

Chapter 7

Decision-Making for Access to AAC Technologies in Late Stage ALS

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Introduction

How important is communication to you?

“There are no words to describe how important it is for me to communicate independently! Personally, I can't imagine life without a communication device. It is as important for me as breathing and nutrition.” —Rich, individual with late stage ALS

Within the past 20 years, augmentative and alternative communication (AAC) intervention has dramatically improved means of expression for people with ALS. Changes have occurred at many levels, from national advocacy and AAC funding policies, to technological advances such as preserving your voice with message and voice banking and the promise of brain-computer interface for computer control. Three significant paradigm shifts have occurred during the past two decades for our global society. First, there is an increased interest in distance or digital communication worldwide. The general population now accesses many services through the Internet, relying on computers for shopping, banking, information retrieval, employment, entertainment, and dyadic communication (Shane et al., 2011). Second, the growth of personal mobile technologies has opened up the acceptability of relying on technology for communication for the general population, reducing the stigma of introducing AAC devices for individuals who cannot use speech for effective message generation (McNaughton & Light, 2013). These advances have benefited people with ALS if they have a means to select content or to interact accurately and efficiently with computers. Finally, access technologies, or the means to

physically interact with communication options, have experienced significant attention and development, especially in the past decade (Fager et al., 2012, 2019).

In this chapter, access technologies for people with ALS in the late stages of the disease are highlighted. This population often has very limited or minimal movement and few reliable, consistent options to connect to technology for communication, recreation, environmental control, social connection, and cognitive stimulation. Information is presented on the classification of late stage ALS, including both impairment-based symptoms and function-based perspectives. Then, a clinical example is offered, describing Rich, a man with late stage ALS who has chosen to undergo tracheostomy and extend his life with the support of mechanical ventilation. Current and emerging access technologies, including eye tracking, switch scanning, multimodal access, and the promise of brain-computer interface as means to interact with AAC technologies is also discussed throughout. Guidelines for clinical decision making are proposed and questions are offered to assist individuals with ALS and their clinical teams motor skills worsen. Decision-making guidelines are illustrated with another clinical case, written in collaboration with a person with ALS.

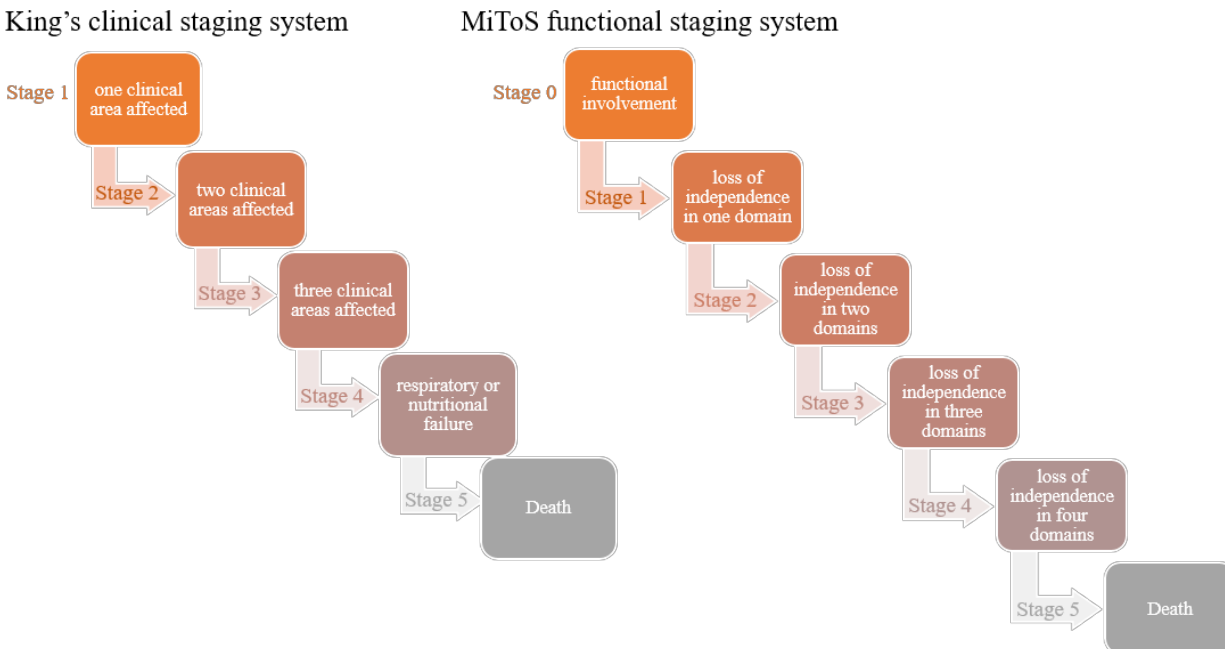
The Presentation of Late Stage ALS

ALS can be discussed according to diagnosis and impairment level, or by function and participation levels. This chapter is grounded in the International Classification of Functioning (ICF) framework (World Health Organization, 2001), using a functional classification.

Individuals with the diagnosis of ALS will be referred to as PALS (persons with ALS). The needs and technologies available for PALS in late stages of the disease follow.

There are many staging processes and group classifications that capture ALS disease progression and the resulting functional impairments of activities in daily living. In clinical neurology, late stages of ALS are defined in terms of how many system regions or system functions (respiratory, swallowing, movement, communication) are impacted by the disease. For instance, scales for staging the disease progression using four to five steps range from no functional impairment to death (Chiò et al., 2015; Roche et al., 2012). King's clinical staging system stipulates five stages based on clinical regions of involvement ranging from stage 1, involvement of one clinical area (e.g., movement, breathing, swallowing, communication), to stage 5, death (Roche et al., 2012). The MiToS functional staging system also includes 5 stages, but is based on loss of independence rather than neurophysiology (Chiò et al., 2015). The six discrete steps progress from stage 0, functional involvement (but not loss of independence) in a single domain, to stage 5, death. Figure 7–1 (adapted from Fang et al., 2017) compares the two rating scales. Late ALS, or the stage prior to death in these frameworks, is defined as either a need for gastrostomy and respiratory support (ventilation) (Roche et al., 2012) or as loss of independent function in the following four domains: swallowing, communicating, breathing, and movement (Chiò et al., 2015).

