

Upper Extremity Orthoses Use in Amyotrophic Lateral Sclerosis/Motor Neuron Disease: A Systematic Review

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Received date: 15 November 2014; Accepted date: 20 March 2015; Published date: 23 March 2015

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Abstract

Background: Orthoses decrease the effects of muscle imbalance, provide greater ease in performance of activities of daily living, prevent joint contracture, and relieve pain. There are no published systematic reviews on the use of upper extremity (UE) orthoses in amyotrophic lateral sclerosis (ALS)/motor neuron disease (MND).

Objective: The aim of this systematic review is to determine common orthosis interventions for ALS/MND.

Methods: The authors performed a systematic review of the literature available on Medline, EMBASE, Google Scholar, PubMed, the Cochrane Database of Systematic Reviews, and CINAHL. Key words used: 1) ALS, amyotrophic lateral sclerosis, Lou Gehrig's disease, MND, motor neuron disease; 2) OT, occupational therapy, hand therapy, PT, physical therapy; 3) splint, brace, orthosis, orthoses, orthotic, orthotic device. Three reviewers critically appraised 22 articles using a standard format.

Results: The reviewers identified no randomized controlled or controlled clinical trials, five level 4 (case reports) and seventeen level 5 (expert opinions) using Sackett's original 5 level pyramid.

Conclusions: Patients with ALS demonstrated improved function, increased range of motion and decreased pain with orthoses. However, there were few studies that met the search criteria. Furthermore, the studies that were reviewed had limited subjects, making it difficult to draw definitive conclusions. Further research is needed to determine appropriate study designs for the use of upper extremity orthoses in ALS/MND. Prospective studies would strengthen the results.

Keywords: ALS; Motor neuron disease; Occupational therapy; Physical therapy; Splint; brace; Orthotic device

Introduction

This article presents a review of published accounts on the use of orthoses that are helpful to persons with Amyotrophic Lateral Sclerosis (ALS) and other Motor Neuron Diseases (MNDs). The authors used the structured review technique of a systematic review [1]. "There are five steps involved in conducting a systematic review:

- Framing questions for a review.
- Identifying relevant work.
- Assessing the quality of studies
- Summarizing the evidence and
- Interpreting the findings.

The question posed in this review is, "In what manner is upper extremity orthoses use beneficial to persons with ALS (PALS)?"

One manner in which Occupational Therapists (OTs) assist patients with activities of daily living (ADLs) and improving function is through orthotic fabrication, fitting, and training. The terminology of orthoses evolved from the term "splint". Hand surgeons were adept at making orthoses from the 1880s continuing into the late 1950s [2]. Due to the polio epidemic and greater need for orthotic devices,

occupational and physical therapists became involved in their fabrication. During World War II, surgeons, nurses, therapists, and medical corpsmen made orthotic devices. By the 1960s, with the onset of the polio vaccine, the need for orthoses decreased. Occupational therapists continued making orthoses while the other professionals reduced their involvement [2]. Orthoses used by PALS may be beneficial to support weak or ineffective muscles and joints, to reposition the hand in a functional position, prevent joint contracture and muscle shortening due to muscle imbalance, and prevent pain associated with resting position.

ALS, involving both the upper and lower motor neurons, is the most common adult motor neuron disease. Upper Motor Neuron (UMN) loss in the motor cortex may cause increased muscle tone. The lower motor neuron (LMN) loss in the brain stem and spinal cord results in muscle weakness with normal or decreased tendon reflexes. The average onset age is 55 to 60 years of age. The ratio of male to female is 1.5 to 1. After age 65-70 the ratio changes to 1 to 1 [3]. The average disease length until death or mechanical ventilation is 3 years from onset of muscle weakness [3]. It is important for OTs and other health care practitioners to be aware of interventions such as orthotic fitting and training that may enhance independence in activities of daily living and quality of life. Due to the rapid progression of the disease process, it is crucial to identify the individual's needs and provide efficient interventions. The purpose of this study is to

synthesize available information on upper extremity orthosis use in ALS.

