pursue hobbies and pastimes [12]. One consistent theme across all studies was the notion of control, with respect to the use of interventions. The theme focused on how people with ALS are cared for by family and caregivers, and their ability to make employment decisions. The studies indicated that for people with ALS to preserve and/or improve QoL, it is important that rehabilitation providers help maintain the person's

independence for as long as possible so that they feel empowered and in control of their lives.

However, it is crucial that rehabilitation providers consider the needs and interests of the person with ALS.

### Choosing When to Use PT, OT and SLP Services

Having control over decisions such as the type and timing of intervention services was important for individuals. Individuals who felt they were losing control over ALS and their lives sometimes attempted to exert control in their use of rehabilitation services. Exerting control over these healthcare services helped patients establish feelings of control over their lives. In addition, while dependency on rehabilitation services was perceived negatively, individuals understood that the use and implementation of these services were important to maintaining a level of control in their lives: *“Well I suppose if I was working outside the home I would be using my voice more whereas I’m here a lot on my own, my only social is playing golf, so I feel that I’m doing ok. I do find it difficult talking but I don’t have to talk. You see that’s why I feel that I don’t need to see a speech therapist [locally] yet”* [11].

### Struggles with Losing Control

Individuals with ALS want to control how they are cared for as it provides them with the sense of control over their lives, which in turn affected QoL. This is significant because the loss of such control was associated with ‘having no choice’ and this left people feeling forced to adapt and accept the disease, as well as the healthcare services, even though they were not yet ready to accommodate for that: *“I know I can’t walk so I have to use a wheelchair. I know I can’t stand up so I have to be lifted up and I have to be hoisted into bed and hoisted onto the toilet... You don’t have a choice. I mean if you are sitting there and you can’t get up, what choice have you*

*only to be lifted up*” [11]. Despite quotes like this, it was clear that accepting loss of control was a challenge.

People with ALS also described feeling powerless in the face of physical impairment(s) and therefore were accepting to the prospect of further loss. This was a strategy used to address the struggle of losing control. Once the individuals were mindful that they had no control over the disease progression, they decided that having a positive outlook gave them a sense of control over it. Allowing people with ALS to have the space and freedom to process the disease progression fostered feelings of control, and also encouraged impartiality towards required care:

*“... it’s just no(t) worth worrying about. I can spend the next few years of my life worrying about this illness, it’s gonna eventually gonna kill me, or I can just go out there and enjoy it, and you know, let it do its worse. I think, having a positive attitude, it helps you in the long run anyway, it sort of keeps you more fitter”* [12].

### Maintenance of Employment

Despite having a degenerative disease, patients acknowledged the importance of maintaining employment in order to feel independent and in charge of their lives, and to maintain QoL by maintaining control wherever possible. More specifically, patients attributed the following benefits to maintaining employment: being involved in something meaningful; attaching their personal identity to their profession; positive experiences created from the workplace environment; and financial benefits [14]. All of these benefits were deemed to increase QoL in patients living with ALS. Although maintaining employment with ALS is quite challenging, people spoke about how rehabilitation services were key to retaining their jobs and allowing them to be independent: *“I felt I still had a lot to contribute and could not face an abrupt*

*transition to retirement...Being able to write again [using AAC technologies] has given my life purpose, a reason to get up and get going every day” [14].*

### **Adapting Interventions to Disease Stage**

Due to the progressive nature of ALS, frequent adjustments are required for overall management. However, these adjustments are not always identified or implemented by patients or healthcare professionals, which can in turn negatively affect QoL.

#### Adapting Exercise Protocols

While persons are still ambulatory, exercise is commonly prescribed to maintain function, muscle strength, and to reduce fatigue [4, 15, 20], which in turn should maintain QoL. The included studies that addressed exercise intervention highlighted the established need to modify exercise interventions from original protocols to accommodate individuals’ needs as their conditions worsen [15]. As physical abilities are different for each patient along the disease continuum, people with ALS want their healthcare providers to pay particular attention to their specific physical abilities and how these abilities change over the course of the disease. Some commented that at times it was easy for them to do more than their body could handle, ultimately resulting in severe aches, pain, and fatigue both physically and mentally. Introducing exercise with the use of assistive devices and/or assistance from an expert with knowledge about ALS, how ALS affects the individual, and prescribing exercise for people with ALS (i.e., by physiotherapists) may allow people with ALS to continue participation in such activities safely and with the necessary adjustments for the disease stage [11, 15]: *“I want a trainer to be familiar with [my] condition and confident to deal with me. [I’m] fed up with constantly having to explain everything when there are changes to my condition” [15].*

### Assistive Devices

The type of assistive device required varies considerably and frequently changes from early to middle stages [11]. Dependence on such devices was regarded as a constant reminder of what patients had already lost. Thus, avoidance of assistive devices until they were absolutely necessary for daily function symbolizes how patients adapted to the loss of functioning throughout disease progression. While patients felt assistive devices were absolutely necessary for achieving the highest possible level of independence [11, 14], adjusting to the abrupt changes of devices was difficult. *“It [ALS] turned me into an old man overnight because suddenly you’re going with a walking stick and then you’re with a rollator, now you’re in this wheelchair and you’ve really, it’s just a robbery of everything”* [11].

### AAC Devices

A commonly identified issue with adapting to AAC devices for patients with ALS was a lack of required skills and adequate training of the therapists using technological equipment [13-14]. On the other hand, patients also commented about the difficulty of learning to use the AAC devices because of limited input and training that was provided from therapists [13-14]. Many people commented that they were provided with instruction manuals to learn from and/or given a quick demonstration but that was still too difficult, complicated, or technical to follow [13]: *“I’ve looked at the manual [grimaces] but I don’t need to use it as comprehensively as that,”* *“Learning something new like this is too hard – but don’t tell my speech language therapist”* [13].

## **Struggles with Rehabilitation Interventions and Accessing Services**

### Rehabilitation Interventions



A major issue identified by patients with ALS was the difficulty involved with incorporating their assistive and AAC devices into their everyday lives, which negatively affected their QoL. For example, fitness facilities, work facilities, bathrooms, etc. were inaccessible for the most part, and finding access routes, doorways for wheelchairs to pass through, and lifts were all impediments to accessing and using mobility devices [12, 14-15].

Some people avoided rehabilitation supports if their condition was not affecting their social interactions. In addition, patients placed significance on maintaining social connections, and believed that using some AAC devices hindered these connections. As explained by one patient with ALS, *“I mean I prefer contact this close [using an alphabet board] rather than trying to type something with the Lightwriter™ . . . to lose that personal contact I think is detrimental really”* [13].

#### Difficulty Accessing Services

Many people with ALS mentioned that they were unsure about the services and options available, specifically after diagnosis. Individuals, families and caregivers expressed that they received very little help navigating available services and information regarding interventions and assistive devices that might have improved their QoL:

*“... [T]hings like wheelchairs and things like that ... my question is, ‘Where does he get that equipment?’ Does he get it from the hospital? Do we get them from the hospital or is it the occupational therapist?”* [12]

*“Most people with ALS are just too devastated to fight for the care, equipment, and support they need . . . to overcome the frustrations associated with the disease, and to be able to actively seek help from others”* [14].

Another identified area of concern occurred when individuals obtained assistive devices or AAC technologies independently or with the assistance from an ALS support worker rather than a rehabilitation specialist. Although there were no direct quotes from participants with respect to obtaining assistive devices independent of a specialist, there was mention of the difficulty trying to obtain equipment in general [12]. Regardless, apparent frustration with the lack of easy access to information about the AAC technologies was abundant across studies [11-14].

### **Barriers between healthcare providers and patients**

#### Healthcare Providers Need to Enhance Knowledge of the Disease

People living with ALS interact with healthcare providers on a regular basis. Patients emphasized concerns regarding a lack of knowledge and understanding of ALS by healthcare professionals and the resulting impact this had on the lives of patients and their caregivers [11, 12, and 15]. Patients were aware that this limited knowledge of ALS amongst some professionals was likely due to the low incidence (1.6 persons per a 100,000 population) of the disease. In addition patients' concerns, some professionals also identified the need to enhance their own knowledge about ALS.

The lack of understanding about the illness was especially troublesome for people with ALS because this led to problems when trying to obtain service entitlements, such as financial support for inability to work, which in turn could influence QoL. Some suggested that having support from a fitness specialist and/or neuro-physiotherapist knowledgeable in ALS exercise protocols would help them feel more confident during the exercise sessions. On the other hand, for patients attending fitness facilities for rehabilitation as prescribed by the health professionals, many reported the lack of confidence in the fitness staff who were assigned to assist them with the

exercises. There was concern regarding the amount of knowledge fitness staff members had about the condition and knowing which exercises were appropriate for them [15].

Some commented that the rarity of the disease meant that people did not always understand the disease and disengaged from their healthcare professionals if they felt that their requirements of care were being ignored. Individuals with ALS did not blame healthcare professionals for their unfamiliarity with ALS as patients themselves admitted, “*some people have never heard of motor neurone disease – I hadn’t*” [12]. They identified that due to the low incidence of the disease, it is not uncommon for therapists to have minimal to no knowledge of the disease.

#### Too Much Information Leads to Confusion

Several participants commented on the importance of receiving the appropriate information at the appropriate time. As diagnosis can be difficult, and health professionals often failing to provide an effective treatment plan, patients and their families often reported ‘seeking information for themselves’ [11-12, 14]. As one patient with ALS explained, this can be difficult because “*you’ve either got the person who wants to throw themselves into it, find out as much information as possible, or the ostriches like me who just want to bury their heads in the sand*” [14]. Therefore, some studies identified that it is important for healthcare professionals to assess their patients before providing them with information regarding their condition. Individuals living with the illness for longer periods were more open to receiving information regarding treatments, therapies and research [12].

## Discussion

The findings in this metasynthesis highlight the perspectives of individuals with ALS regarding their rehabilitation care. The impact of the disease is not only physical but also influences these individuals emotionally and psychologically, especially surrounding concerns of losing control over many aspects of life, including use of rehabilitation services. Additionally, adaptation and struggles with rehabilitation services and interventions were expressed along with how patients envisioned their care from healthcare professionals. Overall, these findings provide detail about how patients with ALS perceive the potential of rehabilitation services in optimizing QoL.

Among patients with ALS, feeling in control was central to making life decisions in terms of accepting the disease, treatment options and employment after diagnosis. It may be important that healthcare professionals first allow these individuals to come to terms with the disease and the necessary adjustments required to maintain QoL before introducing specific interventions, such as exercise interventions, assistive devices, and AAC technologies. By doing this, patients may feel less overwhelmed and insignificant due to having a deteriorating condition, and as a result, they may have a more optimistic perception about the interventions.

Most people with ALS struggled with adapting to the rehabilitation interventions and accessing available services. The main issue with adapting to the interventions was due to the rate at which their condition and interventions changed, where some individuals felt it was a sudden change from a cane to a walker [11], consequently leading to difficulties with adherence to exercise protocols or other interventions. The unpredictability of ALS was too devastating for some to handle and left many struggling to adapt to the interventions.

Additionally, the lack of knowledge about ALS among healthcare professionals and service providers did not make accessibility to rehabilitation services easy, leading to frustration for the patients and their families. Some patients felt that they would have liked to be informed of what to expect regarding the disease after being diagnosed so there would be no surprises at each stage of the disease. Therefore, having health professionals provide patients with the necessary information to obtain such support could reduce the patients' perceived need to look for information and distressing themselves unnecessarily.

