

# Augmentative and Alternative Communication for People with Progressive Neuromuscular Disease

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## KEYWORDS

- AAC • Amyotrophic lateral sclerosis • Duchenne muscular dystrophy
- Myotonic muscular dystrophy • Spinal muscular atrophy • Communication
- Assistive technology

## KEY POINTS

- Augmentative and alternative communication (AAC) is considered standard practice in interventions for individuals with progressive neuromuscular disease.
- Individuals with progressive neuromuscular disease can maintain effective, functional communication by implementing AAC when natural speech no longer meets their needs.
- AAC strategies and systems may be customized to accommodate for individual needs by exploiting intact abilities (eg, spinal muscular atrophy distal movements, amyotrophic lateral sclerosis eye movements).
- New technologies under development promise access to communication systems for individuals with progressive neuromuscular diseases who previously were considered locked-in and unable to retain effective interactions (ie, brain-computer interface).

The purpose of this article was to (1) profile frequently occurring communication impairments associated with neuromuscular disease, (2) identify communication needs within an augmentative and alternative communication (AAC) framework, and (3) identify options for AAC supports. Four neuromuscular diseases are examined: amyotrophic

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Funding sources: None.

Conflict of interest: None.

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Phys Med Rehabil Clin N Am 23 (2012) 689–699

<http://dx.doi.org/10.1016/j.pmr.2012.06.003>

1047-9651/12/\$ – see front matter © 2012 Published by Elsevier Inc.

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lateral sclerosis (ALS), Duchenne muscular dystrophy (DMD), myotonic muscular dystrophy (MMD), and spinal muscular atrophy (SMA). Electronic databases including CINAHL, MEDLINE (PubMed), OVID, and EBM Reviews from January 1995 through March 2012 were queried. Search terms included the name(s) and acronyms of the disease (eg, for ALS: amyotrophic lateral sclerosis, ALS, Lou Gehrig's disease, motor neuron disease; for DMD: Duchenne muscular dystrophy, DMD, Duchenne, pseudohypertrophic; for MMD: myotonic muscular dystrophy, MMD, Steinert disease) AND dysarthria, communication disorder, speech, speech disorder, speaking, communication device, speech generating device, augmentative and alternative communication, technology/computer access, and assistive technology.

The review for ALS (amyotrophic lateral sclerosis, motor neuron disease, Lou Gehrig's disease and dysarthria, communication disorder, speech, speech disorder, speaking, communication device, speech-generating device, augmentative and alternative communication, and technology/computer access, assistive technology) yielded 122 different references and articles. There were no randomized controlled trials identified. Among those reporting data on ALS and AAC, 36 were published in peer-reviewed journals and presented outcome research and individual cohort studies (ie, levels 2b and 2c); these are presented in this document. Additionally, the literature review yielded no results for Duchenne, myotonic, or spinal muscular atrophy related to AAC.

## **AMYOTROPHIC LATERAL SCLEROSIS**

Amyotrophic lateral sclerosis is a rapidly progressive paralyzing disease. Most individuals with ALS die within 2 to 5 years of onset.<sup>1</sup> No cure exists, and limited treatments are available; clinical management consists primarily of symptom management. Many people diagnosed with ALS participate in multidisciplinary clinics where they receive coordinated care from a neurologist and/or physiatrist, physical therapist, occupational therapist, speech-language pathologist, dietitian, social worker, respiratory therapist, and nurse case manager. In general, participation in multidisciplinary clinics is supported by evidence indicating increased use of effective interventions including riluzole, percutaneous endoscopic gastrostomy, and noninvasive ventilation, resulting in fewer hospitalizations.<sup>2</sup> The impact of ALS on a person's participation patterns will vary depending on the individual's life stage, ALS type, and life-extending decisions (eg, gastrostomy, ventilator).<sup>3</sup>

### ***Communication Impairments in ALS***

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Because of the involvement of both upper and lower motor neurons in ALS, motor speech impairments result in a mixed spastic-flaccid dysarthria. Initial speech symptoms among people with ALS (pALS) vary, but a general pattern is often apparent. Initially, speaking rate slows, followed by moderate and then more severe reductions in intelligibility that affect overall communication effectiveness, and continued progression ultimately leaves the person with no functional speech (ie, anarthric).<sup>4</sup> Because there have been no controlled studies examining communication in ALS,<sup>1</sup> the American Academy of Neurology notes that there are insufficient data to support or refute treatment to optimize communication in ALS.<sup>5</sup> Still, because of progressive loss of natural speech, the standard of care for individuals with ALS indicates treatment emphasizing strategies for optimizing natural speech ("communication for as long as possible") and focusing on an individual's expressive language capabilities as well as communication effectiveness among various partners.