Materials and Methods

The authors compiled articles gathered from CINAHL, Cochrane, EMBASE, Google Scholar, Medline and PubMed up to the acceptance date of this article. Key words used include amyotrophic lateral sclerosis, ALS, Lou Gehrig's disease, MND, motor neuron disease, with hand therapy, occupational therapy, OT, physical therapy, physiotherapy, PT, with brace, orthoses, orthosis, orthotic, orthotic device, and splint. The authors included articles that met the following selection criteria: observational, qualitative, and quantitative studies that mention the use of upper extremity orthoses, English only, and a diagnosis of ALS/MND. The use of upper extremity orthoses was determined by the words brace, splint, orthotic device, orthosis, and orthoses concerning upper extremity rehabilitation. The authors eliminated abstracts, book chapters, posters, articles about individuals under the age of 18, and articles about powered orthotics. Fifty-five articles were initially selected and the bibliographies hand searched by two reviewers (MM and SS). If a consensus was not reached amongst the two reviewers (MM and SS), the third author (CI) would also read the article for inclusion and exclusion criteria and a discussion amongst the three authors would ensue until consensus was achieved. Twenty-two articles meeting the criteria are included in the synthesis. All three authors critically appraised each article using a standard format and provided a numerical score based on a 0-10 scale on their impression of the quality of the study based on the critical appraisal form. However, due to the lack of high-level studies, the authors chose to use all articles meeting the selection criteria regardless of the numerical value. The inter rater reliability on the forms was established.

Results

The results of the study yielded 5 case reports and 17 expert opinions (Table 1). In accordance with the 5 level pyramid proposed by Sackett [4,5], this translates to 5 level 4 and 17 level 5 studies that were critically appraised by the reviewers. Randomized controlled trials or controlled clinical trials were not discovered. During critical appraisal discussion, the reviewers chose to eliminate one article [6] from the synthesis, as it was a review of other appraised articles and did not offer new information.

Thirteen articles [7-19] advocate the use of volar wrist supports, with 5 articles specifying that the wrist be placed in 30-35 degrees of extension [9,13-16]. Twelve articles state wrist extension orthoses improve grip strength and/or efficiency therefore improving function [7,8,10,11,13-16,18,20] (Figures 1 and 2).

Eight of the reports advocate the use of resting hand orthoses that include the forearm and the hand [8,11,13,14,21-24] (Figure 3). Two case reports state that a resting hand orthosis may prevent intrinsic minus positioning, also referred to as clawing whereby the metacarpal phalangeal (MCP) joints fall into hyperextension and the Interphalangeal (IP) joints become fixed in flexion [11,21]. Conversely, McDonald and Skalsky and McDonald, recommend the MCPs be placed in neutral and the IPs be placed in flexion within the orthosis for night. The same authors promote full wrist and finger extension positioning for day use. They also emphasize that orthoses should not compromise function or sensation.

Database: EMBASE <1988 to 2015 Week 10>
Search Strategy:

- 1 exp amyotrophic lateral sclerosis/ (18342)
- 2 ALS.mp. (33650)
- 3 exp motor neuron disease/ (22864)
- 4 MND.mp. (1781)
- 5 1 or 2 or 3 or 4 (46327)
- 6 exp physiotherapy/ and exp hand/ (517)
- 7 exp hand function/ (22933)
- 8 hand therapy.mp. (352)
- 9 exp occupational therapy/ (11542)
- 10 OT.mp. (10970)
- 11 6 or 7 or 8 or 9 or 10 (45322)
- 12 exp splint/ or splint.mp. (6502)
- 13 exp brace/ or brace.mp. (6288)
- 14 orthosis.mp. or exp orthosis/ (13193)
- 15 orthoses.mp. (9511)
- 16 exp orthotics/ (2576)
- 17 orthotic device.mp. or exp orthotics/ (2643)
- 18 12 or 13 or 14 or 15 or 16 or 17 (30116)
- 19 5 and 11 and 18 (18)
- 20 5 and 18 (138)
- 21 from 19 keep 3-5, 7-10, 14 (8)
- 22 from 20 keep 5-9, 15, 30-31, 33, 51, 57, 60-61, 76, 80, 105, 107, 120, 123 (19)
- 23 21 or 22 (22)

Figure 1: Sample search strategy used, demonstrating the search on EMBASE. Same strategy used with Medline, Google Scholar, PubMed, Cochrane Database of Systematic Reviews and CINAHL.