Figure 7–1. This figure depicts two ALS staging scales from neurology literature that captures physiological changes associated with the disease and resulting functional impairment. This figure is adapted from a scientific journal article comparing the scales (Fang et al., 2017). King's clinical staging system stipulates (right) five stages based on clinical regions of involvement, ranging from stage 1, involvement of one clinical area (e.g., movement, breathing, swallowing, communication), to stage 5, death (Roche et al., 2012). The MiToS functional staging system (left) progress from stage 0, functional involvement (but not loss of independence) in a single domain, to stage 5, death (Chiò et al., 2015).



Within the field of communication sciences and disorders, PALS have been grouped by the level of reliance on AAC supports secondary to loss of speech, hand, or mobility functions associated with disease progression. Using a six-group model, Yorkston and colleagues (1993) classify PALS according to speech, hand functioning, and mobility. The six groups are:

Group 1: Adequate Speech and Adequate Hand Function. This group is comprised of individuals with either no detectable motor speech disorder, a motor speech disorder without changes to speech intelligibility, or a motor speech disorder with reduced speech intelligibility that does not require the use of AAC. Hand functioning is considered to be adequate to accomplish self-help tasks with minimal assistance.

Group 2: Adequate Speech and Poor Hand Function. This group is composed of individuals with either no detectable motor speech disorder, a motor speech disorder without changes to speech intelligibility, or a motor speech disorder with reduced speech intelligibility that does not require the use of AAC. Hand functioning to support written communication is severely impaired.

Group 3: Poor Speech, Adequate Hand Functioning, Adequate Mobility. This group is composed of individuals with a motor speech disorder characterized by poor speech intelligibility and reliance on AAC. Hand functioning and mobility are adequate to accomplish activities of daily living with minimal assistance.

Group 4: Poor Speech, Adequate Hand Functioning, Poor Mobility. This group is composed of individuals with a motor speech disorder characterized by poor speech intelligibility and reliance on AAC. Mobility is impacted for long distances and individuals require the use of an assistive device (e.g., cane, wheelchair, walker). Hand functioning is considered to be adequate to accomplish self-help tasks with minimal assistance.

Group 5: Poor Speech, Poor Hand Functioning, and Good Mobility. This group is composed of individuals with reduced speech intelligibility, requiring the use of AAC. Hand functioning to support written communication is severely impaired. Ambulation is independent without assistive equipment (e.g., cane, wheelchair, and walker).

Group 6: Poor Speech, Poor Hand Functioning, and Poor Mobility. This group is composed of individuals with a motor speech disorder characterized by poor speech intelligibility and reliance on AAC, hand functioning to support written communication that is severely impaired, and mobility difficulties that require the use of assistive equipment (e.g., cane, wheelchair, and walker). This is the population that this chapter is focused around given the significant complex access challenges that are posed by functional changes at this stage.

What are the clinical challenges presented by PALS in group 6? Below, Rich is described as an example of a PALS presenting in Group 6. Afterwards, motor, vision, cognition and language, and psychosocial conditions common to PALS in Group 6 are reviewed in order to determine how best to determine consistent and reliable access methods for these individuals.

One PALS' Challenge

Rich is a 40-year-old retired physician who was diagnosed with spinal-onset ALS five years ago. He lives at home with his wife and three school-age children, and receives caregiving help from in-home nursing staff and other family members who live nearby. Soon after his diagnosis, he had difficulty with computer access and began using a head mouse and speech-recognition software instead of a mouse and keyboard.

Approximately two years later, Rich received a tracheostomy to provide full-time invasive respiratory support and was no longer able to speak. A speech-language pathologist helped him obtain a speech-generating device, which he controlled using the head mouse. As his ALS progressed, Rich's head control became increasingly unreliable. He fatigued quickly and made frequent errors when using the head mouse with his SGD. Rich and his family wondered whether there were other alternative access options that would work better for him, and how he could continue to communicate as his motor function deteriorated.

Challenge: What factors need to be considered to help Rich communicate effectively at this stage in his disease process?

Motor Challenges in ALS

Changes in motor function, with upper and lower motor neuron signs and symptoms, are clinical hallmarks of ALS. Presentation and progression vary considerably from person to person. A majority of PALS present with spinal-onset ALS, in which the initial symptoms involve weakness or stiffness in the upper or lower extremities. The alternative presentation is bulbar-

onset ALS, in which speech or swallowing difficulties develop first (Kiernan et al., 2011). Weakness and atrophy, stiffness, slowness and incoordination lead to difficulties with hand and/or arm movements that affect performing activities of daily living and manipulation of small objects, typing, or writing. Regardless of the type of onset, ALS is a progressive disease, and therefore, its characteristic upper and lower motor neuron changes will eventually spread to other muscle groups (Borasio & Miller, 2001). Eventually, most or all voluntary motor function is affected, though eye movement is typically preserved (Mitsumoto & Rabkin, 2007). Motor speech skills, as part of motor function changes in ALS, decline during the disease progression (Green et al., 2013). According to Ball, Beukelman, and Pattee (2004) 95% of the people with ALS in the Nebraska ALS Database have reduced speech intelligibility at some time prior to death.

As a result of these progressive changes, many PALS use multiple access methods for AAC or other technologies over the course of the disease. Individuals with bulbar-onset ALS may lose the ability to speak early on, but still may be able to type effectively on a physical keyboard or touch screen. By contrast, those with spinal-onset ALS may maintain some functional speech for months or years, but require alternative access such as a joystick, head mouse, or eye tracker to type on a computer. By the later stages of the disease, most PALS will experience severe impairments in both speech and limb function. Some eventually progress to a locked-in state, in which only eye movements are preserved, or a completely locked-in state, in which all voluntary motor function is lost (Murguialday et al., 2011). These individuals require AAC to meet their communication needs, but may have limited options for alternative access due to their reduced motor capabilities.