Although respiratory failure is the most common cause of death in ALS, mechanical ventilation can prolong life expectancy, but will not halt the relentless progression of

paralysis. Noninvasive ventilation is often the initial treatment to alleviate respiratory symptoms and invasive ventilation via tracheostomy is proposed when noninvasive ventilation is no longer effective owing to disease progression, low tone in bulbar musculature, or difficulty with secretions.<sup>6</sup> When a pALS accepts mechanical ventilation, the clinician must be aware of and resolve incongruent decisions (ie, accepts mechanical ventilation to prolong life but refuses a speech-generating device [SGD] to provide communication even with ongoing functional decline). Similarly, when a pALS accepts ventilation, she or he requires ongoing intervention to ensure appropriate access accommodations to the SGD to maintain effective communication.

Customized AAC systems should be designed for each pALS that include means to communicate basic medical messages as well as new information for daily conversation; a way to “chat” or just casually interact; a means to access and use the telephone; options to call attention for assistance; and ways to express affection, humor, and emotions.<sup>7</sup> The AAC system should change as dysarthria progresses in ALS, with system components ranging from basic alphabet or symbol boards to computer-based SGDs.<sup>6</sup> One key indication for the use of AAC by pALS is that “functional communication is an essential component to improved quality of life for persons with severe physical limitations, such as those experienced by persons with ALS.”<sup>8(p377)</sup> Although outcomes have not been studied systematically, individuals often comment on the value of the AAC system: “My computer has many functions. It is my writing system; my communication system (e-mail, fax), especially for family further away; my information system; my database for addresses and other lists; my financial and legal organizer; my entertainment system; and, lastly, my speech system.”<sup>9(p124)</sup>

### **AAC Acceptance in ALS**

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Caregivers who support individuals with ALS report very positive attitudes toward AAC technology, indicating greater rewards associated with caregiving and increased social closeness to the person with ALS.<sup>7</sup> AAC has a high level of acceptance among pALS, with nearly 96% accepting AAC technology (90% on initial recommendation, 6% following a delay) when provided with options in a timely manner.<sup>10</sup> One departure from this acceptance may be individuals with ALS who have co-occurring severe frontotemporal lobar dementia or accompanying severe health issues, as these may have a higher rejection rate of AAC technology and therefore require careful consideration.<sup>10</sup> Individuals who reject AAC technology may, however, implement strategies that do not rely on technology to support their communication, such as partner-supported scanning, hierarchical yes/no responses, and eye gaze to direct interactions.<sup>4</sup>

### **AAC Use in ALS**

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Individuals with ALS use AAC technology to sustain employment, program computers, access word processing to write documents, provide accounting services, and interact on the telephone and Internet.<sup>11</sup> Advances in technology promise greater access to interactions that support social, recreational, educational, commercial, volunteer, and employment activities.<sup>12</sup> Duration of AAC technology use may be dependent on factors including life expectancy, nutritional status, timelines for AAC provision, and decisions for life-prolonging procedures. In one report, individuals with ALS who acquire and use AAC technology continue to communicate using the same device for a mean duration of 28.4 months, with those who presented with spinal ALS having a mean duration of 32.1 months and those with bulbar ALS having a mean duration of 25.2 months.<sup>8</sup> Individuals with ALS who acquire AAC technology to support their communication continue to use the devices until the end of life, or within a couple weeks before death.<sup>8</sup>