Figure 2: Custom volar wrist orthosis, also known as "wrist cock-up" in 30 degrees of extension. This is optimal positioning for maximum grip and hand function.



Figure 3: Resting hand orthosis with optimal position of wrist in neutral, fingers slightly flexed at the MCP joints and extended at the IP joints. P IP extension helps prevent PIP contracture. Thumb is in available abduction. Loops on straps assist with self-donning and doffing.

Ten of the reports recommend a thumb positioning orthosis to assist with functional pinch [7,11,12,14,16,17,19,21,25,26] (Figure 4). Campbell and Enderby recommend thumb adduction. Goldberg discusses a small thumb orthosis that holds the IP joint in extension. Tanaka, Saura, Houraiya, and Tanimura Sinaki and Sinaki and Mulder

state that the thumb orthosis may increase independence in multiple tasks such as holding a cup, fastening buttons, writing, and pouring water. Tanaka, Saura, Houraiya and Tanimura published an algorithm for clinical decision-making. They recommend early use of a wrist cock-up splint, long thumb spica (Figure 5), short thumb spica, and simple web spacer. Other authors advocate the use of a thumb spica orthosis for opposition [11] and improved grasp in those with thumb

abduction and extension weakness [14]. Combining the wrist and the thumb in a single orthotic to address both weak wrist extension as well as weak abduction may be useful in some cases [12,16,19,21] (Figure 5). Additionally, Lenox and Sinak discuss use of dynamic finger extension assist orthoses. Lenox also notes that a dorsal placement of the wrist orthosis may allow for increased finger motion.

Author (s)	Methodology	Level of Evidence	Type of Orthosis/Orthoses	Outcome/Conclusion
Campbell & Enderby	Expert Opinion	5	<ul style="list-style-type: none"> Wrist cock-up orthosis First webspace orthosis for adduction 	<ul style="list-style-type: none"> Improved grip, pinch and grasp strength
Dal Bello-Haas, Kloos & Mitsumoto	Case Report	4	<ul style="list-style-type: none"> Wrist thumb orthosis Resting hand orthosis 	<ul style="list-style-type: none"> Assist in reducing pain due to contractures Wrist thumb orthosis used for activities Resting hand orthosis for prevention of clawing Support weak muscles
DeLisa, Mikulic, Miller & Melnick	Expert Opinion	5	<ul style="list-style-type: none"> Wrist support orthosis Resting static orthosis 	<ul style="list-style-type: none"> Provide improved wrist extension and improved hand function Wrist cock-up may assist to grasp objects and maintain optimal joint positions May reduce contractures which lead to pain and decreases functional use Consideration should be given to the patient's perceived value of the device
Francis, Bach & Melnick	Expert Opinion	4	<ul style="list-style-type: none"> Wrist cock-up orthosis in 30-35° extension 	<ul style="list-style-type: none"> Improve grip efficiency
Goldberg	Case report	4	<ul style="list-style-type: none"> Small thumb orthosis to hold the IP in extension 	<ul style="list-style-type: none"> Orthosis deemed unsuccessful; unable to grasp the remote control while wearing the orthosis
Goldblatt	Expert Opinion	5	<ul style="list-style-type: none"> Wrist cock-up orthosis 	<ul style="list-style-type: none"> Brings fingers into position to exert maximum grip strength
Ivy, Smith & Materi	Case report	4	<ul style="list-style-type: none"> Prefabricated including figure of eight and dorsal wrist orthoses Custom orthoses including finger orthosis, hand based thumb spica, resting hand, and sttachable hand component for extension 	<ul style="list-style-type: none"> Can improve meaning and quality of life Therapists play a crucial role on the team and can recommend and fabricate orthoses Prevent PIP buckling during use of mouse Dorsal wrist orthosis maximizes use of sensory surface Dorsal wrist orthosis lighter Circumferential finger orthoses prevent contractures, decrease pain and add leverage for activities requiring pressure force Attachable digit night extension piece may be used with day wrist orthoses
Janiszewski, Caroscio & Wisham	Expert Opinion	5	<ul style="list-style-type: none"> Opponens orthosis Cock-up orthosis 	<ul style="list-style-type: none"> Reports orthoses helpful Supprots weak muscles Recommend prompt referrals
Johnson, Florence & Abresch	Expert Opinion	5	<ul style="list-style-type: none"> Neutral resting position during sleeping hours 	<ul style="list-style-type: none"> Goal of limiting contracture Suggests early intervention
Lenox	Expert Opinion	5	<ul style="list-style-type: none"> Palmar and dorsal cock-up orthoses Static and dynamic orthoses 	<ul style="list-style-type: none"> Dorsal cock-up improves mobility in hand and fingers Static orthoses support weak and immobile fingers and reduce painful contractures Dynamic orthoses are costly and may be difficult to obtain
Lewis & Rushanan	Expert Opinion	5	<ul style="list-style-type: none"> Wrist cock-up orthosis in approximately 30° extension Resting hand orthosis 	<ul style="list-style-type: none"> Wrist cock-up can improve grip function, reach and grasp Resting hand orthoses may be used to prevent contractures