Cognition and Language Challenges in Late Stage ALS

Cognitive and behavioral changes in PALS have been consistently documented (Elamin et al., 2013; Goldstein & Abrahams, 2013) and are accepted as common features of the disease. Recent estimates note that approximately 50% of PALS experience cognitive impairment, of whom 15% present with frontotemporal dementia (FTD) (Crockford et al., 2018). Changes vary widely and may range from mild impairments detected on neuropsychological testing, to profound FTD. Executive function, behavior, and language are the most likely areas to be involved (Achi & Rudnicki, 2012). Clinicians report that if PALS pursue all treatment measures to live as long as possible, the risk for developing FTD is higher. Crockford et al. (2018) conducted a large multicenter observational cohort of 161 patients with ALS and report that cognitive deficits and behavior impairment are more prevalent in severe disease stages, and by the end stages of the disease, only a small percentage of PALS present with no neuropsychological impairment. The impact of cognitive impairment affects both acceptance of, and ability to learn, initiate, and use AAC (Ball et al., 2007).

It is very difficult to determine if language functions, separated from cognition, are affected in late stage ALS, because motor skills, including oral motor speech skills for responding and upper extremity motor skills for pointing or typing, are significantly impacted. The ALS Functional Communication Scale lists seven communication abilities and activities that change over the course of the disease progression (<http://www.amyandpals.com/als-communication-scale/>). The scale was designed to document, longitudinally, the cognitive communication skills to be targeted as treatment goals for intervention, from initial diagnosis through late stage of the disease.

Table 7–1. ALS Functional Communication Scale (Roman, 2004)

The ALS Functional Communication Scale developed by Amy Roman (2004) lists seven communication abilities and activities that change over the course of the disease progression (www.amyandpals.com/als-communication-scale/). The scale was designed to document, longitudinally, the cognitive communication skills that should be targeted as treatment goals for intervention, from initial diagnosis through late stage of the disease.

See next page for table

Communication Ability	Short term goals	Baseline	Projected	Achieved today
1. Alerting/ Emergency	Patient (and caregiver) can demonstrate/describe the method by which patient can alert others, not in his/her immediate environment, to a need or emergency	+/-	+/-	+/-
2. Communication Strategies	Patient (and caregiver) demonstrate patient and partner strategies that improve communication success, efficiency, speed, and reduce fatigue	+/-	+/-	+/-
3. Nonvoiced (Low Tech) Communication	Patient demonstrates the ability to communicate novel messages via spelling or combining words using a low tech AAC method	+/-	+/-	+/-
4. Speech Generation	Patient demonstrates the ability to communicate a novel message with a voice (speech or SGD)	+/-	+/-	+/-

5. Communicate with those at a Distance	Patient demonstrates abilities to use all methods s/he requires to communicate with partners at a distance	+/-	+/-	+/-
6. Independently Set-Up and Customize AAC systems	Patient (and caregiver) demonstrate the ability to independently use, set up, and customize low and/or high tech augmentative communication equipment	+/-	+/-	+/-
7. Prepared for Future Changes	Patient (and caregiver) can describe one or more pro-active strategies designed to prepare for typical changes associated with ALS in speech/access.	+/-	+/-	+/-
Total Functional Communication Score		/7	/7	/7

Vision Challenges in End Stage ALS

It is critical to consider visual skills in PALS, especially in late stages of the disease, since AAC frequently involves visual interfaces (Chen & O’Leary, 2018). PALS who choose mechanical ventilation, thereby prolonging life in the face of continuing deterioration of motor function, may rely on eye gaze or BCI when other muscle groups are no longer capable of serving as control sites. Extraocular motor neurons are often spared until late stage ALS. There are some reports, however, of visual impairments presenting in ALS. Moss and colleagues (2012) compared neuro-ophthalmic evaluations of 37 PALS followed in a multidisciplinary ALS clinic with matched participants in the control group without disabilities. They found that visual acuity was lower in PALS versus participants in the control group. Some PALS also presented with gaze impersistence, moderately to severely restricted voluntary up gaze, and moderate-to-severe eyelid opening apraxia, or severely saccadic horizontal smooth pursuits. Additionally, oculomotor abnormalities, including smooth pursuit eye movements, optokinetic nystagmus, and visual suppression of vestibular nystagmus were noted in a sample of nine patients with ALS in the early stages (Ohki et al., 1994). AAC clinicians should seek assistance from professionals who can conduct a visual screening and a comprehensive ophthalmologic evaluation when appropriate when considering eye tracking and eye gaze as an access technology in late stage ALS (Fried-Oken et al., submitted).

Psychosocial and Emotional Challenges in Late Stage ALS

Coping with the late stages of ALS, where activities of daily living are significantly restricted by the functional limitations of the disease process is challenging both for PALS and their families.

In a large sample of PALS, 37% of patients reported mild-to-moderate depression symptoms, 13% reported moderate-to-severe depression symptoms, 6% reported severe depression symptoms, and 18% reported clinical-level anxiety symptoms (Wicks et al., 2007). Likewise, in a separate sample, depression and anxiety levels were reported in 20% of the family members of PALS (Lillo et al., 2012). Perceived social support and appraisal of one's coping potential and independence is a predictor of depressive symptom severity for PALS (Matuz et al., 2015). The most important stressors identified by PALS and family members are existential concerns (e.g., worries about the future, loss of faith in religion, worries about disease progression) and physical concerns (e.g., muscle weakness, mobility problems, ADL) (Trail et al., 2004). Caregiver burden affects the psychosocial well-being of PALS and their family members. The strongest predictor of caregiver burden appears to be cognitive/behavior changes (reported between 10% to 40% of sample) rather than physical limitations of the disease (Lillo et al., 2012).

AAC clinicians must consider the significant emotional and psychosocial burdens on PALS and their families that present challenges in the later stages of the disease process. Often, the multidisciplinary team includes a social worker, psychologist, or palliative care specialist who can work with the family and patients at this time. Costello (2009) suggests that common roles for speech-language pathologist on palliative care teams and in end of life care include: (1) supporting autonomy whenever possible; (2) providing care information and encouraging questions; (3) encouraging patients to talk about their experience of illness; (4) supporting self-expression through use of unaided and aided communication strategies; (5) selecting vocabulary for aided communication systems that is personalized and reflects medical, social-connectedness, and psychosocial needs; (6) assisting with message and voice banking; (7) supporting

maintenance of social and emotional ties through communication; and (8) supporting patients to express concerns for those left behind.