Across all studies, rehabilitation interventions positively contributed in managing the goals of individuals with ALS, especially in terms of independence and employment, which could in turn be seen to optimize QoL. Dependence on assistive and communicative devices was not perceived negatively when individuals were still able to perform daily living activities without too much dependence on a family member or caregiver. Although some patients did not agree that exercise and physical activity would benefit their condition, they participated in such activities because they enjoyed the social interactions [14-15]. Social contact was identified as an important part of QoL, and AAC devices played a significant role in the maintenance of communication.

However, AAC technologies were noted to be difficult to learn to use due to the technological difficulties; yet many patients found them to be very beneficial specifically when talking became tiresome.

This metasynthesis highlights the importance of rehabilitation interventions particularly assistive and communication aids that significantly facilitate maintaining a 'normal life' despite having ALS. QoL, a multifaceted concept, was supported through the increased ability to remain independence and adequately function to carry out daily life activities. ALS is a progressive disease that requires a variety of coping strategies at the different disease stages [16]. Therefore,

it is important that people with ALS and health professionals recognize both the short and long term goals and the challenges that will be associated with them.

### **Limitations**

The reporting of this metasynthesis was guided by the ENTREQ guidelines, with detailed information on each of the reporting criteria shared in Appendix 3. All reporting criteria to demonstrate the quality of the metasynthesis were addressed. The low score calculated using Cohen's Kappa was possibly due to a small sample size or low variance and differences in raters in judging those articles for which there was some degree of uncertainty about the application of inclusion/exclusion criteria. In the 2x2 table used to calculate kappa, it is apparent that one rater (AYS) had a much higher tendency to include articles than the other rater; the resulting asymmetry in the table likely resulted in a lower kappa score than might typically be expected considering an 85% overall agreement between raters[17]. Additionally, restricting this synthesis to articles published in English meant that articles published in other languages (e.g., Spanish, Dutch and Japanese) were excluded which may have excluded other perspectives. Another limitation is associated with the data analyses being conducted primarily by one investigator. However, data credibility was enhanced by having two investigators extract the data, and triangulation was implemented when the emerging themes were confirmed by a second investigator.

This metasynthesis was restricted to the small number of qualitative and mixed-methods studies available in the literature related to PT, OT and SLP interventions. As none of the included studies were performed exclusively to identify if any relationship existed between rehabilitation and QoL, the topic emerged from the conduct of the studies. Thus, interpretation of how

rehabilitation influences QoL was challenging. Additionally, the selected studies recruited their samples specifically to target their primary objectives. Therefore, this metasynthesis may not have captured all shared experiences from people with ALS who used rehabilitation services to maintain QoL. While suggesting the potential contributions that rehabilitation can make to support QoL for people with ALS, targeted qualitative investigations specifically aimed at understanding the experiences of people with ALS receiving rehabilitation would contribute confirmation and perhaps new insights.

### **Conclusion**

The study integrates qualitative evidence concerning rehabilitation interventions for maintaining and/or improving QoL for people with ALS. The metasynthesis findings show the potential rehabilitation interventions have to enhance the QoL for individuals with ALS; the information may help healthcare providers and researchers to expand practice and research in this area. The professions of PT, OT and SLP have the ability to influence QoL, as these rehabilitation professions have expertise in methods to maintain a person's activities of daily living throughout the course of the disease i.e. mobility, communication, and home safety, which may in turn support control over life, an important concept emerging from the papers reviewed. Although more research is needed, this metasynthesis identifies some benefits and drawbacks of rehabilitation. Nevertheless, the evidence suggests that from the perspective of patients, PT, OT, and SLP interventions have potential to be beneficial in managing ALS to optimize QoL.

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<b>Table 1: Details of Included Studies</b>						
Author & Publication year and place	Sample size (age, sex)	Purpose of the study	Design of the study	Participants' type of ALS and/or duration with disease	4 levels of Trustworthiness	RF-QRA Level of Qualitative Evidence
Foley et al., 2014 (Ireland)	N = 34 (37 – 81 years, 17 men and 17 women)	The study focused on understanding the meaning of loss for individuals with ALS and how they exert control in their care decisions in response to loss.	Grounded Theory	Type of ALS: 10 participants used AAC devices to communicate Duration: 4 months to 13 years	Credibility: Y (audio-recorded and information of the authors roles for transcribing and analyzing findings all described) Transferability: Y (detailed information of participants and settings provided) Dependability: Y (Clear explanation of data collection and analysis) Confirmability: Y	Level: I
Hughes et al., 2005 (United Kingdom)	N = 9 (30 – ≥ 70 years, 6 men and 3 women)	This study aimed to understand the impacts of MND and how services can be improved to meet people's needs.	Phenomenology	Type of ALS: Not reported Duration: Less than 1 year to over 3years	Credibility: Y (Field notes, audio-taped interviews) Transferability: Y (settings, time of interview, and participant	Level: I

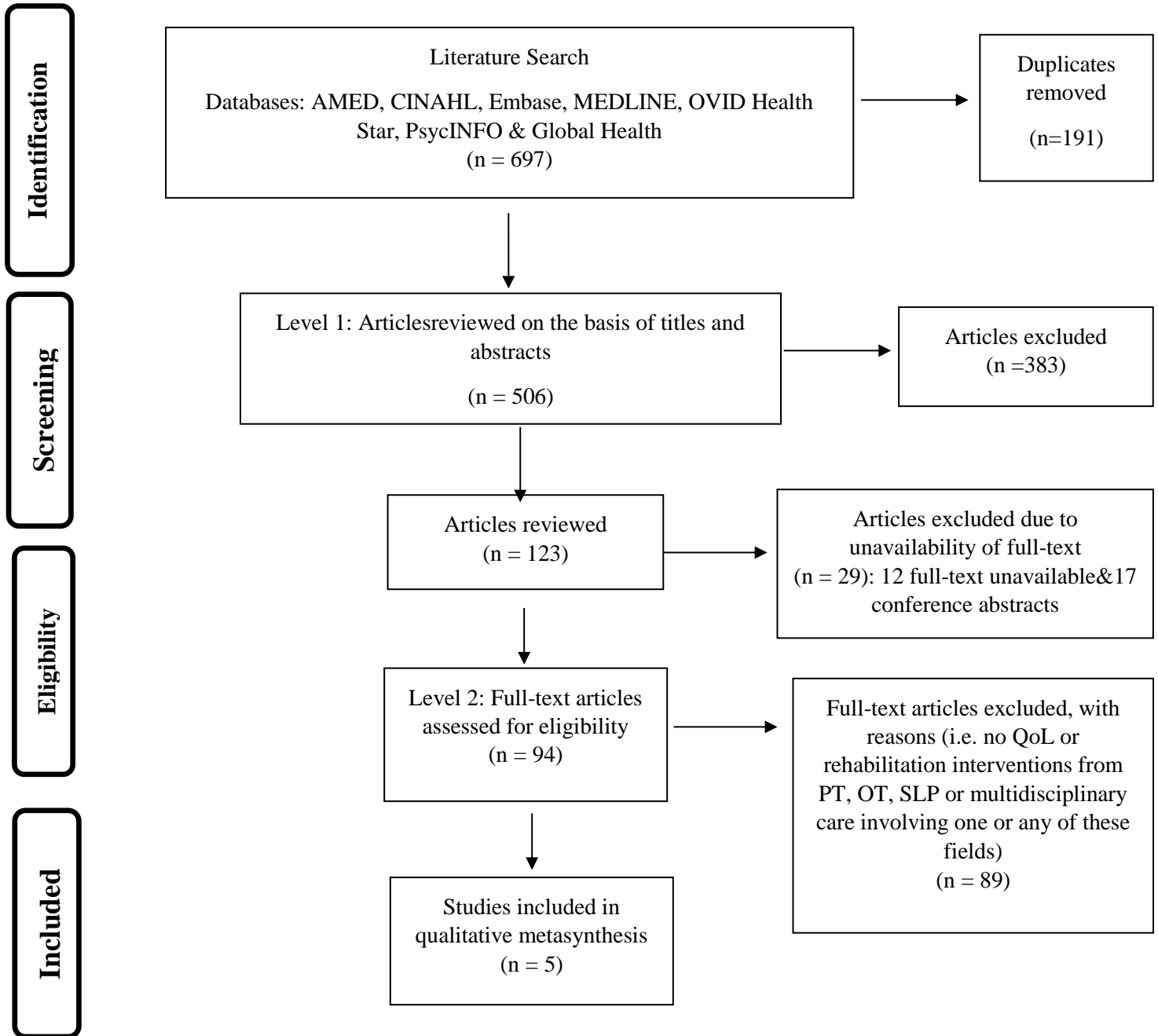
					information provided in detail) Dependability: Y (Clear explanation of data collection and analysis provided) Confirmability: Y (limitations addressed and procedures to minimize bias noted)	
Elsworth, et al., 2009 (United Kingdom)	N = 24 (mean age 54 years) N (MND) = 6	The study aimed to determine the opinions of individuals with neurological conditions on factors facilitating their physical activity participation.	Mixed-methods	Type and duration: Not reported	Credibility: Y (Notes, independent note analysis by researchers to identify major themes) Transferability: Y (settings, interview content and questions, and participant information provided) Dependability: Y (quantitative approach for data analysis of developed questionnaire)	Level: I

					results, clear explanation of qualitative approach for focus group interviews) Confirmability: Y (limitations addressed and procedures to minimize bias noted)	
McNaughton et al., 2001 (USA)	N = 5 (52 – 57 years, 2 men and 3 women)	This study investigated the experiences of five individuals with ALS who required AAC, including the types of employment activities in which they were engaged and their use of AAC systems.	Phenomenology	Type of ALS: 2 not reported Bulbar = 2 Spinal = 1 Duration: 1 to 8 years	Credibility: Y (focus group conducted via internet, details of data analysis explained) Transferability: Y (details of all participants and their work settings provided) Dependability: Y (data analysis and collection procedures provided) Confirmability: Y	Level: I
Murphy, 2004 (United Kingdom)	N = 15 (45 – 78 years, 8 men and 7 women)	The purpose of the communication and the use of augmentative and alternative	Qualitative case study methodology was used and involved (a)	Type of ALS: Bulbar = 13 Spinal = 2	Credibility: Y (video recorded, field notes, and narratives used, researcher assistant	Level: II

		communication (AAC) according to the perceptions of people with MND and their partners were examined.	using video recordings of participants in open conversation, (b) semi-structured interviews in order to gain rich and detailed narrative information from a number of sources, and (c) field notes.	Duration: Not reported	transcribed data, author and SLP discussed findings to determine themes) Transferability: Y (detailed information of participants and settings provided) Dependability: Y (information of data collection and analysis provided) Confirmability: N (limitations addressed, however no information of peer review)	
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Abbreviations: AAC – Augmentative & Alternative Communication, ALS – Amyotrophic lateral sclerosis, MND – motor neuron(e) disease

**Figure 1: PRISMA flow diagram for study selection**



## **Appendix 1: Keywords and Medical Subject Headings (MeSH) terms**

### **Group 1: Sample**

1. Amyotrophic lateral sclerosis (MeSH exploded not focused) (keyword)
2. ALS search as keyword
3. motor neuron disease
4. motor neurone disease
5. Charcot disease (keyword)
6. Lou Gehrig's disease (keyword)
7. Combine 1 or 2 or 3 or 4 or 5 or 6 → 7

### **Group 2: Phenomenon of Interest**

8. Rehabilitation (explode not focused) (keyword)
9. Therapy
10. Physiotherapy
11. Physical therapy
12. Occupational therapy
13. speech and language pathology/therapy
14. Group 8 or 9 or 10 or 11 or 12 or 13 → 14
15. Combine 7 and 14 → 15
16. ALS – MeSH + rh, th [Rehabilitation, Therapy] → 16
17. Combine 15 or 16