McDonald	Expert Opinion	5	<ul style="list-style-type: none"> Nighttime resting orthosis in wrist extension and MCP extension, PIP flexion Daytime position emphasis on wrist and finger extension 	<ul style="list-style-type: none"> Early diagnosis and initiation of orthoses contractures are still mild Should not compromise function *See discussion
Majmudar, Wu & Paganoni	Expert Opinion	5	<ul style="list-style-type: none"> Resting hand (light weight; wrist in neutral) Anti-claw (intrinsic plus) Volar cock-up in 20-30° of wrist extension Short opponens with thumb in abduction and opposition 	<ul style="list-style-type: none"> Reduce hand pain from finger contractures Allow for increased MP flexion for functional positioning during activities Improved grasp when wrist extensors weak Improved grasp with weak intrinsic muscles Useful day and night Recommends controlled studies for impact and safety
Morris	Expert Opinion	5	<ul style="list-style-type: none"> No specific orthoses were described 	<ul style="list-style-type: none"> Can help to prevent and decrease impairment such as muscle strain, or disuse atrophy Prompt referral are needed to optimize function Publication of therapy outcomes needed for reimbursement and accountability
Rocha, Reis, Simoes, Fonseca & Medes Ribeiro	Expert Opinion	5	<ul style="list-style-type: none"> Wrist extension orthosis in 30° of extension 	<ul style="list-style-type: none"> Improved grip function
Sinaki	Expert Opinion	5	<ul style="list-style-type: none"> Wrist support Thumb orthosis lacing the thumb in apposition 	<ul style="list-style-type: none"> Wrist supports improve extension and allow for a better grip Thumb orthosis in which the thumb is placed in apposition allows for more effective use of hand for fine motor movements
Sinaki & Mulder	Expert Opinion	5	<ul style="list-style-type: none"> Thumb orthosis Wrist immobilization orthosis Dynamic MCP extension orthosis 	<ul style="list-style-type: none"> Thumb shell extends to the IP joint of the thumb allowing for thumb opposition, enabling a 3-point pinch; function
Smith & Norris	Expert Opinion	5	<ul style="list-style-type: none"> Wrist orthosis 	Orthosis may help with grasp
Skalsky & McDonalds	Expert Opinion	5	<ul style="list-style-type: none"> Nighttime resting with MP extension and IP flexion Daytime wrist and finger extension 	<ul style="list-style-type: none"> Use orthoses before contractures present and while contractures mild Orthosis use should not compromise function or sensation *See discussion
Tanaka, Houaiya, Akagi & Kihoin	Expert Opinion	5	<ul style="list-style-type: none"> Short thumb spica Long thumb Spica Simple webspacer Cock-up orthosis 	<ul style="list-style-type: none"> Simple webspacer improved tip to tip pinch strength, improved FIM score, and improved ability to fasten buttons and write Cock-up for inability to extend wrist Hand orthoses useful for increasing independence Algorithm can help providers choose what orthosis is best for particular patients
Tanaka, Saura, Houraiya & Tanimura	Expert Opinion	5	<ul style="list-style-type: none"> Neoprene orthosis as simple webspacer 	<ul style="list-style-type: none"> Orthoses improve function Orthoses must be fabricated in a timely fashion Ease of application important Usefulness diminishes as disease progresses Soft material suggested with intact sensation *See discussion

Table 1: Case reports & experts opinion.