Possible AAC Solutions

Finding appropriate AAC technology solutions for PALS and their care providers, even in late stage ALS, remains an important clinical goal. Ball et al. (2004) reported that about 96% of people with ALS in the Nebraska ALS Database who were recommended AAC in a timely manner accepted and used AAC, with only 6% delaying but eventually accepting technology use. Those who rejected AAC presented with compounding severe health challenges, such as cancer, or concomitant dementia (Ball et al., 2004). The availability of access technologies has been shown to improve quality of life for adults with ALS and has the potential to reduce the burden placed on care providers (Hwang et al., 2014). It is important to note that low-tech solutions with different access options are critical to present, and often are preferred by PALS during the final stage of the disease. Possible access technologies (both high tech and low tech) appropriate for AAC for people who experience late stage ALS are discussed below.

Using Eye Gaze Technologies in Late Stage ALS

Eye tracking technologies have been widely used internationally with PALS (Käthner et al., 2015; Pasqualotto et al., 2015; Spataro et al., 2014). The volitional muscles of the eye, innervated by the nuclei of the oculomotor (CN III), trochlear (CN IV) and abducens (CN VI) nerves, typically remain functional longer than muscle groups innervated by upper and lower motor neurons. These technologies range from eye transfer boards (also known as ETRAN)

(Fried-Oken, 2000), and the Speakbook, an eye gaze flip book (Joyce, 2011), to digitized speech devices, such as the Megabee™ Eye Pointing Communication Tablet (*MegaBee: Electronic hand-hand communication tablet*, 2010), and finally to computer-based speech generating devices. All eye gaze-based technologies require PALS to look at different parts of the page or screen, and a partner or the computer speaks out what is selected by eye gaze.

The ETRAN board and SpeakBook both involve printed letters or messages, typically presented on a piece of paper or Plexiglas frame. A communication partner looks at the PALS through a hole in the paper or frame, and observes the PALS' gaze as she or he looks at letters or phrases. A coding system is typically used, for example, looking at the top left corner and then the pink dot selects the pink letter in the top left corner. The Megabee™ is an electronic version of the same concept. A communication partner holds up the Megabee and watches as the PALS looks at colored targets in different locations around the central window, then the communication partner presses a colored button for each selection. The Megabee decodes these button-press inputs and prints the resulting message on a small screen.

High-tech eye tracking technologies that provide direct selection to language on communication devices typically collect data on eye gaze position using infrared pupil-corneal reflection. Devices that utilize these methods often include an eye tracking module with a built in infrared light source and camera. As the user looks at various points on the display during a calibration, the camera detects the changing distance between the pupil and a “glint” or reflection of the infrared light-source from the cornea.

Challenges With Eye Tracking Technologies

What is challenging about using your eye tracker?

“Using eye tracking with dwell click is] mentally and physically stressful, frustrating, and exhausting. There is so much strain involved that it increases my heart rate and respiratory rate. I have to take frequent breaks before continuing.” -Rich

Eye tracking technologies are prone to error at various levels including user eye-conditions, shifting position of the user or device, and environmental factors. For example, changes in the ambient light conditions in the environment can significantly impact performance, and PALS regularly report difficulties accessing their eye tracking speech-generating devices outdoors due to competing infrared light sources. Even when indoors, changing lighting conditions can result in problems with reliable eye gaze use.

Positioning changes also have the potential to significantly alter performance of eye gaze technologies. People with late stage ALS experience regular positioning changes as care providers must shift their positioning to prevent bedsores or manage incontinence. Sometimes the device itself must be moved away from the PALS for care activities. Chen and O’Leary (2018) point out that a device positioned too high will result in eyelid fatigue or eyestrain due to looking upwards, yet an eye gaze device positioned too low may not read the angle of the user’s eyes correctly due the upper lid obscuring part of the pupil from the camera.

Several tools and strategies may support reliable and consistent positioning for use of eye tracking speech-generating devices. Mounting solutions may affix the device directly onto a hospital bed and offer the ability to swing away to a locked position to address the PALS’ medical needs, then to swing back and lock in the original position. Some PALS benefit from a rolling-floor mount that allows the device to be positioned more flexibly. A challenge of these tools is to return them to the same position after being removed. Historically, clinicians adjusted

multiple levers to approximate the necessary position. Novel float arm mounts have been developed that allow the clinician or caregiver to easily move the device into the necessary position without adjusting multiple levers. Although current mounting solutions are somewhat limited, emerging solutions are in development that may detect a user's head position and adjust the position of the device to match.

Person-centered factors may also result in reduced reliability in eye gaze technology for PALS. Many PALS who utilize eye gaze technologies use a dwell-to-select method of button selection. That is, PALS will look at a button for a predetermined period of time that causes the button to activate. For those using this method, Chen and O'Leary (2018) suggest an ideal dwell time of 0.5 seconds for general eye tracking use, but optimal dwell times have not been explored for PALS. It is likely that PALS with reduced ocular motility will have difficulty directing their gaze from target to target quickly enough to avoid frequent errors with a short dwell time. For example, a PALS who is able to consistently move a body part, such as their finger, toe, jaw, or eyebrow, can activate a switch for a click-to-select method of activation (much like using a computer mouse). As an alternative to activating a switch, a PALS may utilize a blink-to-select activation method. In this case, the user looks at a desired button then blinks their eyes to select it (blinks for selection are longer in duration than normal involuntary blinks). However, PALS often report that this activation method is fatiguing and can become less reliable as the disease progresses.

A critical factor associated with effective use of eye tracking technologies is the quality of the PALS' visual skills as it relates to visual access. Visual skills such as fixation, acuity, ocular motility, binocular vision, convergence, field of vision, and perceptual abilities have all been identified as fundamental for successful use of visually based assistive technology such as

AAC (Federici & Scherer, 2012). Specifically, an impairment of one or more of these visual skills may affect the ability of the PALS to effectively analyze and manipulate visual information in the environment, influencing their ability to also understand, problem solve, and execute a plan when utilizing AAC technologies (Warren, 1993).

Using Switch Scanning in Late Stage ALS

Another common access method to a speech-generating device for PALS is scanning. This is the process where items are presented in a pattern and the user makes a selection indirectly by selecting a switch. Scanning on a speech-generating device involves many features that a clinician has to consider during the feature matching process including number of switches, switch placement, switch type, scanning pattern, and page set organization. These features may all be optimized for a person with late stage ALS.