### **Group 3: Design**

18. Questionnaire
19. Survey
20. Interview
21. Focus group
22. Observ\*
23. Case stud\*
24. Combine 18 or 19 or 20 or 21 or 22 or 23 → 24

### **Group 4: Evaluation**

25. Quality of life
26. QoL
27. disease management
28. functional status/ ability
29. experience



- 30. health status
- 31. activities of daily living
- 32. view
- 33. opinion
- 34. attitude
- 35. feel\*
- 36. perce\*
- 37. belie\*
- 38. combine 25 or 26 or 27 or 28 or 29 or 30 or 31 or 32 or 33 or 34 or 35 or 36 or 37 → 38

**Group 5: Research type:**

- 39. Qualitative
- 40. Mixed method
- 41. Combine 39 or 40 → 41

Add limitations Humans, 1980 – Current, English

**Appendix 2: Data Extraction Form****General Information**

Title of study/ID	
Reference citation (e.g. CINAHL)	
Study author details (e.g. _____ et al., 2010)	
Publication type (e.g. phenomenology)	
Date form completed (dd/mm/yyyy)	
Name of person extracting data	

**Study eligibility**

Study Characteristics	Eligibility criteria		Eligibility criteria met?		
			Yes	No	Unclear
Type of study Circle one →	Qualitative	Mixed-methods	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Sample	Adults (18 years and older) diagnosed with ALS		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
	Any stage of the disease → stage identified in study?		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Phenomenon of Interest	Rehabilitation interventions received or have received through physiotherapy, occupational therapy, speech and language pathology or the combination of interventions from these rehabilitation fields		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
	- PT		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>

	- SPL	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
	- OT	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
	- Combination of PT/OT/SLP	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Design	Interviews, questionnaires, surveys, observations, focus groups	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Evaluation	- Quality of life	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
	- Positive and negative aspects of rehabilitation interventions	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
	- Functional status	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
	- Disease management	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Results	Qualitative data analysis of the impact of rehabilitation interventions on QoL	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
INCLUDE <input type="checkbox"/> EXCLUDE <input type="checkbox"/>				
Reason for exclusion				

**STOP HERE IF THE STUDY DOES NOT MEET INCLUSION CRITERIA**

**IF INCLUDED, THEN PROCEED TO THE NEXT PAGE AND/OR THE CRITICAL APPRAISAL FORM(S)**

**Characteristics of included studies****Methods& Results**

	Description of the study	
Study purpose		
Design		
Setting		
Duration of participation		
Main Findings		
Ethical approval obtained for study	<input type="checkbox"/> Yes <input type="checkbox"/> No <input type="checkbox"/> Unclear	Withdrawals from study:

**Participants Characteristics**

	Description	
Population description (from which study participants are drawn)	Male	Age
	Female	Age
Total no. of participants		

Ethnicity information (circle one)    Yes    No  If yes, describe →	
Participant information (e.g. time since diagnosis, stage of disease, treatments undergoing/undergone, experience with rehabilitation services etc.)	

<b>Appendix 3: Enhancing transparency in reporting the synthesis of qualitative research: the ENTREQ statement</b>		
No	Item	Description
1	Aim	The research question to be answered by this metasynthesis is: how do patients with ALS perceive the potential of rehabilitation services to optimize QoL? (p.3)
2	Synthesis methodology	Qualitative metasynthesis → provide a comprehensive review of existing literature to consider implications for healthcare professionals in the treatment of ALS (4 steps) (p. 3-6)
3	Approach to Searching	The search was conducted using the SPIDER approach (Appendix 1) (p.4) and a research librarian provided consultation support
4	Inclusion criteria	<ul style="list-style-type: none"> <li>- Studies (in English only)</li> <li>- 1980 to March 2015</li> <li>- Qualitative methods of data collection and analyses (i.e. either a stand-alone study or a part of a larger mixed-method study)</li> <li>- 18 years and older diagnosed with ALS at any stage of the disease</li> <li>- Participants should be receiving or have received interventions provided by rehabilitation specialists through PT, OT, SLP and/or the combination of interventions from these professions</li> <li>- Studies that addressed domains or themes that patients considered important to improving QoL in ALS (p.5)</li> </ul>
5	Data Sources	<ul style="list-style-type: none"> <li>- AMED, CINAHL, Embase, MEDLINE, OVID Health Star, PsycINFO and Global Health</li> <li>- Search was performed in March 2015 (p.4)</li> </ul>
6	Electronic Search Strategy	Appendix 1 & p.5
7	Study Screening Methods	A two level screening process to independently identify relevant studies. In level 1, both investigators individually screened citations, excluded articles only available as conference abstracts. In level 2, the full articles were independently reviewed by the same two investigators, who made recommendations for inclusion (p. 5). Figure 1 (p.26)
8	Study characteristics	Table 2 (p. 22)
9	Study selection results	The obtained search results are presented in Figure 1 (p. 6 & 26)
10	Rationale for appraisal	The McMaster guideline [6] and the RF-QRA [7] based on Guba's four aspects of trustworthiness to grade the level of evidence [8]. Using both tools ensured a greater level of trustworthiness and relevance than would have been achieved using either tool alone (p.5)
11	Appraisal items	The quality of the studies was assessed using McMaster University's Guidelines for Qualitative Review Version 2.0 [6] and the Rosalind Franklin – Qualitative Research Appraisal (RF-QRA) instrument [7] (p. 5)
12	Appraisal process	Table 2 (p.26)
13	Appraisal results	Table 2 (p. 26)

14	Data extraction	Data extraction and synthesis methods described by Sandelowski and Barroso [9] (p.6)
15	Software	No software was used for the organization of themes (p. 6)
16	Number of reviewers	two investigators (AYS and LL) implemented a two level screening process to independently identify relevant studies (p. 5). AYS and LL also independently completed data extraction forms for selected studies.
17	Coding	Once studies were identified, the next step involved synthesizing findings across studies using data extraction and synthesis methods described by Sandelowski and Barroso [9] (p. 5)
18	Study comparison	Once themes were identified, we identified of similar concepts, making comparisons of similarities and differences, and developing an interrelated explanation across all studies [10] (p. 6)
19	Derivation of themes	Inductive reasoning was used by making generalizations through observing patterns and describing interpretations (p. 6)
20	Quotations	(p.7 to p.14)
21	Synthesis output	The metasynthesis findings show the potential rehabilitation interventions have to enhance the QoL for individuals with ALS; the information may also help healthcare providers and researchers to expand practice and research in this area. The evidence suggests that from the perspective of patients, PT, OT, and SLP interventions are beneficial in managing ALS to optimize QoL (p. 17)

## CHAPTER 3: STUDY TWO

### **The impact of rehabilitation interventions on quality of life in individuals with amyotrophic lateral sclerosis: A Systematic Review**

The following paper has been formatted for submission in the Journal of Critical Reviews™ in Physical and Rehabilitation Medicine.



**The impact of rehabilitation interventions on quality of life in individuals with  
amyotrophic lateral sclerosis: A Systematic Review**

(Note: Written and formatted for Publication in the journal of Critical Reviews™ in Physical and Rehabilitation Medicine. The article has not been published elsewhere and it has not been simultaneously submitted for publication elsewhere. Further editing is required to meet the word count for publication.)

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## Abstract

**Background.** Many factors can affect a person's outlook on his/her life; rehabilitation interventions vary greatly and little is known about the effect of rehabilitation to maintain and/or enhance quality of life (QoL) in people with amyotrophic lateral sclerosis (ALS). **Objective.** This systematic review aimed to determine the effectiveness of interventions within the scope of rehabilitation including physiotherapy (PT), occupational therapy (OT) and speech and language pathology (SLP) on QoL in people with ALS. **Methods.** A comprehensive search of the following databases was performed: MEDLINE; CINAHL; PsycINFO; The Physiotherapy Evidence database (PEDro); Occupational Therapy Systematic Evaluation of Evidence (OT seeker); AMED; EMBASE; OVID Health Star; Global Health; and LILACS EN. Inclusion criteria were adults diagnosed with ALS who received rehabilitation interventions from the PT, OT, and/or SLP professions reporting QoL outcomes measured by the Short-Form 36 Questionnaire or Amyotrophic Lateral Sclerosis Assessment Questionnaires. We included quantitative and/or mixed-methods studies. We appraised risk of bias using the Cochrane Risk of Bias tool for randomized controlled trials (RCT), and the New-Castle Ottawa scale for the non-RCTs. **Results.** A narrative synthesis of interventions was conducted as data extracted were insufficient for conducting a meta-analysis. Six studies enrolling 460 ALS patients met inclusion criteria: three RCTs, two cohort studies, and one cross-sectional study. Interventions included exercise, communication devices, and multidisciplinary care. The studies found that people using rehabilitation interventions have a higher QoL compared to those who do not receive any. The studies provided a moderate level of evidence indicating the potential benefit of rehabilitation interventions in maintaining and/or improving QoL. **Conclusion.** The roles of PT, OT and SLP have the potential to play an important role in delivering appropriate management of the disease

to improve QoL. However, based on the limited evidence available, it is difficult to determine the comparative effects of the interventions from each rehabilitation field on improving QoL for people with ALS. More research needs to be carried out in this field to identify the most effective interventions to sustain QoL in ALS.

### **Key Words**

Amyotrophic lateral sclerosis (ALS), physiotherapy (PT), occupational therapy (OT), speech and language pathology (SLP), quality of life (QoL), rehabilitation

## **I. BACKGROUND**

Amyotrophic lateral sclerosis (ALS) is a degenerative, disabling and fatal motor neuron disease affecting 450,000 adults worldwide (32). The prognosis of ALS is affected by age at disease onset, rate of progression, and presence of bulbar signs early in the disease (1, 2). Currently there is no cure for ALS and the cause of the disease remains unknown. Therefore, treatment is based on symptom management to optimize lifespan and maximize QoL (3, 4).

Individuals with ALS experience muscle weakness due to the progressive degeneration of motor neurons in the spinal cord, brainstem, and cerebral hemispheres (5). This ultimately results in activity limitations and participation restrictions for these individuals. About 50% of people experience initial signs and symptoms in the upper extremities, whereas for 40% of individuals the onset occurs in the lower extremities (6). In the remaining 10%, onset may be observed in areas of the bulbar region showing symptoms of changes in voice and speech, poor articulation, a decrease in range of pitch and loudness of voice, spasms in muscles of the jaw, face, voice box, throat and tongue (7).

As the disease progression can occur rapidly, it is essential that people with ALS have quick access to services from a range of health professionals that are skilled and effective in the management of the physical and psychosocial developments that occur with this condition (10, 11). Several interventions from physiotherapists (PTs), occupational therapists (OTs), and speech and language pathologists (SLPs) can directly target functional impairments, physical activity, and address patients' limitations with regard to participation in social, home, work, leisure, and community roles (5). The nature of ALS can significantly affect a person's feelings of hope and self-worth. Additionally, one's personal activities, fulfillment of life goals, and continuing employment are also affected (12). Changes in self-esteem, loss of independence, and

psychosocial stress all play important roles in an individual's experiences of QoL throughout the disease process (1). To maintain independence and maximize QoL, many individuals seek the services of rehabilitation professionals for the purposes of “advice, education, movement rehabilitation, speech/swallowing rehabilitation, environmental modification, workplace adaptation, and the provision of assistive devices” (5) for environmental control and to assist with communication.