### **ALS case study**

Gil is a 39-year-old man who works at a large regional hospital as a data entry specialist in the Information Technology Department. Recently diagnosed with bulbar-onset ALS, Gil hopes to continue working as long as possible. At his most recent ALS clinic visit, the speech-language pathologist completed a brief evaluation and identified slowed speaking rate and mild dysarthria; she then referred him for an AAC evaluation. During the AAC evaluation, Gil had the opportunity to examine and try a variety of SGDs and together with his wife and teenage daughter, planned for his communication strategy. He already was using an iPad to keep his schedule, plan meetings, surf the Web/e-mail, and for entertainment. To minimize out-of-pocket expense but remain functionally communicative, his initial plan was to add a communication app (eg, Verbally [Intuary, Inc, San Francisco, CA, USA]) with an onscreen keyboard and synthesized voice output to his existing iPad. He purchased a small external speaker that was embedded in an iPad case so that the voice output would be loud enough to be heard in most of his communication situations. He used this system to interact with others at work, to participate in religious services, and when he went out to play cards with his friends. As Gil's ALS progressed, he began having difficulty using his hands to access the iPad keyboard and relied on a walker to steady himself while ambulating. At that point, his respiratory function had declined and he made the decision with his family to proceed with mechanical ventilation; subsequently, he also acquired a dedicated SGD to provide more access options. As he was also in the process of acquiring a power wheelchair (eg, Permobil [Permobil, Inc, Lebanon, TN, USA]), he selected a large-format SGD with eye gaze access (eg, Dynavox Eyemax [DynaVox Mayer-Johnson, Pittsburgh, PA, USA]) that could be mounted to the wheelchair and easily moved to another mount in his home. He used the SGD to communicate with coworkers until he resigned (10 months from initial iPad implementation) and continued using the SGD until approximately 1 week before death. During the final days of life, Gil used the SGD intermittently, but also used a partner-supported hierarchy of questions to which he responded yes by looking up and *no* by looking down.

### **DUCHENNE MUSCULAR DYSTROPHY**

DMD is an X-linked recessive degenerative disease caused from absence of the dystrophin protein that stabilizes and protects muscle fibers.<sup>13</sup> Standards of care were recently developed by the DMD Care Considerations Working Group under the auspices of the US Centers for Disease Control and Prevention.<sup>14</sup> Among others, the recommendations included the multidisciplinary approach and the multidisciplinary team, including a speech-language pathologist, as a key aspect of care.

#### ***Communication Impairments in DMD***

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Speech intelligibility decreases because of deteriorating respiratory support. Compensatory strategies, speech amplification, and AAC become appropriate as communication becomes increasingly limited.<sup>14</sup>

### **DMD case study**

RJ is a 26-year-old man with DMD who resides in an adult foster home in a rural community. RJ graduated from the local high school 5 years ago without any means to control his environment, no computer for writing, recreation, or employment. He did not have a means to explore the Internet for information. He is currently quadriplegic with strained respiration. He can move his head and has control of his eye movements and eye blinks. He does have a significant head tilt to the right, which affects his

face-to-face eye contact for conversation. RJ uses a wheelchair for mobility that is pushed by his caregivers. He leans forward in his chair to facilitate respiration. RJ relies on speech with significantly reduced volume for spontaneous personal communication (Melanie Fried-Oken, 2010), as well as for calling attention, talking on a speakerphone, and managing his medical needs. When he is travelling in his adapted van, he does not have an effective means to speak with the driver or other passengers. RJ came to the AAC clinic looking for a means to control his computer to surf the Internet and write. Language and cognitive testing indicated that this young man had receptive and expressive language skills that were within normal limits for adults. He was an adequate speller and enjoyed reading magazines when someone held them up and turned pages for him. With assistance from a clinical consultant at Words+, RJ tried to use an SGD accessed through advanced eye gaze technology. A computer with a low-voltage USB camera (EyePro GS [Words+, Inc, Lancaster, CA, USA]) was set up in front of him, and he learned how to use his eyes to control a mouse and select letters from an on-screen keyboard. After an initial evaluation, RJ worked with the consultant through remote training sessions. The consultant and RJ would meet on Skype, where she could watch RJ practice eye control with different computer applications over multiple sessions. He learned how to set up an e-mail account, surf the Internet, and compose text with specialized word processing software (called E Z Keys [Words+, Inc, Lancaster, CA, USA]) that provided keystroke enhancement techniques for word prediction, macros, and text storage. RJ acquired the necessary technology through his medical insurance. He now has an effective AAC system that includes an SGD for writing and environmental control, head nods, facial expression, and smiles for face-to-face communication, and a simple voice amplifier (the Chattervox, Indian Creek, IL, USA) for increasing his speaking volume when he is in a room alone and needs medical assistance or is in the van with a driver or other passengers. He still relies on caregivers to set up the computer and EyePro so that he can use it for different tasks, but his access to the world, his acquisition of new knowledge through the Internet, and his quality of life have been enhanced by current assistive technology.