When considering switch scanning as an access method, three questions must be asked:

1) What muscles have consistent, reliable movements for placement of the switch? 2) What switches are available that can be reliably activated with the residual movement? 3) What type of scanning settings should be offered for indirect selection? Often in late stage ALS, PALS may produce only a single consistent, reliable movement, such as protrusion of the jaw or movement of a thumb, for motor control. Either automatic or inverse single switch scanning may be options. However, due to fatigue associated with keeping a switch in an activated position, inverse scanning is often not a feasible option.

To optimize automatic scanning for those with late stage ALS, it is necessary to consider that these individuals often present with long latencies in motor responses. As such, when utilizing automatic scanning with a PALS with late stage ALS, it may be beneficial to

significantly reduce the scan speed to improve selection accuracy. However, in trading accuracy for speed, it may take PALS significantly longer to express desired messages. When this is the case, clinicians may consider prioritizing high-frequency vocabulary and messages to be presented early in the scan cycle.

To optimize speed, organizing vocabulary by frequency of use may be beneficial. The placement of vocabulary is based on the frequency of their selection with commonly used words or phrases placed such that they will be offered earlier in the scanning pattern, reducing the time required for selection. For example, consider a PALS who utilizes linear automatic scanning. To select the first symbol, the user has to wait only for the first presentation. To select the second symbol, the PALS has to wait twice until the desired symbol is offered. Letter frequency patterns are commonly used with row/column scanning. In quadrant scanning, the user is presented first with a quadrant, then may be presented with a linear scan pattern of individual symbols. For both of these scan patterns, the lowest number of presented stimuli is two. Thus, the most frequently used message should be presented in the upper left of the display. Some quadrant scanning patterns utilize row-column scanning after a selection of a quadrant. As a result, the user is presented with a minimum of three stimuli including the quadrant, the row, and the individual stimulus.

When scanning using a high-tech communication device, many people with late stage ALS experience difficulty selecting items that are presented first (e.g., if row/column scanning, then the first row; if linear scanning, then the first-item). This is often due to minimal time available to anticipate a selection that needs to be made, or having to visually reorient to the scan location after having cycled through all options and returning to the first presented item. As a

potential solution to this issue, many high-tech AAC devices include a feature to increase the scan time of the first presented item or item group.

When it comes to accessing communication devices using scanning, there are a variety of switches that can be used to detect even the most minimal movements. This is particularly critical for late stage ALS as movements can become so limited that they may not close the distance necessary for a proximity switch, or further limited to even a single muscle twitch. Fager, Fried-Oken, Jakobs, & Beukelman (2019) developed a novel access method involving a custom 3-D printed housing containing accelerometers, gyroscopes, and magnetometers. This tool was ‘trained’ such that given several repetitions of intentional and unintentional movements, it could learn when the user meant to activate their switch. These custom-made solutions likely represent the future of switch access technologies, though the availability of these tools is currently limited to participants in research studies.

Switch solutions that clinicians may currently access include mechanical switches, as well as those that detect movements by electromyography (EMG), electrooculography (EOG), proximity sensors, or fiber-optic sensors. The switch solution selected depends on the residual motor control of the individual. For those with the ability to generate a movement, but who lack the strength to use a mechanical switch, proximity switches are often used. These switches require the user to create a movement of a body part within a certain distance of the switch. If the user is unable to generate the movement necessary to activate a proximity switch, an alternative switch type may include fiber-optic switches. These switches emit a beam of light from the end of the switch. The clinician may select to have the switch activate when the beam of light is interrupted, such as the user moving their finger into the light), or when the beam of light is

uninterrupted, such as the user keeping their finger in the light and moving it out to make a selection.

An EMG switch may be a solution for PALS with single muscle control. The EMG switch is placed over the area where the muscle fiber is active. Once the PALS initiates activation of the muscle, the switch is closed. Alternatively, for individuals who have residual eye gaze movements, but with conditions such as ptosis interfering with accurate eye gaze tracking, an EOG switch may be used.

Assessing the appropriateness of a particular switch type can be challenging for clinicians. Even for clinicians who regularly practice with PALS who have complex access needs, objective assessments of switch access are often not completed. Clinicians may develop their own criterion-referenced measures, evaluating reliable switch access to some percentage of acceptability. However, this can be a very time-consuming process, and if not completed, can result in low reliability of switch activation. Koestner and Simpson (2019) recommend the use of Scanning Wizard software, an iOS application and web-based application used to improve the setup of switch scanning systems.

Scanning Wizard software guides the PALS as well as the clinician through a series of tasks evaluating the user's (1) optimal switch placement/set-up, (2) optimal scanning time, (3) the need for an extra delay during scanning tasks, (4) optimal scanning initiation pattern (automatic or manual), (5) scanning loop count, and lastly (6) optimal keyboard setting. A small study of ten people with severe physical and communication needs demonstrated that use of Scanning Wizard software was an effective resource to objectively improve the configuration of their scanning system (Koester & Simpson, 2019).

Challenges to Switch Scanning

Extremely slow rates of selection, leading to reduced speed of communication, offer the greatest challenge to PALS who rely on switch scanning. Even when more than one switch is used, the rate of message generation does not come close to directly selecting letters on a keyboard (Koester & Arthanat, 2017). Many PALS reject scanning because of this challenge. Additionally, poor switch placement, unintentional movement of the switch or the body part used to activate it, and difficulty finding an adequate mounting solution at the switch site also cause frustration. Currently, most switches do not offer any feedback or information to support problem solving when challenges arise. If there is a problem in the setup, it is difficult to know if it is at the SGD level, the switch level, or the PALS level. With this degenerative disease, it is difficult to know when to change switches or locations, and often failure is the only indication, leading to frustration and even abandonment of the SGD. There are many creative and unique switch placement solutions, where the clinician, family, and PALS experimented with materials to increase the reliability of switch placement or mounting reliability. The AAC clinician, working with PALS with late stage ALS will need to consider many different solutions for switch use if the PALS agrees to stick with this access technology.

Using Multimodal Access Technologies

Although access methods have been presented in isolation, it is critical to recall that many PALS will use multiple input methods across the day. For example, the PALS who has strong eye gaze function in the morning and worsening ptosis across the day might utilize eye gaze access in the morning and scanning in the afternoon and evening. Further, if switch access movements

become unreliable, partner assisted auditory scanning may be another method to express desired messages without the operational demands of a high-tech communication system.