Rehabilitation is a process aimed at enabling people with disabilities to attain and maintain the best “physical, intellectual, psychological and social functional levels” possible by offering the tools necessary to manage independence and control of their life (31). Rehabilitation professionals use their knowledge and expertise to assist individuals with ALS to understand the disease progression, control symptoms, manage expectations, address communication issues, identify and address end of life issues, fulfill equipment needs, provide counselling and support, and advise on social assistance (13). This broad range of tasks fulfilled by rehabilitation professionals collaboratively can maintain and/or enhance QoL of the person with ALS. QoL has been defined as “the extent to which our hopes and ambitions are matched by experience” (12), thus refers to a person's satisfaction with the way his/her life is progressing, despite the diagnosis of a disease such as ALS, and with the care measures being taken (12). Although there are many other factors that can affect a person's outlook on his/her life after diagnosis, this study focuses specifically on those related to rehabilitation. This review specifically addresses interventions from PT, OT, and SLP, and multidisciplinary care that includes one or all of these therapies to examine the impact on QoL in people with ALS. Our research question was: In individuals with ALS, what interventions within the scope of rehabilitation are effective for improving QoL?

## **II. METHODS**

### **A. Search Strategy**

The search was conducted by author AYS in March 2015 with the following restrictions: English articles only, dated from 1980 to March 2015, and studies with human participants. We chose 1980 because Mikulic, DeLisa, and Miller (33) proposed the idea of using rehabilitation for people with ALS in 1979. The search was conducted using the PICO approach (Appendix 1) in consultation with a health sciences librarian and included the following databases: MEDLINE; CINAHL; PsycINFO; The Physiotherapy Evidence database (PEDro); Occupational Therapy Systematic Evaluation of Evidence (OT seeker); AMED; EMBASE; OVID Health Star; Global Health; and LILACS EN. The main search terms included *amyotrophic lateral sclerosis, motor neurone disease, motor neuron disease, physiotherapy, physical therapy, occupational therapy, speech language pathology, rehabilitation, therapy, and quality of life*.

### **B. Types of Studies**

The authors searched for all quantitative study designs that included interventions delivered by PT, OT and/or SLP for people with ALS. Inclusion criteria: people with a diagnosis of ALS (duration and stage of the disease did not have to be reported); rehabilitation intervention(s) (Appendix 3) delivered by at least one of the disciplines of PT, OT, SLP, or delivered by another person or discipline and assigned by one of these 3 rehabilitation professionals; QoL outcomes (see below) and quantitative and/or mixed-method designs (i.e. randomized control trials (RCTs), cohort, cross-sectional, and case-control studies). We included studies if part of a sample included people with ALS and met our other inclusion criteria.

## **Outcome Measures**

The primary outcome of this study was quality of life (QoL) as measured by the 36 item Short-Form Survey (SF-36) questionnaire (14) or the Amyotrophic Lateral Sclerosis Assessment Questionnaire (ALSAQ-40) (15). In the SF-36 questionnaire (14), there are eight sections; vitality, physical functioning, bodily pain, general health perceptions, physical role functioning, emotional role functioning, social role functioning, and mental health. In this measure higher scores reflect higher QoL (i.e., a score of zero is equivalent to poor QoL and a score of 100 is equivalent to excellent QoL).

The ALSAQ-40 has 40 questions specific to the ALS population with 5 discrete scales: physical mobility, activities of daily living and independence, eating and drinking, communication, and emotional reactions. Individuals with ALS are asked questions about the difficulties experienced during the last two weeks (e.g. I have fallen over while walking). The responses are collected by selecting one of five options (Likert scale): never/rarely/sometimes/often/always or cannot do at all (15). The responses are reported and collected where a higher number of responses in the “never/rarely/sometimes” section indicates a better QoL and, subsequently, responses in the “often/always or cannot do at all” correlates to a lower QoL. Thus, in contrast to the SF-36, higher scores on the ALSAQ – 40 indicate lower QoL.

## **C. Data Extraction and Quality Appraisal**

### *a. Selection of Studies*

Two review authors (AYS and LL) independently screened titles and abstracts of all citations. All potentially relevant articles selected by either author were obtained as full-text

articles, assessed for inclusion and exclusion criteria, and documented reasons for exclusion.

Where there were differences between reviewers, a discussion occurred to arrive at consensus.

Kappa was calculated to determine the level of inter-rater agreement (16). Once the studies were identified, the primary author (AYS) used a data extraction form (Appendix 3) to describe the study characteristics, study participants, and conclusions. Data extracted provided information about the study's purpose, ethical approval, methodology, outcome measures, and results. All findings were compiled in a single chart.

#### *b. Quality Appraisal and Assessment of Risk of Bias*

Two review authors (AYS and LL) independently evaluated the quality of the reporting of RCTs using the CONSORT Checklist (19) and risk of bias using the Cochrane Risk of bias tool (20). We assessed risk of bias as “high”, “low” or “unclear”. The STROBE Statement (21) and the New-Castle Ottawa scale (22) were used to assess quality of reporting and risk of bias in the non-RCT studies. In the case of disagreement between the two authors, each reassessed the studies and agreement was reached through discussion.

#### *Statistical Analysis*

A meta-analysis on the data was planned should data allow, as well as the generation of forest plots to illustrate the estimates and confidence intervals for each study. We would pool data by QoL outcome if the studies were clinically (e.g. similar intervention) and statistically (e.g. all SF-36 results reported by the mean of each group) homogeneous. This method would follow that described in The Cochrane Handbook for Systematic Reviews of Interventions (17). The authors contacted included studies' authors for data if required.



### III. RESULTS

After removing duplicates, the search yielded 276 articles, and 7 articles of 6 unique studies were initially included (Figure 1); however, one article was an abstract (25) and it was removed as it was a preliminary data report of another study (26), resulting in a total of 6 unique articles and studies. Three studies were RCTs (24, 26, 27), two were cohort studies (29, 30), and one was a cross-sectional study (28). Five studies reported SF-36 (24, 26, 28, 29, 30) and one study reported ALSAQ-40 (27). There were no disagreements between the authors for included studies; using Cohen's Kappa (16), the level of agreement was calculated as 1, representing perfect agreement.

Following contact with study authors for additional data, we did not conduct a meta-analysis as the nature of the studies was not sufficiently similar and thus the data was not presented with the same variables from the outcome measures. Thus, we conducted a narrative synthesis of the interventions. Table 1 presents a summary of all included studies, including QoL results. Table 2 presents the evaluation of the quality of the studies based on the CONSORT and STROBE criteria (Appendix 2), where a higher percentage in total score is interpreted as higher quality of reporting. Table 3 presents the results of the risk of bias assessments.

#### **A. Description of Studies and Effect of Rehabilitation Interventions on QoL using SF-36 and ALSAQ-40**

##### SF-36

Of the three RCTs enrolling 25, 27, and 132 ALS patients respectively (24, 26, 27), two (24, 26) focused on exercise and one (27) on multidisciplinary ALS care with case management. Two studies (24, 26) observed a decline in QoL using the SF-36 (Table 1). Drory et al. (24)

compared people that performed exercise to those whose physical activities only included activities of daily living (ADLs). The exercise program targeted muscle endurance and resistance against a modest load for people with ALS (24). This study had no statistically significant differences in QoL between the two groups (Control Group Mean = 81.2, Exercise Group Mean = 72.7). Dal Bello-Haas et al. (26) found that at 6 months, the resistance exercise group presented a significantly higher QoL (SF-36) score than the control group (stretching exercise). The resistance exercise group were given a stretching exercise program with an individualized upper (U/E) and lower extremity (L/E) resistance exercise program with moderate load and intensity. The control group received a program that consisted of daily U/E and L/E stretching exercises to be performed once a day. The studies were mainly limited by sample sizes, brief duration of intervention, and drop-outs thus affecting the statistical power of results.

Interventions delivered by SLPs were addressed in one study (29). Korner et al. (29) conducted a cohort study of patients with ALS who received speech therapy alone or speech therapy combined with communication devices. The response rate was 100%, and included 38 participants. This study found that speech therapy with communication devices positively affected QoL, however there were no significant between group differences on any component of the SF-36. Nevertheless, the study found that communication devices and speech therapy appeared to stabilize QoL in people with ALS.

Jensen et al. (30) conducted a survey the nature of pain and pain treatments (including rehabilitation interventions such as muscle strengthening exercises) used for people with neuromuscular disease, including those with ALS. The response rate was 30% from people with ALS and 86% response rate overall. Of 193 participants, 30 had ALS. Across all the neuromuscular conditions, physiotherapeutic approaches were reported to provide the greatest

pain relief, however this did not show an impact on the QoL score measured using the SF-36 questionnaire. The results of this study were reported for the whole sample, limiting the ability to identify the importance of physiotherapy for pain management in people with ALS. However, the authors noted that people with ALS appeared to be the most sensitive to the effects of pain, thus potentially influencing QoL. Therefore, the study was unable to link the effectiveness of physiotherapy interventions for pain to improved QoL.

The cross-sectional study performed by Van den Berg et al. (28) assessed the effect of multidisciplinary ALS care from a team and according to the Dutch consensus protocol for rehabilitation management in ALS compared to general care. The ALS team was led by a consultant in rehabilitation medicine and included a PT, an OT, an SLP, a dietician, or a social worker. . This study found better SF-36 mental-QoL among people who received multidisciplinary care compared to those who only received general care; however this was not statistically significant in the multivariable analysis. Yet in the multivariate analysis performed by Van den Berg et al. (28) to determine the factors that contributed to changes in mental-QoL, it remains unknown which factors contributed to improved mental-QoL. Despite the discussion sections of the studies reporting the positive influence of rehabilitation services to impact QoL, use of the SF-36 questionnaire typically resulted in non-statistically significant results comparing outcomes between groups.

#### ALSAQ-40

Creemers et al. (27) conducted a cluster RCT of multidisciplinary interventions in 132 people with ALS to establish the effect of case management for patients and caregivers compared to usual multidisciplinary ALS care. The study reported no benefit of case

management to improve QoL measured by the ALSAQ-40, unless people with ALS were struggling with using and/or accessing rehabilitation interventions or other care services. This study reported that during the intervention period, people with ALS reported one issue related to a rehabilitation intervention whereas other reasons for seeing a case manager was for emotional support. Despite this noticeable trend among the participants, the emotional function scores on the ALSAQ-40 measure had no significant difference than the usual multidisciplinary ALS care group.

### **B. Quality of the Evidence**

We assessed the quality of evidence using the CONSORT statement, STROBE statement, and the Cochrane Risk of Bias Tool as appropriate to each study design. The evidence base obtained to address the research question of this study resulted in the overall quality of evidence where five of the six included studies had overall quality percentages above 75% (Table 2). The study that scored less than 75% (29) was due to not meeting certain criteria from the results section i.e. information was not provided regarding reasons for participants not participating at each stage, participant characteristics were incomplete, and additional details were missing to describe the study methods.

From the Cochrane Risk of bias assessments, the authors determined that the Drory et al. (24) study was at unclear risk of bias, and that the Dal Bello-Haas et al. (26) and Creemers et al. (27) studies were at low risk of bias. In two studies (26, 27), there were no baseline differences between participant groups in regards to age and disease duration. In Drory et al. (24), the institution statistician performed the randomization using computer software, Dal Bello-Haas et al. (26) reported that opaque envelopes with group allocation was used, and the Creemers et al. (27) reported that all ALS teams were invited to participate in the study then teams were matched

based on three factors. In two studies (24, 26), allocation concealment was not done, as it would be difficult to blind participants to the exercise intervention, however in the cluster RCT (27) allocation concealment was performed at the cluster level. The outcome evaluator was not blinded to group allocation in Drory et al. (24); however in Dal Bello-Haas et al. (26) and Creemers et al. (27) 2007, the evaluators were blinded to group allocation.