In addition to suggestions on positioning for prevention of contractures and efficiency in grip and pinch strength, many of the reports address specific functional improvements with orthotic use. Seven of the articles mention functional gains with orthosis use [8,11,13,16,17,21,26]. Described functional improvements include using technology, holding objects including a hairbrush, walker, cup, and utensils and the ability to complete personal hygiene. In addition,

case reports discuss improvement in sociability and participation in leisure activities such as crocheting [11].

Case reports also show relief of pain with orthoses [11,21]. In one of the reports, two case studies explicitly mention circumferential finger and figure of style orthoses, prefabricated dorsal wrist supports, and nighttime forearm resting hand orthoses for pain management and/or comfort [11] (Figures 3, 6-8).



Figure 4: Custom hand based thumb orthosis with IP joint free allowing opposition.



Figure 5: Custom combined wrist and thumb orthosis as recommended by several authors.

This systematic review reveals that the published literature on the use of upper extremity orthosis with ALS/MND is anecdotal and based on expert opinion and case report. Most of the level 4 studies are single patient case studies. Furthermore, the outcomes that were used to measure success in the studies are not consistent or comparable. Based on these facts, there is not enough information to assess bias between studies. There is not enough data to compare the recommendations between studies mathematically to look for bias. To compare the studies mathematically, it would be necessary to compare studies with more than one patient that measure the same outcome.



Figure 6: Custom circumferential finger orthosis for comfort and function. This patient used this orthosis for using the tablet that also became her communication device.



Figure 7: Custom figure of 8 style orthoses for comfort and function. This stabilizes fingers with weak intrinsic muscles.



Figure 8: Commercial dorsal wrist orthoses with commercial figure of 8 (arrow). This style may be preferred as it leaves volar surface free for sensory input and is lighter weight.

Discussion

Despite not finding higher level of evidence studies, there were a number of case reports and expert opinions on the topic of orthoses with motor neuron disease documented in the literature. No prospective studies and no consistent methodology amongst the authors in reaching the conclusions that they reported in the papers are the biggest limitations of this systematic review. Because of this, it is impossible to completely synthesize the information. It is clear however, that there is a trend toward the usefulness of wrist extension orthoses, night orthoses, and thumb positioning orthoses. Persons with ALS/MND could benefit from further published accounts on the use of upper extremity orthoses to decrease the effects of muscle imbalance, provide greater ease in performance of ADLs, prevent joint contracture, and relieve pain.

Published accounts could come in the form of various prospective methods. A case series may be one useful method. This is a descriptive study that could be arranged prospectively to monitor change in function, pain, and range of motion over time using consistent outcome measures. Planning to report cases ahead of time would allow for increased detail and uniformity in the data that is collected. This in turn would allow increased ability to analyze the usefulness and best timing of orthotic intervention. Completing a randomized controlled trial on the effectiveness of orthotic use could occur by comparing various orthosis designs. A comparison might be made between volar and dorsal wrist orthoses, full extension of the wrist versus partial extension and /or prefabricated versus custom orthoses. Outcome measures for these types of studies could include functional outcome

measures, pain assessments, and patient satisfaction surveys. Existing valid and reliable measurement tools are pain scales such as a Likert scale [27] range of motion with goniometry [28,29], grip and pinch strength [30,31]. The Disability of the Arm, Shoulder and Hand (DASH) questionnaire or the shorter version of this (QuickDASH) [32], the Upper Limb Functional Index [21,33], the Canadian Occupational Performance Measure [34] and quality of life scales developed specifically for ALS [35] or others with advanced disease [36-38]. Pre- and post-photographs could document success in positioning and reduction of resting contracture Biomechanical studies may be useful to further evaluate the impact of imbalance between the intrinsic and extrinsic musculature and how best to accommodate to the imbalance with external support and positioning.