Historically, AAC devices have only been designed to expect input from a single access method (e.g., eye gaze only, switch access only). There are significant challenges associated with the use of only a single access modality to access a communication device, including fatigue, over-use injuries, change in positioning, and inefficiency (Fager et al., 2019). In complex access challenges such as in late stage ALS, users often require the use of more than one access method to reduce fatigue, conserve energy, and to optimize the speed and accuracy of the communicative exchange.

Fager and colleagues at the Rehabilitation Engineering Research Center on Augmentative and Alternative Communication propose the development of systems that can expect input from more than one access method, such as voice recognition and typing, or single switch scanning and eye gaze (Fager et al., 2019). Fager, Beukelman, Jakobs and Hosom (2010) have designed a typing system where an individual first uses eye gaze to locate a target on the screen (whether that is a group of symbols or single symbol), activating a switch to begin a scan thereby switching from eye gaze to switch scanning, and then activating the switch a second time to select the target. This type of system accounts for difficulties with dwell activations such as reduced range of motion and precision in ocular movements.

Multimodal access methods are currently available in some commercially available dedicated speech generating device systems. This allows for trialing of possible combinations of access methods with people with late stage ALS. Important considerations for trialing include (1) accuracy and efficiency of a multimodal access method in comparison to a single access method alone; (2) effort and fatigue associated with a single access method in comparison to a

multimodal access method; (3) cognitive demands of a multimodal access method in comparison to a single access method; and (4) user preference. Implementation of multiple access methods also has considerations for clinical practice, especially in regards to care team training, such as identifying with a PALS and their care team when specific access methods are challenging (due to time of day, circumstantial considerations, medication, pain, etc.), coming up with communication strategies to request assistance for changing an access method, and training the care team in positioning and implementation of multimodal access methods.

Challenges to Multimodal Access Technologies

As multimodal access options are relatively new within AAC technologies, many speech-generating devices currently have limited access to these features. It can be difficult to determine the advantages of one access method over another given the complex inherent variability in the contributing factors to access challenges (e.g., fatigue, medication, pain, alertness, mood). This can also make it difficult to determine when and if it is appropriate to change access methods. There is not yet software with dynamic recognition of these changes in functioning throughout the day that might be used as a tool by a clinician to change access methods or to prompt the individual with the option to change access methods themselves.

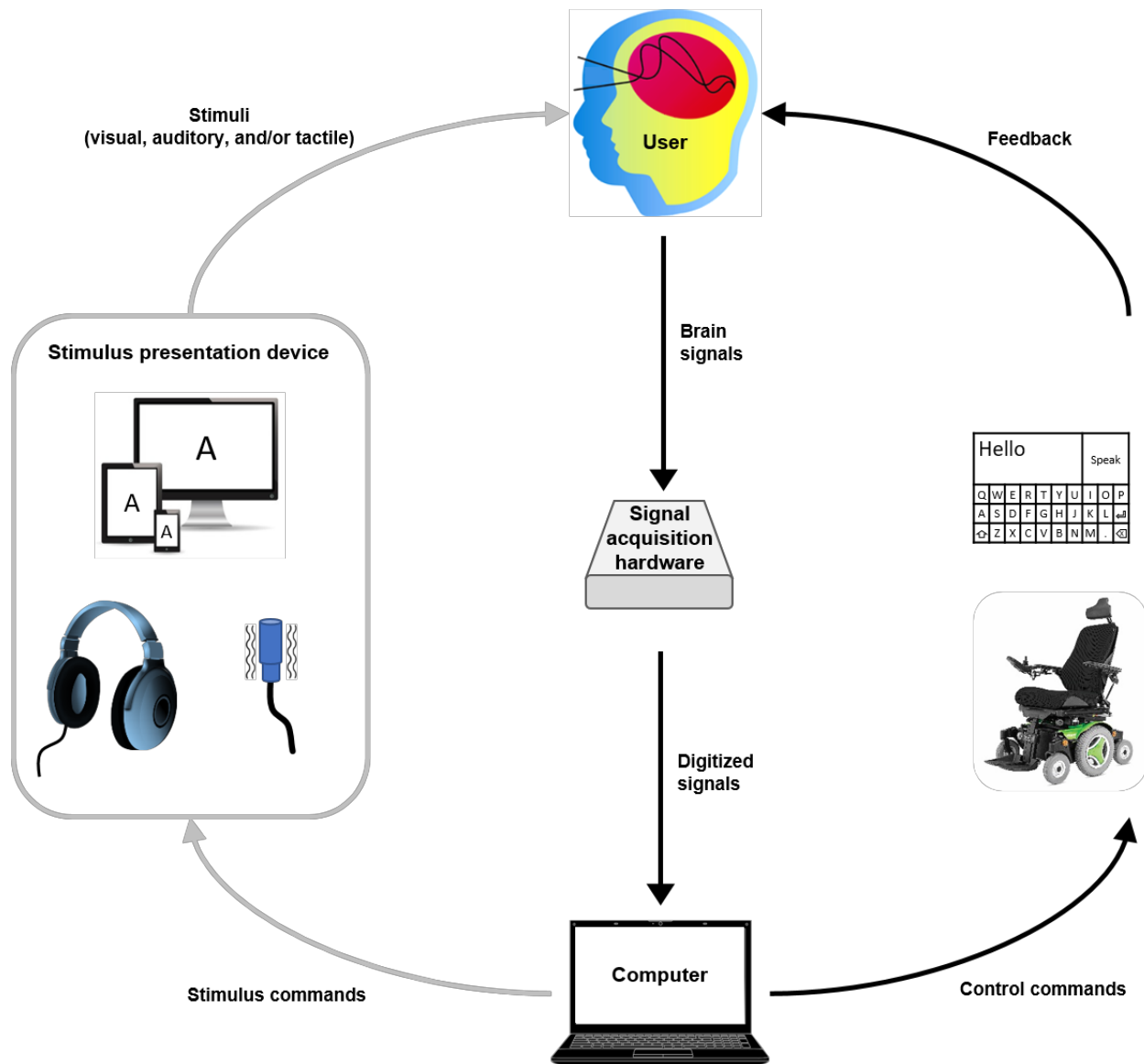
The Potential of Using Brain-Computer Interface (BCI) in Late Stage ALS