For non-RCT studies, the two cohort designs (29, 30) were identified as low and unclear risk of bias respectively and the cross-sectional study (28) was low risk of bias. This was concluded based on participants, outcomes, comparing baseline characteristics, and detection of reporting and/or attrition biases (Table 3).

#### **IV. DISCUSSION**

Currently there is little evidence quantifying positive and sustained effects of interventions from PT, OT and SLP on QoL in the ALS population. Across 5 studies, exercise and pain interventions by physiotherapists, communication devices and speech therapy and assistive aids have shown potential to impact functional outcomes in the management of people with ALS. However, only one study reported a statistically significant improvement in QoL, measured by the SF-36 (26). The summary of quality of reporting the evidence (Table 2) suggests a reasonable quality as most studies were above 70%. Additionally, the assessed risk of bias from the included studies were mainly low (26, 27, 29, 30) and unclear (24, 28) risk of bias presenting a high quality of evidence overall. Nevertheless, the quantity is limited.

Drory et al. (24) and Dal Bello-Haas et al. (26) have both demonstrated that exercise, when appropriately prescribed and performed, can positively affect physical function which may in turn impact QoL, particularly when individuals are still in the early stages of the disease. In

fact, Dal Bello-Haas et al. (26) found QoL to be significantly higher in the exercise group at 6 months, compared to a control group who completed usual activities. Even though exercise does not have the ability to improve strength of the muscles weakened by the condition, moderate regular exercises may have positive effects and can play an important part in the disease management. The lack of strong evidence regarding the effectiveness and benefits of exercise on QoL in the ALS population may influence the availability and quality of rehabilitation interventions provided (5), and thus suggests a need for more research in this area.

Our included studies focused on interventions and/or the influence of PT and SLP in managing people with ALS (24, 26, 27, 28, & 29), rather than QoL. Such have shown that PT improves individual muscle strength and decreases fatigue and spasticity (add references here too). Augmentative and alternative communication (AAC) devices are shown to be helpful among individuals with ALS as there is an emotional aspect associated with communication that can affect one's QoL (10). Although there is some overlap in the interventions provided by professionals from PT, OT, and SLP, the studies performed to date do not specifically address the impact of interventions by OTs in people with ALS. For example, assistive aids and appliances are often used in the management of ALS and can be recommended by both PTs and OTs (e.g., walkers, seating and positioning techniques, wheelchairs etc. (10)). Moreover, OT interventions can also be helpful for individuals with ALS in regards to employment, as engaging in activities that are essential to one's health and well-being. Yet, there is a lack of evidence establishing the impact such rehabilitation interventions have on a person's QoL, suggesting a need for further research.

Based on the results of this review, we identified few studies from the PT, OT and SLP professions to conduct controlled experiments that measured QoL outcomes of therapy

interventions for people with ALS. We believe this may partly be due to the nature of ALS e.g., heterogeneity and rate of disease progression in the ALS population. Researchers who have performed RCTs in this area have consistently faced the challenge of high drop-out rates due to the quick progression of the disease (24, 26, 27). Furthering ALS research through a longitudinal cohort study to examine which rehabilitation interventions have the potential to impact QoL would be beneficial. The literature available regarding the effectiveness of rehabilitation interventions is limited; however, there is growing evidence supporting the importance of multidisciplinary care for the management of ALS.

The strengths of this systematic review include a defined process and structured critical appraisal. The possibility of rejecting relevant studies was reduced by having two authors review all studies for inclusion using criteria agreed upon prior to extracting the data and reporting the quality of included studies. The quality of the studies was assessed using the CONSORT Checklist (19), Cochrane Risk of bias tool (20), Newcastle-Ottawa Scale (22), and the STROBE Statement (21), thus allowing simple comparison of the results to identify study quality. Additionally, this study adds to the evidence by appraising the available literature to date and underlining the potential of rehabilitation services from PT, OT and SLP to maintain QoL when managed by a multidisciplinary team of specialists in rehabilitation.

## **V. LIMITATIONS**

Research in ALS related specifically to PT, OT and SLP interventions is limited, and thus this systematic review is limited to a small number of studies, which had to be analysed descriptively. In addition, bias may have been introduced by restricting the review to only studies published in English.

## **VI. CONCLUSION**

The overall finding of this narrative review supports interventions from the professions of PT, OT and SLP have the potential to impact QoL in people with ALS. It is difficult to determine the relative effects of rehabilitation interventions due to the disease progression in ALS, especially when the rate of progression differs across individuals, and evidence associated with rehabilitation and stage of ALS is very limited. Rehabilitation interventions can affect QoL, and offering interventions that cater to the needs of people with ALS could contribute to the maintenance of QoL. Future research should be directed toward developing an approach to measure the effectiveness of different rehabilitation interventions in people with ALS and evaluating short- and long-term effects of impairments on QoL.



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<b>Table 1: Details of Included Studies</b>						
Author & Publication year, study design, and total sample size (evaluated for QoL/ enrolled)	Purpose of the study	Intervention (evaluated/ enrolled)	Control (evaluated/ enrolled)	Outcome measures (QoL measure in bold)	Overall Study results	QoL Results
Drory et al., 2001 Randomized Controlled Trial; N = 14/25 (males) <b>Moderate daily exercise (DE) vs. No exercise or physical activity other than daily life activities (PA)</b>	To determine the effect moderate exercise under professional guidance have on motor deficit, disability, fatigue, MSK pain and QoL	Daily exercise N = 8/14 (males)	Daily activities N = 6/11 (males)	MMT, Ashworth spasticity scale, ALSFRS, FSS, VAS, <b>SF-36</b>	QoL deteriorated slightly with a non-significant trend towards the control group	<b>Mean at Baseline:</b> PA – 84.1 DE – 82.9 <b>Mean at 3months:</b> PA – 83.0 DE – 80.4 <b>Mean at 6 months:</b> PA – 81.2 DE – 72.7
Dal Bello-Haas et al., 2007 Randomized Control Trial; N = 27 <b>Resistance exercise (RE) vs. Stretching exercise (SE)</b>	To determine the effects of resistance exercise on function, fatigue, and QoL	Resistance exercise = 13	Stretching group = 14	ALSFRS, FSS, <b>SF-36</b> , Muscle strength (MVIC)	No adverse effects occurred with relation to intervention and decline in leg strength was lower in resistance exercise group	<b>3 months:</b> No difference between groups (SE and RE) <b>6 months:</b> RE significantly higher than SE t = 2.58 df = 16 p = 0.02
Creemers et al., 2013 Cluster Randomized	To determine the effect of case management on QoL, caregiver strain, and	Case management and usual care ALS patients N =	Usual care ALS patients N = 61, Caregivers N = 60)	ALSFRS-R, <b>ALSAQ-40</b> , CSI, QOC	Introducing case management for people with ALS showed	Mean (SD), n (Emotional Functioning) – <b>Baseline MDC + CS:</b> 21.3 (18.2), 70

<p>Controlled Trial; N = 298 <b>Multidisciplinary ALS care with case management (MDC+CS) vs. Usual multidisciplinary ALS care (MDC)</b></p>	<p>perceived quality of care (QOC) in patients with ALS and their carers</p>	<p>71, Caregivers N = 66)</p>			<p>no benefit in QoL scores.</p>	<p><b>Baseline MDC:</b> 19.4 (17.0), 61 <b>p-value:</b> 0.524</p> <p><b>4 months MDC + CS:</b> 19.8 (14.6), 43 <b>4 months MDC:</b> 19.4 (16.6), 39 <b>p-value:</b> 0.35</p> <p><b>8 months MDC + CS:</b> 21.5 (13.5), 28 <b>8 months MDC:</b> 20.0 (16.1), 22 <b>p-value:</b> 0.33</p> <p><b>12 months MDC + CS:</b> 22.8 (16.4), 30 <b>12 months MDC:</b> 19.1 (14.7), 27 <b>p-value:</b> 0.68</p>
<p>Jensen et al., 2005 Survey study N = 193 ALS = 30 Physiotherapy consisting of a variety of interventions (e.g. passive range of motion and</p>	<p>To examine the nature and scope of pain in persons with neuromuscular disorder (NMD)</p>	<p>(not applicable)</p>	<p>No comparison group</p>	<p>Neuropathic pain scale (NPS), Brief Pain Inventory (BPI), <b>SF-36</b></p>	<p>Pain appeared to be particularly severe in people with ALS affecting their QoL significantly. However, no response from contacted</p>	<p><b>Physical Functioning Scale (Past 3 months – one time measure):</b> Physical functioning – 50.41 (15.98) Role functioning-physical – 62.28 (17.77) General health perception – 58.76 (20.37) Body pain – 51.50 (15.74)</p>

<p>stretching, massage, specific muscle strengthening exercises)</p>					<p>author regarding ALS-specific data</p>	<p><b>Mental Health (Past 3 months – one time measure):</b>                  Vitality – 47.95 (17.10)                  Social functioning – 67.12 (20.93)                  Role functional-emotional – 79.01 (21.44)                  Mental health – 73.23 (14.94)</p>
<p>Korner et al., 2013 Cohort study; N = 38 (26 females, 12 males) Speech therapy and communication devices</p>	<p>To investigate and compare the impact speech therapy and communication devices have on QoL and mood in ALS patients</p>	<p>Speech therapy with communication device(s) N = 4/16 (males)</p>	<p>Speech therapy only N = 8/22 (males)</p>	<p>Beck Depression Inventory (BDI), SF-36, ALSFRS-R</p>	<p>Patients described a high impact of the communication device on QoL yet the impact of speech therapy was rated lower. Additionally, multiple regression analysis confirmed that an independent positive effect of communication device use on depression</p>	<p><b>Speech Therapy Only (S); Speech Therapy + Additional Communication Device (S+C)</b></p> <p><b>Physical Functioning Scale:</b>                  Physical functioning – S: 33.18                  S + C: 35.38</p> <p>Role functioning-physical – S: 31.26                  S + C: 28.97</p> <p>General health perception – 34.84                  Body pain – 70.24</p> <p><b>Mental Health:</b>                  Vitality – 38.99</p>



					and mental QoL.  Time point not mentioned, however data was collected post-intervention.	Social functioning – 51.90 Role functional-emotional – 47.51 Mental health – 57.28  General health perception – 41.31 Body pain – 63.00  <b>Mental Health:</b> Vitality – 33.21 Social functioning – 51.85 Role functional-emotional – 41.39 Mental health – 54.98
Van den Berg et al., 2005 Cross-sectional study N = 208 Multidisciplinary ALS care vs. Non- specialized general ALS care	To examine the effect of multidisciplinary care (MDC) on QoL in ALS patients and their carers	Multidisciplinary ALS care N = 133	General ALS care N = 75	<b>SF-36, VAS</b>	Providing people with ALS a high standard of care improves their mental-QoL.	<b>Adjusted difference, 95% CI, p-value</b>  <b>Physical Functioning Scale (data collected once, time point not mentioned):</b> Physical summary scale – (-1.22), -4.2 to 1.7, 0.42 Physical functioning – (-0.63), -8.6 to 7.4, 0.88 Role functioning-physical – (-1.1), -11.5 to 9.3, 0.83 General health perception – (0.94), -5.2 to 7.0, 0.76 Body pain – (-2.49), -10.9 to 5.9, 0.56