## MYOTONIC MUSCULAR DYSTROPHY

MMD is an autosomal dominant disease that may be evident at birth (the congenital form is more severe) or more commonly in teenage or adult years.<sup>15</sup> Myotonia is characterized by slow relaxation of muscles following voluntary contraction that may require repeated attempts to ultimately return to a neutral position (ie, prolonged muscle contractions).<sup>16</sup> Cognitive impairment, executive dysfunction, and avoidant personality traits, eventually deteriorating with age, have been described.<sup>17</sup>

### *Communication Impairments in MMD*

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In addition to the motor deficits, up to 51% of children with MMD may demonstrate symptoms of autism spectrum disorder, 83% to 95% have moderate to severe learning disability, and 94% to 100% have problems with social interaction. The relationship between MMD type and cognitive function illustrate that with more severity, the IQ (ie, Wechsler Intelligence Scale for Children—III full-scale IQ [severe M = 40.3, mild M = 27.9]) and adaptive skill scores (ie, Vineland Adaptive Behavior Scales; severe M = 36.5, mild M = 44.8) may be lower.<sup>18</sup> Although impaired, children with MMD have relative strengths with verbal understanding (eg, vocabulary, similarities) and receptive language.<sup>18</sup> For AAC, it is important to consider that the myotonia may influence an individual with MMD regarding access, particularly direct access that requires muscle contraction and release. AAC systems should be programmed

to support expressive communication and exploit stronger receptive language abilities.

### **SPINAL MUSCULAR ATROPHY**

SMA is a recessively inherited neuromuscular disease characterized by degeneration of spinal cord (ie, lower) motor neurons, resulting in progressive muscular atrophy and weakness. There is a broad spectrum of severity ranging from early infant death to normal adult life span with only mild weakness. There are unique clinical features of each SMA type and heterogeneous clinical features are used to classify various phenotypes. Some features of SMA that may affect communication include impaired head control, tongue atrophy, weakness and hypotonia in limbs and trunk, typical cognitive and emotional development, delayed motor milestones (proximal more than distal), and severe respiratory weakness.<sup>19</sup>

#### ***Communication Impairments in SMA***

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Flaccid dysarthria and swallowing impairments occur among individuals with SMA, the severity of which will vary based on the SMA type and overall disease severity.<sup>20</sup> The natural history of SMA involves inspiratory, expiratory, and bulbar weakness that may affect communication, particularly in types 1 and 2. Because strength tends to be preserved in distal (eg, extremities) more than proximal structures (eg, head/neck), a focus on access to AAC via distal body movements (eg, hands, fingers, toes) may be recommended. The young age of the most severe forms of SMA also implies a need for adapting AAC for preliterate children by using transparent symbols and photographs that are readily associated with the item.

### **AAC TECHNOLOGIES FOR COMPLEX COMMUNICATION NEEDS**

Because the progressive nature of neurodegenerative disorders often ultimately results in the complete loss of functional upper extremity and lower extremity skills, access to traditional AAC systems can become challenging. Although the preponderance of research in AAC for people with neuromuscular diseases has focused on ALS, the technologies described may be used to provide support to people with other neuromuscular impairments (eg, DMD, MDD, SMA).

#### ***Eye Gaze Tracking***

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Because of the ongoing progression of ALS and subsequent loss of upper extremity function, direct access to AAC technology is most commonly accomplished using eye gaze technologies at present.<sup>21</sup> Eye movements of individuals with ALS are typically spared but occasional impairments are identified (eg, oculomotor apraxia, reduced saccade velocity, eyelid opening/closure) making trial use of eye gaze-based AAC technology essential in determining effectiveness.<sup>22</sup> Previous studies have examined eye movement in ALS and identified decreased saccadic velocity and some abnormal saccadic patterns (eg, overlapping, low velocity, long duration) along with fewer eye movements.<sup>23</sup> Assessment of eye fatigue has been limited in ALS research, but reports indicate that eye gaze can be an effective means of AAC input.<sup>24</sup>

Infrared eye-tracking technology projects a safe, invisible infrared light that causes a reflection on the pupil of the person's eye. Paired with the infrared light, AAC technology uses software to triangulate eye movement to track items on screen. When the person's eye(s) dwells on a location, the system selects the item and thereby assembles a message; which, when paired with synthesized voice output will "speak." Compared with scanning through items on screen and selecting the desired item

with a switch, eye control has been found to produce faster messages with reduced error rates for people with ALS.<sup>25</sup>

### ***Head Movement Tracking***

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Tracking of head movements may be accomplished by infrared or video cameras, with the most common application using infrared technologies. Head movement tracking uses similar methods of capture, but track movement of a reflective surface (eg, dot, sphere) instead of eye movements. Essentially, this technology allows the head movements to “take over” the computer mouse and operate the AAC technology. Selections, as with eye tracking, are made using a dwell function or a secondary switch.<sup>26</sup>