“If I could converse as close to normal as possible, I could be myself and express my personality much more than I’m currently able to. I hope that the BCI system can

accomplish that. Otherwise, my dreams will stay as dreams.” —Rich

Some individuals with ALS eventually will lose all voluntary motor function, leaving them unable to use eye tracking or switch scanning for AAC access. Brain-computer interface (BCI) technology is being investigated as a new alternative access method that bypasses the motor system, supporting computer control with brain signals alone. As the name suggests, a brain-computer interface system involves three main components: (a) the brain, (b) the computer, and (c) the interface. The user, of course, supplies the brain and the intention to control an assistive technology. The computer may be a laptop, desktop, tablet, or even a smartphone, and runs software that analyzes brain signals and converts them into signals for controlling the desired application. The interface is the means of recording the brain signals and sending them to the computer. For most noninvasive BCIs, this involves electrodes that detect brain activity, and hardware that amplifies the signals and converts them to a format that can be processed by the computer. Figure 7–2 provides a schematic of the basic design of a BCI for AT control.

Figure 7–2. This figure provides a schematic of the basic design of a BCI for AT control. A BCI system involves three main components: (a) the brain, (b) the computer, and (c) the interface. The user, of course, supplies the brain and the intention to control an assistive technology. The computer may be a laptop, desktop, tablet, or even a smartphone, and runs software that analyzes brain signals and converts them into signals for controlling the desired application. The interface is the means of recording the brain signals and sending them to the computer. For most noninvasive BCIs, this involves electrodes that detect brain activity, and hardware that amplifies the signals and converts them to a format that can be processed by the computer.



In some BCIs, auditory or tactile stimuli are presented to the user via a presentation device (e.g., display, headphones, or tactors), and the user's reactions produce brain signals related to the intention or lack of intention to select a given stimulus. This is analogous to automatic switch scanning, with brain activity taking the place of muscle activity that would typically be used to activate a switch. Other BCIs require the user to spontaneously produce brain signals (e.g., imagined movements of the left and right hands to move a cursor left and

right). Depending on the system, control may work similarly to either multiple-switch scanning or to cursor control with mouse emulation, again with brain signals replacing movement-based signals. In either case, the brain signals are collected and digitized by signal acquisition hardware and sent to the computer for analysis. If the computer has sufficient evidence to infer the user's intent, it sends a control command to the AT software or device. If not, it awaits additional information from the user. In many systems, the user simultaneously receives input from the stimulus presentation device and feedback from the AT.

Brain signals for BCI may be acquired either invasively or noninvasively. Invasive BCI systems require surgery to implant an electrode array directly onto the cerebral cortex. Noninvasive systems capture brain signals from outside the scalp using a variety of methods. Electroencephalography (EEG), which involves electrodes placed against the scalp, often in a cap or headset, is the most common method of noninvasive brain signal acquisition. Research studies have investigated the use of BCI to control both integrated, custom-built BCI communication software and off-the-shelf, commercially-available AAC programs (Gosmanova et al.,(no et al. list all 4 authors) 2017 Thompson et al.,(no et al. list all 3 authors) 2014). Below, some of the invasive and noninvasive BCI systems that have been explored for use by PALS are reviewed.

A variety of brain responses have been used as control signals in noninvasive BCIs for AAC (Akcakaya et al., 2014; Brumberg et al.(no et al. list all 4 authors)., 2019). Many rely on the detection of the P300 event-related potential, a positive change in EEG signals elicited by a rare or unique stimulus in a stream of stimuli. Perhaps the best-known P300-based BCI is the P300 speller that is based on Farwell and Donchin's (1988) original mental prosthesis. The P300 speller interface consists of a matrix of letters and other symbols or words (e.g., space,

backspace, or commands for computer control), with rows and columns (or individual characters) flashing in random order. The user focuses on the desired symbol and mentally counts each time it flashes; the flashing of the desired symbol should elicit a P300 response. Each row or column is flashed multiple times in order to elicit repeated P300 responses, providing enough information for the computer to determine the desired letter (Farwell & Donchin, 1988). As of this writing, the BCI communication systems currently available to consumers rely primarily on the P300 paradigm.

A number of studies have demonstrated successful use of a P300 matrix speller by individuals with ALS (Guy et al., 2018; Schettini et al., 2015; Silvoni et al., 2013; Thompson et al., 2014). In a study of long-term independent home use of a P300 BCI system, PALS and their caregivers reported that the benefits outweighed the burdens, and most participants elected to keep the system at the study's end (Wolpaw et al., 2018). Another study demonstrated that some PALS can maintain the ability to operate a P300 BCI over a period of several years (Silvoni et al., 2013). In addition to spelling interfaces, PALS have used BCI systems (using the P300 and/or other brain responses) to answer yes/no questions, activate emergency call bells (Lim et al., 2017), select predetermined phrases (Hwang et al., 2017), and create visual art (Zickler et al. (no et al. list all 5 authors), 2013).

PALS also have participated in invasive BCI research studies, though these are less common due to the need for surgical implantation of the electrodes. BrainGate is a long-term, multicenter project exploring invasive BCI for communication and computer control. As part of the BrainGate program, individuals with ALS and other conditions have intracortical micro-electrode arrays implanted directly onto the motor cortex. Study participants, including PALS, have successfully used this invasive BCI system to control a cursor for text-based

communication and other applications on a commercial tablet (Nuyujukian et al., 2018). Another research group has reported on a PALS who successfully used her invasive BCI system in her home over a period of 3 years (Pels et al., 2019; Vansteensel et al., 2016).