						<p><b>Mental Health (data collected once, time point not mentioned):</b>                  Mental summary scale – (4.28), 1.2 to 7.4, 0.01                  Vitality – (4.02), -2.2 to 10.3, 0.21                  Social functioning – (15.0), 6.8 to 23.3, &lt;0.001                  Role functional-emotional – (5.1), -7.7 to 18.0, 0.43                  Mental health – (4.5), -0.2 to 9.2, 0.06</p>
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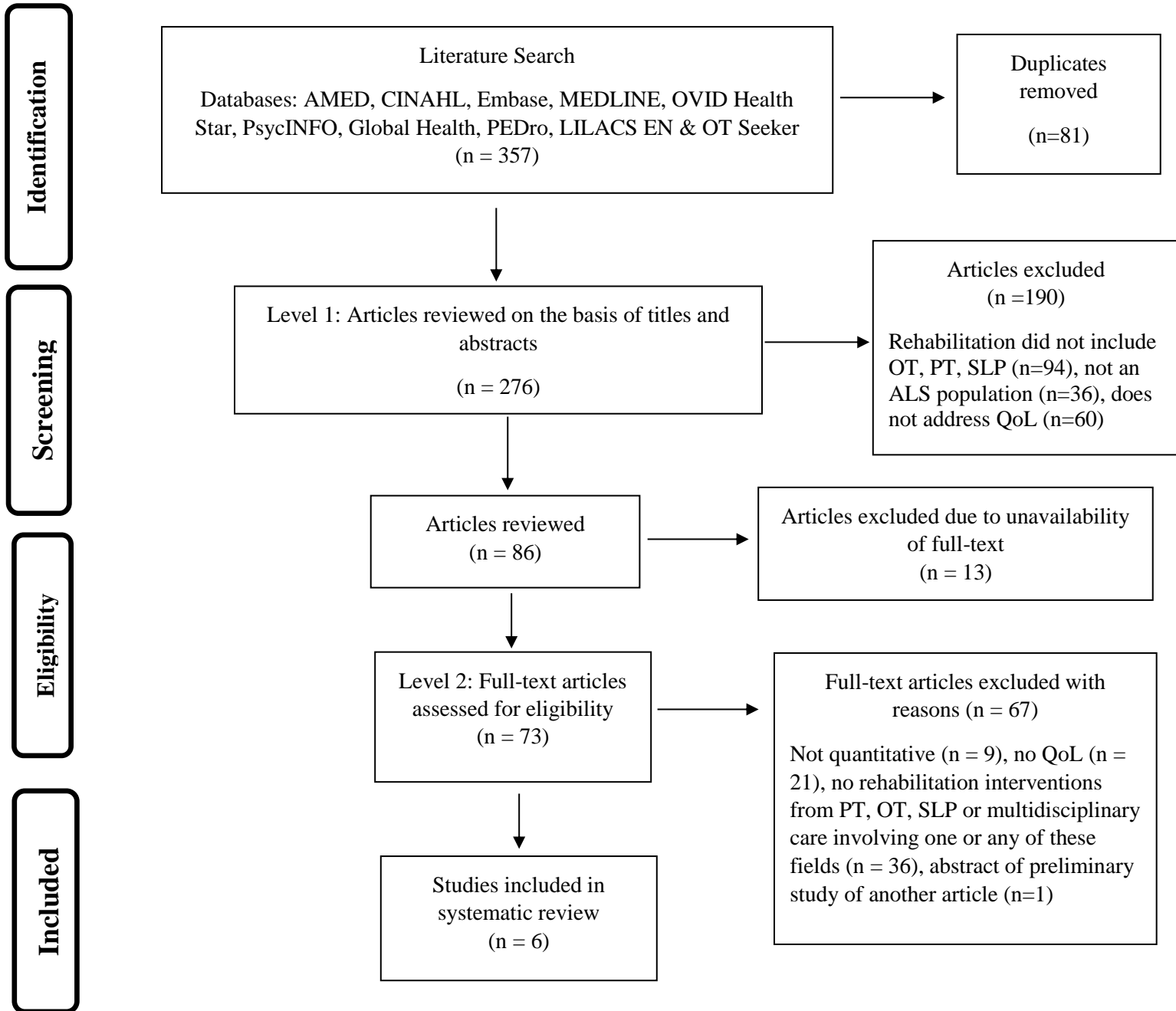
**Abbreviations:** MMT – Manual Muscle Testing; ALSFRS – Amyotrophic lateral sclerosis Functional Rating Scale; FSS – Fatigue Severity Scale; VAS – Visual Analogue Scale; SF-36 – Short Form-36; Muscle strength MVIC - Maximum voluntary isometric contraction; ALSAQ-40 – Amyotrophic lateral sclerosis Assessment Questionnaire – 40; CSI – Caregiver Strain Index; QOC – Quality of care; NPS- Neuropathic pain scale;BPI – Brief Pain Inventory; BDI – Beck Depression Inventory; MDC – Multidisciplinary care; CM – case management; RE – resistance exercise; SE – stretching exercise; DE – moderate daily exercise (DE); PA – no exercise or physical activity other than daily life activities

<b>Table 2: Study reporting quality of the included studies based on CONSORT (Studies 1-3) and STROBE criteria (Studies 4-6)</b>										
Author and year of publication	Title & Abstract	Introduction	Methods	Results	Discussion	Other information	Score obtained	Total score	% of Total score	
	<b>CONSORT</b>	<b>1</b>	<b>1</b>	<b>10</b>	<b>7</b>	<b>3</b>	<b>3</b>			
1.	Drory et al., 2001	0.5	1	7	6	3	2	19.5	25	78%
2.	Dal Bello-Haas et al., 2007	1	1	8	6	3	3	22	25	88%
3.	Creemers et al., 2013	1	1	8	7	2	3	22	25	88%
	<b>STROBE</b>	<b>1</b>	<b>2</b>	<b>9</b>	<b>5</b>	<b>4</b>	<b>1</b>			
4.	Jensen et al., 2005	1	2	6	3	3.5	1	16.5	22	75%
5.	Korner et al., 2013	0.5	2	7	3	3	0	15.5	22	70%
6.	Van den Berg et al., 2005	1	2	7	4	4	1	19	22	86%

<b>Table 3: Results of Risk of Bias Evaluation</b>						
Author & Publication year	Random sequence generation (selection bias)	Allocation concealment (selection bias)	Blinding (performance bias and detection bias) All outcomes	Incomplete outcome data (attrition bias) All outcomes	Selective reporting (reporting bias)	Overall Risk of Bias
Drory et al., 2001	Low risk- The institution statistician performed the randomization using computer software	Unclear risk – not reported	Unclear risk – blinding not reported	High risk - Both groups lost a lot of people – reasons for drop outs not reported – numbers are	Low risk - All outcomes reported	Unclear risk
Dal Bello-Haas et al., 2007	Low risk – random sequence reported	Low risk - Opaque envelopes with group allocation used; note that participants were not blind to group allocation and some revealed their allocation to therapist collecting data	Low risk - Participants completing intervention were unblinded; outcome evaluators were blinded to group allocation	Low risk - Participants were lost to follow up in both groups – reasons clearly stated and missing data were imputed for analyses.	Low risk - All outcomes reported	Low risk
Creemers et al., 2013	Low risk - All ALS teams were invited to participate; teams were matched	Low risk - Allocation concealment was at the cluster level	Low risk - blinded outcome evaluators at all follow-up times	Low risk - Although they lost quite a few participants from both groups, the losses are explained and many were because of death.	Low risk - All outcomes reported	Low risk

Jensen et al., 2005 Note: Data presented does not clearly identify people with ALS	Low risk – People with neurodegenerative conditions were allowed to complete the survey	Low risk – Limited restrictions on who could participate on the survey	Low risk - No outcomes based on performance, all self-report measures	Low risk – Onetime survey therefore no attrition	Low risk – data on all outcomes reported	Low risk
Korner et al., 2013	Low risk – Participant selection mentioned	Low risk – Groups assigned based on allocation	Low risk – all outcome measures are self-report	Low risk – data collected when patients visited the clinic post-intervention	Unclear risk - Can't identify dependent and independent variables in regression	Low risk
Van den Berg et al., 2005	Unclear risk - No random selection and not clear how patients were recruited or selected	Unclear risk – not clear how allocations were determined	Low risk – nurses collecting outcome data blinded to treatment group	Unclear risk – no clear description of dropouts nor if patients were followed over time	Low risk – all outcome measures reported	Unclear risk

**Figure 1: PRISMA flow diagram for study selection in Systematic Review**



**Appendix 1: Keywords and Medical Subject Headings (MeSH) terms****Group 1: Population**

42. Amyotrophic lateral sclerosis (MeSH exploded not focused) (keyword)
43. ALS search as keyword
44. motor neuron disease
45. motor neurone disease
46. Charcot disease (keyword)
47. Lou Gehrig's disease (keyword)
48. Combo of keywords and subject
49. Group 1 or 2 or 3 or 4 or 5 or 6 → 7

**Group 2: Intervention**

50. Rehabilitation (explode not focused) (keyword)
51. Therapy
52. Physiotherapy
53. Physical therapy
54. Occupational therapy
55. speech and language pathology/therapy
56. combo of keyword and subject
57. Group 8 or 9 or 10 or 11 or 12 or 13 → 14

**15. Combine 7 and 14****16. ALS – MeSH + rh, th [Rehabilitation, Therapy]****17. Combine 15 or 16****Group 3: Outcome**

58. Quality of life
59. QoL
60. disease management
61. functional status/ ability
62. physical endurance
63. health status
64. activities of daily living
65. health level
66. psychiatric status
67. combo of keyword and subject
68. Group 18 or 19 or 20 or 21 or 22 or 23 or 24 or 25 → 26

**26. Combine 17 and 26**

Add limitations Humans, 1980 – Current, English

## Appendix 2: CONSORT & STROBE Statements



### CONSORT 2010 checklist of information to include when reporting a randomised trial\*

Section/Topic	Item No	Checklist item
<b>Title and abstract</b>	1a	Identification as a randomised trial in the title
	1b	Structured summary of trial design, methods, results, and conclusions (for specific guidance see CONSORT for abstracts)
<b>Introduction</b>		
Background and objectives	2a	Scientific background and explanation of rationale
	2b	Specific objectives or hypotheses
<b>Methods</b>		
Trial design	3a	Description of trial design (such as parallel, factorial) including allocation ratio
	3b	Important changes to methods after trial commencement (such as eligibility criteria), with reasons
Participants	4a	Eligibility criteria for participants
	4b	Settings and locations where the data were collected
Interventions	5	The interventions for each group with sufficient details to allow replication, including how and when they were actually administered
Outcomes	6a	Completely defined pre-specified primary and secondary outcome measures, including how and when they were assessed
	6b	Any changes to trial outcomes after the trial commenced, with reasons
Sample size	7a	How sample size was determined
	7b	When applicable, explanation of any interim analyses and stopping guidelines
Randomisation:		
Sequence	8a	Method used to generate the random allocation sequence
generation	8b	Type of randomisation; details of any restriction (such as blocking and block size)



Allocation concealment mechanism	9	Mechanism used to implement the random allocation sequence (such as sequentially numbered containers), describing any steps taken to conceal the sequence until interventions were assigned
Implementation	10	Who generated the random allocation sequence, who enrolled participants, and who assigned participants to interventions
Blinding	11a	If done, who was blinded after assignment to interventions (for example, participants, care providers, those assessing outcomes) and how
	11b	If relevant, description of the similarity of interventions
Statistical methods	12a	Statistical methods used to compare groups for primary and secondary outcomes
	12b	Methods for additional analyses, such as subgroup analyses and adjusted analyses
<b>Results</b>		
Participant flow (a diagram is strongly recommended)	13a	For each group, the numbers of participants who were randomly assigned, received intended treatment, and were analysed for the primary outcome
	13b	For each group, losses and exclusions after randomisation, together with reasons
Recruitment	14a	Dates defining the periods of recruitment and follow-up
	14b	Why the trial ended or was stopped
Baseline data	15	A table showing baseline demographic and clinical characteristics for each group
Numbers analysed	16	For each group, number of participants (denominator) included in each analysis and whether the analysis was by original assigned groups
Outcomes and estimation	17a	For each primary and secondary outcome, results for each group, and the estimated effect size and its precision (such as 95% confidence interval)
	17b	For binary outcomes, presentation of both absolute and relative effect sizes is recommended
Ancillary analyses	18	Results of any other analyses performed, including subgroup analyses and adjusted analyses, distinguishing pre-specified from exploratory
Harms	19	All important harms or unintended effects in each group (for specific guidance see CONSORT for harms)
<b>Discussion</b>	20	Trial limitations, addressing sources of potential bias, imprecision, and, if relevant, multiplicity of analyses
Limitations		
Generalisability	21	Generalisability (external validity, applicability) of the trial findings
Interpretation	22	Interpretation consistent with results, balancing benefits and harms, and considering other relevant evidence
<b>Other information</b>		

Registration	23	Registration number and name of trial registry
Protocol	24	Where the full trial protocol can be accessed, if available
Funding	25	Sources of funding and other support (such as supply of drugs), role of funders

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\*We strongly recommend reading this statement in conjunction with the CONSORT 2010 Explanation and Elaboration for important clarifications on all the items. If relevant, we also recommend reading CONSORT extensions for cluster randomised trials, non-inferiority and equivalence trials, non-pharmacological treatments, herbal interventions, and pragmatic trials. Additional extensions are forthcoming: for those and for up to date references relevant to this checklist, see [www.consort-statement.org](http://www.consort-statement.org).