There was some conflicting information in the reports on the positioning for resting orthoses. Two papers described placing the hand in the intrinsic minus position, which is with the MCPs extended, and the IPs flexed [23,24]. This is a common position of deformity with intrinsic weakness, frequently observed in ALS/MND, therefore one would typically place the hand in an alternate position with the MCPs flexed and IPs extended in order to place the presumably shortened soft tissue structures on stretch. If however, spasticity or a comorbid condition such as rheumatoid arthritis leads to laxity of the MCPs or subluxation of the extensor digitorum communis over the metacarpal heads, this would be an optimal position for the hands. Another conflicting recommendation was placing the thumb in adduction, also a common deformity in ALS/MND due to muscle atrophy [7]. Typically, most grasp patterns favor the thumb in opposition. If however, the provider wishes to gain a strong lateral or appositional pinch, also known as “key pinch” as in grasping a car key, adduction of the thumb may be desirable. Tanaka et al. recommend soft material if the patient has intact sensation. It is important to try to make all orthoses as comfortable and lightweight as possible and sometimes a softer material achieves these goals. If there is not intact sensation, which would typically be from a co-morbid diagnosis such as an ulnar nerve compression, soft material may also be important to lessen the chance of pressure related ulcers that may go unnoticed due to the lack of sensation.

Summary

There were several themes apparent in the appraisal of the papers. One overriding theme noticed in the 21 papers is emphasis on early intervention and a multidisciplinary approach. The importance of frequent evaluations in order to have the most beneficial timing of orthotic intervention is another theme. Authors also regularly suggested individualizing the treatment with particular attention to the unique functional goals and needs. Figure 9 provides a quick reference for common ALS/MND upper extremity problems and some possible orthotic solutions. Wrist hand finger and thumb orthoses in a resting position, worn at night or at rest during the day, may help to maintain tissue length. A wrist orthosis may increase grip strength and provide support for functional use and grasp during activities of daily living. Finger extension orthoses may assist with preventing and decreasing contractures at the proximal interphalangeal joints, thus preventing or diminishing pain. An opposition style thumb orthosis may be helpful in positioning the hand in a functional posture. It is essential to be aware of problems arising with sensation or function when using an orthotic intervention. One may achieve this through periodic assessments by the occupational therapist for proper fit as well as training and discussion of the purpose and use of the orthosis. It is

important to assess the patient’s function throughout the course of the disease process due to the progressive nature of the disease and thus the changing needs of the patient. Future published accounts of the benefits of upper extremity orthotic intervention for ALS/MND will help guide timing of fitting and fabrication as well as decisions on the most appropriate upper extremity orthoses to consider.







Problem	Solution
Weak finger extension with overdrive of the flexors and/or weak hand intrinsic muscles	<p>A</p>  <p>•Oval 8 or custom circumferential orthosis</p>
Decreased grip and/or weak intrinsic muscles	<p>B</p>  <p>•Universal cuff in shown; patient using a custom device to scratch her chin •Universal cuffs may be used with eating utensils, combs, make-up wands and hygiene devices</p>
Joint contractures, joint pain, median nerve compression, and risk of developing joint contractures	<p>C</p>  <p>•Custom circumferential for PIP joints (see A) •Resting forearm based orthosis (nighttime)</p> <p>D</p>  <p>•Commercial orthosis pictured •Custom added resting finger extension piece to commercial dorsal wrist orthoses</p>
Intrinsic wasting/loss of opposition	<p>E</p>  <p>•Hand based thumb spica (shown) •Resting forearm based orthosis (nighttime) in intrinsic plus positioning of fingers (see C & D)</p>
Wrist-drop/decreased grasp	<p>F</p>  <p>•Commercial dorsal orthosis, such as Quad Quip shown, consider volar wrist orthosis or long thumb spica</p>

Figure 9: Quick reference for common ALS.

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