A second form of head tracking is accomplished using laser pointers mounted to the head or other body part (eg, headband, glasses frame, cap, sock, hand). The laser beam is directed to the item for communicating a message. With concerns regarding the eye safety of laser beams, a Safe-Laser was developed. The Safe-Laser System (Invotek, Inc, Alma, AR, USA.) operates at a low, eye-safe level until directed uninterrupted at a laser safe surface. The surface can then be programmed with AAC technology for message formulation and may be an effective strategy for some individuals with ALS.<sup>27</sup>

### ***Gesture Tracking***

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Currently, eye-tracking and head-tracking technology uses a dwell function to select items on a screen to assemble messages. Gesture input strategies are now emerging that allow more precise targeting and activation of items on a display, but typically require touchscreen access (eg, SWYPE [Nuance Communications, Inc, Burlington, MA, USA]).<sup>28</sup> Recently, a prototype (ie, Gesture-Enhanced Word Prediction [Invotek, Inc, Alma, AR, USA]) has been developed to take head and hand movement tracking and translate it into the gesture interface for faster, more accurate message assembly and production by reducing the number of precise dwells that are required to generate text.<sup>29</sup>

### ***Speech Recognition***

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Use of speech recognition has increased and has been incorporated into mainstream technologies (eg, smart phones, tablets); however, commercially available speech recognition has been developed for a market of typical (nondisabled) users). For individuals with dysarthria, this technology remains largely inaccessible, as it does not recognize moderate to severely dysarthric speech<sup>30–32</sup>; however, technology is being developed that uses speech recognition based on models of dysarthric speech.<sup>33–37</sup> Applications include environmental control and specific software programs with limited recognition vocabulary sets. Others have investigated the use of dysarthric speech recognition along with electromyography signals to increase accuracy,<sup>38</sup> or the use of alphabet supplementation and language modeling to develop a functional writing and communication system for individuals with moderate and severe dysarthria.<sup>39</sup> As researchers and developers begin to look at speech recognition as part of an “input” system to computerized technology, individuals with neuromuscular conditions will be able to functionally use the technology to support writing and communication.

### ***Brain-Computer Interface***

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Another input system that is receiving considerable research attention for individuals with severe neuromuscular impairments is the novel brain-computer interface (BCI).<sup>40</sup>

The technology for BCI relies on a signal detection system that translates neuronal activity into device demands. The components necessary for functional BCI have matured sufficiently that we can imagine devices that are powered by mere thoughts. There are 3 recognized categories for BCI: the invasive BCI records neuronal action potentials (spikes) or local field potentials when a circuit board is actually placed directly on to the cortex.<sup>41</sup> One publicized invasive BCI is the Braingate Neural Interface System where intracortical microelectrode sensors read control signals directly from the motor cortex.<sup>42</sup> Noninvasive BCI relies on recording sites at the scalp for electroencephalography or on magnetic brain forces: magnetoencephalography. A final technique is a “partially invasive” BCI: electrocorticography, which uses sensors placed within the skull but outside gray matter. The noninvasive methods must filter artifact from a signal that is far from its source, but filtering techniques are improving rapidly so that the advantages of lower cost, portability, lack of concern for infection, avoiding surgery, and faster application are improving their appeal.

BCI for communication is being examined worldwide. Reports of functional use for daily tasks are beginning to appear for individuals with ALS or spinal cord injuries as a means to significantly improve quality of life and productivity.<sup>43,44</sup> The RSVP Keyboard (under development by Cognitive Systems Laboratory, Northeastern University, Boston, MA, USA) is one noninvasive system that incorporates language models into signal acquisition to monopolize on word prediction and completion for spelling.<sup>45</sup> The P300 speller is another noninvasive BCI that has been used by individuals with ALS.<sup>46</sup> BCI will one day be a clinically available tool for increasing independent daily function. It has much to offer as a promising access method for people with severe neuromuscular impairment as a tool to improve quality of life and support participation for communication, environmental control, and computer access.

## SUMMARY

Individuals with progressive neuromuscular disease often develop complex communication needs and consequently find that interaction using their natural speech may not sufficiently meet their daily needs. Increasingly, assistive technology advances provide accommodations for and/or access to communication. Although research evaluating the use of AAC across all neuromuscular disease is severely lacking, studies in ALS suggest that use of an AAC is generally well accepted and improves quality of life. AAC systems continue to be designed and implemented to provide targeted assistance based on an individual’s changing needs. Advanced technologies using BCI are becoming more readily available and have the potential to extend access to communication to even the most severely disabled patient with neuromuscular disease.

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