STROBE Statement—checklist of items that should be included in reports of observational studies

	<b>Item No</b>	<b>Recommendation</b>
<b>Title and abstract</b>	1	(a) Indicate the study's design with a commonly used term in the title or the abstract (b) Provide in the abstract an informative and balanced summary of what was done and what was found
<b>Introduction</b>		
Background/rationale	2	Explain the scientific background and rationale for the investigation being reported
Objectives	3	State specific objectives, including any pre-specified hypotheses
<b>Methods</b>		
Study design	4	Present key elements of study design early in the paper
Setting	5	Describe the setting, locations, and relevant dates, including periods of recruitment, exposure, follow-up, and data collection
Participants	6	(a) <i>Cohort study</i> —Give the eligibility criteria, and the sources and methods of selection of participants. Describe methods of follow-up <i>Case-control study</i> —Give the eligibility criteria, and the sources and methods of case ascertainment and control selection. Give the rationale for the choice of cases and controls <i>Cross-sectional study</i> —Give the eligibility criteria, and the sources and methods of selection of participants (b) <i>Cohort study</i> —For matched studies, give matching criteria and number of exposed and unexposed <i>Case-control study</i> —For matched studies, give matching criteria and the number of controls per case
Variables	7	Clearly define all outcomes, exposures, predictors, potential confounders, and effect modifiers. Give diagnostic criteria, if applicable
Data sources/ measurement	8*	For each variable of interest, give sources of data and details of methods of assessment (measurement). Describe comparability of assessment methods if there is more than one group
Bias	9	Describe any efforts to address potential sources of bias
Study size	10	Explain how the study size was arrived at

Quantitative variables	11	Explain how quantitative variables were handled in the analyses. If applicable, describe which groupings were chosen and why
Statistical methods	12	<p>(a) Describe all statistical methods, including those used to control for confounding</p> <p>(b) Describe any methods used to examine subgroups and interactions</p> <p>(c) Explain how missing data were addressed</p> <p>(d) <i>Cohort study</i>—If applicable, explain how loss to follow-up was addressed</p> <p><i>Case-control study</i>—If applicable, explain how matching of cases and controls was addressed</p> <p><i>Cross-sectional study</i>—If applicable, describe analytical methods taking account of sampling strategy</p> <p>(e) Describe any sensitivity analyses</p>
<b>Results</b>		
Participants	13*	<p>(a) Report numbers of individuals at each stage of study—eg numbers potentially eligible, examined for eligibility, confirmed eligible, included in the study, completing follow-up, and analysed</p> <p>(b) Give reasons for non-participation at each stage</p> <p>(c) Consider use of a flow diagram</p>
Descriptive data	14*	<p>(a) Give characteristics of study participants (eg demographic, clinical, social) and information on exposures and potential confounders</p> <p>(b) Indicate number of participants with missing data for each variable of interest</p> <p>(c) <i>Cohort study</i>—Summarise follow-up time (eg, average and total amount)</p>
Outcome data	15*	<p><i>Cohort study</i>—Report numbers of outcome events or summary measures over time</p> <p><i>Case-control study</i>—Report numbers in each exposure category, or summary measures of exposure</p> <p><i>Cross-sectional study</i>—Report numbers of outcome events or summary measures</p>
Main results	16	<p>(a) Give unadjusted estimates and, if applicable, confounder-adjusted estimates and their precision (eg, 95% confidence interval). Make clear which confounders were adjusted for and why they were included</p> <p>(b) Report category boundaries when continuous variables were categorized</p>

		(c) If relevant, consider translating estimates of relative risk into absolute risk for a meaningful time period
Other analyses	17	Report other analyses done—eg analyses of subgroups and interactions, and sensitivity analyses
<b>Discussion</b>		
Key results	18	Summarise key results with reference to study objectives
Limitations	19	Discuss limitations of the study, taking into account sources of potential bias or imprecision. Discuss both direction and magnitude of any potential bias
Interpretation	20	Give a cautious overall interpretation of results considering objectives, limitations, multiplicity of analyses, results from similar studies, and other relevant evidence
Generalisability	21	Discuss the generalisability (external validity) of the study results
<b>Other information</b>		
Funding	22	Give the source of funding and the role of the funders for the present study and, if applicable, for the original study on which the present article is based

\*Give information separately for cases and controls in case-control studies and, if applicable, for exposed and unexposed groups in cohort and cross-sectional studies.

**Note:** An Explanation and Elaboration article discusses each checklist item and gives methodological background and published examples of transparent reporting. The STROBE checklist is best used in conjunction with this article (freely available on the Web sites of PLoS Medicine at <http://www.plosmedicine.org/>, Annals of Internal Medicine at <http://www.annals.org/>, and Epidemiology at <http://www.epidem.com/>). Information on the STROBE Initiative is available at [www.strobe-statement.org](http://www.strobe-statement.org).

### Appendix 3: Data Extraction Form – Systematic Review

#### General Information

Title of study/ID	
Reference citation (e.g. CINAHL)	
Study author details (e.g. _____ et al., 2010)	
Publication type (e.g. RCT)	
Date form completed (dd/mm/yyyy)	
Name of person extracting data	

#### Study eligibility

Study Characteristics	Eligibility criteria		Eligibility criteria met?		
			Yes	No	Unclear
Type of study Circle one →	Quantitative	Mixed-methods	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
	Experimental study including randomized controlled trials (RCTs) or cluster-randomized trials (CRTs)		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
	Quasi-experimental studies including quasi-randomized trials, controlled before-after studies		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
	Observational studies including cohort, case-control and cross-sectional studies		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Participants	Adults (18 years and older) diagnosed with ALS		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
	Any stage of the disease → stage identified in study?		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>

Types of intervention	Provided by rehabilitation specialists through physiotherapy, speech and language therapy, and occupational therapy and/or the combination of interventions from these professions	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
	- PT	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
	- SPL	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
	- OT	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
	- Combination of PT/OT/SLP	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Types of comparison	Comparative studies (e.g. Clinic –based physiotherapy intervention vs. physiotherapist recommended home-based exercises)	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Types of outcome measures	- Quality of life measures	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
	- Functional status measures	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Results	Quantitative results of the impact of rehabilitation interventions on QoL	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
INCLUDE <input type="checkbox"/> EXCLUDE <input type="checkbox"/>				
Reason for exclusion				

**STOP HERE IF THE STUDY DOES NOT MEET INCLUSION CRITERIA**

**IF INCLUDED, THEN PROCEED TO THE NEXT PAGE AND/OR THE CRITICAL APPRAISAL FORM(S)**

**Characteristics of included studies****Participants**

	Description
Population description (from which study participants are drawn)	Male Age Female Age
Total no. of participants	
Ethnicity information (circle one) Yes No If yes, describe →	
Baseline information (e.g. time since diagnosis, stage of disease, treatments undergoing/undergone)	

**Methods& Results**

	Descriptions of the study
Study purpose	
Design	
Setting	
Duration of participation	
List measures used	



Ethical approval obtained for study	<input type="checkbox"/> Yes <input type="checkbox"/> No <input type="checkbox"/> Unclear	Withdrawals from study:
Statistical Results		

**Other information**

	Description
Key conclusions of reviewer	
References to other relevant studies	

## CHAPTER 4: DISCUSSION & CONCLUSIONS

## **Chapter Four: Discussion and Conclusions**

### **Overview of Thesis Paper Results**

Individuals who are diagnosed with amyotrophic lateral sclerosis (ALS) ultimately experience difficulties with many areas of function, including mobility, communication, activities of daily living, and instrumental activities of daily living, leisure and productivity roles, all of which have the potential to affect quality of life (QoL). Although ALS remains incurable and irreversible, rehabilitation could help individuals and their families adjust, manage and cope with the disease to maintain or improve QoL (Dal Bello-Haas, 2002; Mitsumoto & Del Bene, 2000). The signs and symptoms of ALS are wide-ranging and need to be carefully assessed and managed, as availability and/or the effectiveness of interventions and options may be limited with disease progression. Rehabilitation services for people with ALS, in particular occupational therapy (OT), physiotherapy (PT), and speech and language pathology (SLP), are the main focus of this thesis. This chapter summarizes and discusses the conclusions of the reviews of qualitative and quantitative evidence with the intent that health professionals, people with ALS, and their families and carers will better understand the potential influence rehabilitation services have on QoL.

This thesis consists of two manuscripts for submission to peer reviewed journals for publication. Chapter 2 was a qualitative meta-synthesis of five studies that explored the perceptions of people with ALS regarding rehabilitation interventions and their impact on QoL. This study identified four themes: the concept of control; adapting interventions to disease stage; struggles with interventions; and barriers between healthcare providers and patients. Overall, this study highlights that people with ALS perceive PT, OT, and SLP interventions to have potential

benefits in helping them manage their condition and enhance QoL. Chapter 3, was a systematic review of the effectiveness of rehabilitation interventions in maintaining or improving QoL in people with ALS. Six studies met inclusion criteria: three randomized controlled trials (RCTs), two cohort based surveys, and one cross-sectional study. Although some studies from the review were able to establish an improvement in QoL, data could not be pooled to conduct meta-analysis as there was clinical and statistical heterogeneity of the results. This review found that it was difficult to determine the relative effects of interventions from PT, OT, and SLP or combined interventions from these fields on improving QoL for people with ALS.

Strengths and limitations of each of the two reviews in this thesis can be identified. In Chapter 2, I used four levels of trustworthiness for qualitative evidence (Henderson & Rhealt, 2004) and identified four of the included studies as Level I evidence, and one study as Level II evidence. Although one study had limitations in the confirmability component due to no mention of being peer reviewed, overall there were no significant problems identified with the credibility, transferability, and dependability aspects of the included studies. Thus, the data extracted for the qualitative meta-synthesis came from a body of original evidence deemed trustworthy and relevant to answer the research question. However, none of the included studies were designed to specifically examine the impact of rehabilitation interventions on QoL or to determine if rehabilitation was important to maintaining and/or enhancing QoL. Thus, it was necessary to interpret findings from the studies to identify topics related to QoL and rehabilitation interventions. We defined patients' perceptions of QoL by occurrences which these individuals consider important through the course of their disease i.e. continuing employment, ability to communicate, sense of importance, and the highest level possible of independence.

In Chapter 3, the studies included in the systematic review provided a limited quantity of evidence, but with reasonable quality to support the objective of the study. The strengths of the review included two authors independently reviewing all studies for inclusion prior to data extraction, and a defined and structured critical appraisal. We assessed the reporting completeness of included studies using the CONSORT Checklist, and the STROBE Statement, and risk of bias in RCTs with Cochrane Risk of bias tool and in non-RCTs using New-Castle Ottawa Scale. These assessments ultimately resulted in a rigorous overview of reporting and of internal validity to interpret the findings and results. Talk about reporting here. Talk about risk of bias here. Talk about inconsistent results here. Despite being unable to definitively determine the relative effects of therapy, this systematic review highlights the potential of rehabilitation interventions to facilitate the maintenance of QoL.

Across both studies, we identified few rehabilitation studies performed in the ALS population, and none of our studies specifically focused on QoL outcomes. In the following section, we consider whether our literature searches identified all available evidence.

### **SPIDER and PICO Methodologies**

The methods used to conduct the literature searches were the SPIDER (Cooke, Smith & Booth, 2012) and PICO (Higgins & Green, 2013) strategies for the meta-synthesis (Chapter 2) and systematic review (Chapter 3), respectively. The term SPIDER stands for Sample, Phenomenon of Interest, Design, Evaluation, and Research type (Cooke et al., 2012). The term PICO stands Population, Intervention, Comparison, and Outcome (Higgins & Green, 2013). The intention of each type of strategy was to identify relevant studies to make the search of literature specific and straightforward.

SPIDER was first designed by Cooke et al. (2012) in response to encountering limitations of the PICO strategy to search for qualitative and mixed-methods studies. The components of the SPIDER tool were adapted from PICO with the intention of making it suitable for qualitative research. In the meta-synthesis conducted in this thesis, the SPIDER approach yielded many studies that were unrelated to the search objective. This caused a problem with specificity, resulting in hundreds of inappropriate hits. We excluded 383 of 506 potentially relevant articles at Level 1 (screening based on titles and abstracts) because the majority of the studies were not performed within an ALS population and/or were not qualitative studies. Based on the search strategy, it is not clear how these hits were obtained; however, it is noted that more hits resulted from SPIDER in Chapter 2 than the PICO search in Chapter 3.

Methley, Campbell, Chew-Graham, McNally & Cheraghi-Sohi (2014) reported that even though some qualitative keywords are necessary to identify qualitative evidence, including the word ‘qualitative’ and the type of inquiry (e.g., phenomenology), may be too restrictive because the word “qualitative” may not be listed by authors or even indexed as a keyword by the database searched. Given the unstandardized and inconsistent use of the qualitative index term, this might explain the broad range of studies identified by SPIDER in comparison to PICO.

In Chapter 3, the PICO search yielded 276 articles and 190 were excluded at Level 1. This was primarily due to ALS genetic or animal model studies, non-rehabilitation (e.g., not PT, OT, and SLP related) interventions, or ineligible study designs (e.g., case reports). Overall, the two searches yielded evidence of QoL in ALS. However, due to the limited amount of research available that combined ALS rehabilitation and QoL, the majority of the retrieved articles were inapplicable to answer the research questions of both reviews, resulting in limited evidence for the reviews. Due to the search strategy applied in the databases of the keywords and MeSH

headings to only specific terminology (e.g., amyotrophic lateral sclerosis instead of motor neuron(e) disease) and other variations could have possibly result in locating more precise results.

### **Impact of Thesis Papers**

The purpose of performing both a qualitative and a quantitative review was to have a comprehensive overview of the impact of rehabilitation on QoL from the perspectives of people with ALS. As ALS is currently incurable, and research exploring rehabilitation interventions with this population is limited, rehabilitation professionals, are left to respond to individual concerns and symptoms to help someone with ALS manage their condition. Across the two studies, one overarching theme was observed: the struggles of adjusting to the changing interventions as the disease worsened. The metasynthesis illustrated the frequent and challenging adjustments to interventions with disease progression. The results of the systematic review were inconsistent and we could not identify the benefits of PT, OT, and/or SLP or a combination of these interventions. Nevertheless, this study was able to conclude that there are potential benefits of these interventions in ALS however they were brief, likely because of the disease progression. Only one study (Dal Bello-Haas et al., 2007) detected a significant overall improvement in QoL resulting from rehabilitation interventions for people with ALS.

For people with ALS, the notion of control was a common underpinning in the study designs across both the qualitative and quantitative studies. Dal Bello-Haas et al. (2007) noted that participation in an exercise program may provide a greater sense of control over what people with ALS experience during the course of the disease. Korner et al. (2013) described that QoL was higher in people with communicative devices and recommended that such interventions be

introduced early in the disease process to lessen rejection or negativity towards the intervention. Ultimately, the use of such devices could lead to individuals feeling empowered and in control (Foley, Timonen, & Hardiman, 2014). Additionally, using these devices may enable people with ALS to preserve their functional independence to sustain and/or enhance QoL throughout all stages of the disease.

The two reviews complement each other by recognizing that expertise from PT, OT and SLP professionals can play a role in influencing QoL in people with ALS. The metasynthesis revealed that barriers between healthcare professionals and people with ALS existed when professionals do not possess adequate knowledge and understanding of the disease. Rehabilitation professionals are generally knowledgeable about their scope of practice (Ng, Kahn & Mathers, 2009) and are aware of suitable interventions for a person given his/her impairments. Healthcare professionals who have encountered the challenge of treating someone with ALS have identified the need to improve their knowledge about the disease to benefit these individuals (Elsworth et al., 2009). Although the metasynthesis only included x studies with multidisciplinary care, people with ALS receiving both OT and SLP interventions together highlighted the importance of these services in maintaining QoL (McNaughton, Light, & Groszyk, 2001; Hughes, Sinha, Higginson, Down, & Leigh, 2005).

Ideally, people with ALS should receive multidisciplinary care from a variety of health professionals who are skilled and knowledgeable in managing ALS and the challenges associated with the disease (Hardiman, 2007). However, multidisciplinary ALS care is not always accessible or even available to each person with ALS; this could be tied to the low prevalence of the disease, which may make such specialized care difficult to provide (Van den Berg et al., 2005). Therefore, if a person with ALS cannot receive multidisciplinary care, communication



among rehabilitation professionals, even among non-ALS specific professionals, would be key when making decisions to address concerns, offering advice, education, and/or providing rehabilitation interventions.

The deteriorating nature of ALS is a constant struggle for people with this disease that does not become easier over time, and providing people with ALS with rehabilitation services may ease some of the difficulty (Chapter 2 & 3). However, struggles with accessing services and interventions have led to frustration (Hughes et al., 2005). In order to address this issue, assigning people with ALS a case manager after diagnosis has been suggested (Morris et al., 2009). Interestingly, a cluster RCT (Creemers et al., 2014) reported that multidisciplinary case management had no benefit for people with ALS despite concerns regarding delivery of assistive products and technology, restrictions in activities and participation, symptom treatment, psychosocial problems, and preparation for palliative care. More research would need to be conducted before validating that case management is a solution for people with ALS to receive better access to information and assistance of services when required.

The two review approaches helped in identifying the potential benefits of rehabilitation from a qualitative and quantitative lens. According to Pluye and Hong (2014), qualitative synthesis helps to explain some results of the quantitative synthesis, where findings of qualitative studies are integrated using thematic analysis and interpretation of the results uncovers new explanations and reveals knowledge gaps from quantitative studies. Therefore, the purpose of using two different methods lies in the fact that each review is answering a distinct question and therefore each study has their own research method. If only one method was used, the study would be restricted to the study design and data collection tools available specifically for that research method and may not answer both questions as accurately as possible. The rationale for

using two distinct reviews rather than a mixed methods review was related to the challenges associated with the management and/or reduction of the data, e.g., combining the two types of data. Additionally, the compromises in sample size, questions regarding generalizability of the data, timing of the sampling, difficulty in compiling and analysis of data all contribute to the challenges to facilitate this process in order to make it more effective and efficient (Driscoll Appiah-Yeboah, Salib & Rupert, 2007). Within the context of the thesis, the two reviews were conducted as distinct reviews, with the advantage of integrating the findings in this final chapter.

It was challenging to arrive at conclusions regarding the relationship between rehabilitation and QoL, since no qualitative study directly explored this relationship, and data in the quantitative review did not focus on QoL but reported it as a secondary outcome measure. Interpretation from the available qualitative studies had to be drawn to explore the relationship between rehabilitation and QoL. From the 5 studies reporting SF-36 questionnaires, patients with ALS reported QoL scores in a very low range of possible values compared to population-based reference norms. These low scores occurred particularly in the physical function components, which is likely to occur due to the nature of the disease. Although the Short-Form 36 (SF-36) is a generic measure that does not target a specific age, disease, or treatment group, it is more widely used as it is simple to administer, complete and ultimately shows how the disease impacts a person's overall QoL physically and mentally (Ware, Snow, Kosinski, & Gandek, 1993). In contrast, the Amyotrophic lateral sclerosis Assessment Questionnaire (ALSAQ-40) is ALS-specific and is described as a measure of QoL, however the items seem to measure function predominantly (Jenkinson, Fitzpatrick, Swash & Jones, 2007). The ALSAQ-40 has 40 questions specific to the ALS population with 5 discrete scales: physical mobility, activities of daily living and independence, eating and drinking, communication, and emotional reactions. Individuals

with ALS are asked questions about the difficulties experienced during the last two weeks (e.g. I have fallen over while walking). The responses are collected by selecting one of five options (Likert scale): never/rarely/sometimes/often/always or cannot do at all (Jenkinson et al., 2007), where a higher number of responses in the “never/rarely/sometimes” section indicates a better QoL and, subsequently, responses in the “often/always or cannot do at all” correlates to a lower QoL and thus, higher scores indicate lower QoL.

### **Future Research**

Due to the deteriorating nature of ALS, performing RCTs to evaluate the effectiveness of rehabilitation interventions that require significant time to deliver the intervention and follow-up may not be feasible with this population. People with ALS would struggle to remain enrolled and participate in rehabilitation through the entire duration of the study, resulting in high drop-out rates as seen in the included studies of the systematic review (Drory, Goltsman, Goldman Reznik, Mosek, & Korczyn, 2001; Dal-Bello Haas et al., 2007; & Creemers et al., 2013). The ability of rehabilitation to be effective for enhancing and/or maintaining QoL in the long-term has yet to be established as it is necessary to adapt interventions in order to respond to disease progression in a timely manner.

The ALS research literature could benefit from a longitudinal cohort study carried out from disease diagnosis to end of life, to determine if there is long-term potential of rehabilitation interventions to impact QoL. It is also important to understand the personal perspectives of people with ALS throughout the course of the disease. The incorporation of participant perceptions into such a cohort study could contribute to researchers’ and clinicians’ understanding of which interventions are suitable at each stage. Therefore, efficiency of

rehabilitation during all stages of the disease requires solid guidelines and evaluation measures if QoL improvements are to be demonstrated in the future.

### **Conclusion**

Introducing rehabilitation interventions early in the disease process is believed to have potential benefits in the overall management of people with ALS and stabilizing QoL (Ng, Talman, & Khan, 2011). The gap in current available evidence calls for future research to be focused on both the perceptions of people with ALS and measurable changes in QoL resulting from rehabilitation interventions. This will ultimately lead to the determination of which rehabilitation interventions would be the most beneficial to maintain and/or improve QoL in people with ALS.

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