Challenges to BCI Research and Development

“When BCI reaches a more advanced stage, my main concern would be the placement of the cap, the electrodes and calibration of the equipment to make it effectively functional. The current system, in my opinion, is practically impossible to use by patients without significant assistance.” —Rich

Several studies have found that some PALS are unsuccessful with BCI communication systems (McCane et al., 2014; Wolpaw et al., 2018) though it is unknown whether these same PALS might perform better with a different type of BCI or interface. Studies involving both PALS and healthy control participants often demonstrate significant differences in results between the two groups, with better BCI performance for participants in the control group (Geronimo et al., (no et al. list all 3 authors) 2016; Ikegami et al. (no et al. list all 5 authors), 2014; Oken et al., 2014). However, this performance gap is absent in other studies (e.g., McCane et al., 2014; Thompson et al., 2014). Individuals with ALS may take medications that affect attention, vigilance, or brain signals (Meador, 1998; Polich & Criado, 2006). Fatigue, cognitive changes, or visual impairments associated with ALS may also affect BCI performance (Geronimo et al., 2016; McCane et al., 2014). Disease progression or overall level of disability (as measured by the ALS Functional Rating Scale-Revised) does not appear to correlate with BCI performance (McCane et

al., 2014; Silvoni et al., 2013), and studies involving PALS in a locked-in or completely locked-in state have produced mixed results (De Massari et al., 2013; Okahara et al., 2018).

In addition to patient factors, system as well as environmental factors may present potential barriers to successful clinical implementation of BCI technology. Many noninvasive systems use wet electrodes, which require a small amount of gel to be added to each electrode to improve signal quality. This gel can be a problem for users who have their hair washed infrequently. Dry electrodes can have inferior signal quality and may cause discomfort for some users. The setup, configuration, and electrode application required for BCI use may be complicated and challenging for caregivers and clinicians (Peters et al., 2015). PALS at end stages of the disease may have electrical equipment that interferes with the data acquisition system of the BCI system. The technical support required by BCIs that are being developed and evaluated is not trivial, as these are sophisticated devices. The coordination required between the signal acquisition hardware and software, the signal processing software, and the assistive technology outcomes can cause challenges at many levels (Chavarriaga et al., 2017). Having a knowledgeable team is of paramount importance if this new access technology is going to be available for PALS. Comfort, ease of use, training for PALS, caregivers, and clinicians must be addressed to facilitate clinical implementation of BCI technology.

The Future for Access Technologies in Late Stage ALS

Access to AAC technologies is challenging in late stage ALS due to the complex interaction of contributing disease factors. Although recent advancements in technology have made AAC systems more accessible, additional research and development of AAC/AT tools for increasing access to technology in this population are needed.

The need is particularly great in increasing the flexibility and adaptability of existing technology to meet the complex access needs of this population. Proposals for new advancements in access technologies are grounded in a multi-modality theory of AAC using multiple input signals (e.g., eye gaze, EMG, EEG, switch, direct selection) at one time. Possible advancements include: (1) increasing machine learning techniques in AAC software (active querying) with the goal of improving the system detection of selection versus non-selections; (2) optimizing multimodal signal fusion to dynamically use the most reliable physiological intent evidence by a user, including brain-based signals (EEG, SSVEP, P300, VEP, SMRS, EOG), neural impulses given by muscle activation (EMG), eye gaze (position, velocity of movement), head tracking and other inputs; and (3) providing new vocabulary sources to SGD language models based on partner input to personalize language enhancements and reduce message generation time for functional communication.

The ethics of offering access technologies to PALS in late stage ALS must be considered by the AAC clinician and treating team. Earlier in this chapter, it was questioned whether failure with an access technology is the only way to know that it's time to change that option and explore others. It is paramount to honor PALS preferences in this process with the role of the speech-language pathologist to present information and answer questions. Are there other ways to determine best fit that may not lead to frustration and abandonment even for a clinician with limited experience in AAC?

Decision-Making Guidelines

Up to this point, this chapter has presented access technology options and challenges. What follows is a list of clinical questions that should be considered when making decisions about

which access technologies to introduce to PALS in late stages of the disease. Many of these guiding questions should be asked during every full AAC evaluation. For late stage ALS access questions, the chapter focuses on unique, specific considerations for PALS. The questions are presented in three categories: (1) PALS-focused, (2) environment-focused, and (3) clinician-focused skills and competencies. The rule here is flexibility and creativity. PALS in late stage ALS present with complex communication needs that are not easily addressed with a standard intervention timeline. A basic knowledge of disease history, motor progression and timing, the PALS's trajectory, past treatment choices, and reported functional variability are the principles that guide decision making. Throughout the decision-making guide, an illustrative case example is highlighted using an individual with late stage ALS.

I. PALS-Focused Considerations

- What are the PALS' **goals** for communication and alternative access?
- What is the PALS' **medical status & plan of care**?

Marty is a 60-year-old man with late ALS. Approximately 12 years ago, he noticed instability in his balance, followed by mild dysarthria a few months later. He received a formal diagnosis of ALS one year after his initial symptom presentation. The following year, he elected to undergo tracheostomy and has used mechanical ventilation since then. He relies on AAC to meet his communication needs in all situations, including social interaction, maintaining relationships with family and friends, interacting with health care providers, and directing his everyday care.

- What are the PALS' **motor skills**? Consider history, timing of progression, trajectory, and variability.
- What are the PALS' **hearing acuity** and **auditory skills**? Consider history, timing of progression, trajectory, assistive technologies (e.g., hearing aids) and variability.
- What are the PALS' **visual acuity and oculomotor skills**? Consider history, timing of progression, trajectory, assistive technologies (e.g., single vision or bifocal glasses) and variability.
- What are the PALS' **cognition & language skills**? Consider history (including cultural and linguistic considerations), timing of progression, trajectory, and variability.
- What is the PALS' **emotional/ psychosocial** status? Consider relationships with family and friends, stability of care team and staff turnover, and familiarity with technology.

Marty now has limited remaining voluntary motor function, but can produce small movements of his eyes, chin, and left thumb. These movements are restricted in range and have significant (>1 second) and variable latency. Marty uses horizontal eye movements to give yes/no responses, but has limited vertical eye movement. He can protrude his chin slightly to activate a proximity switch to control a calling device to alert care providers he needs assistance, though the switch requires frequent repositioning due to trach-care activities or small, unintentional changes in Marty's head position. Finally, Marty can activate a proximity switch by abducting his left thumb, though this switch also requires frequent repositioning. His eye

movements and switch activations movements become slower and more unreliable with fatigue.

Marty's cognitive and language skills have remained relatively stable since his diagnosis. However, members of his care team have recently noticed inconsistency with yes/no responses, increased latency in following spoken directions, and occasional lack of response to care provider's questions. Although it is difficult to formally assess cognitive status at this stage given Marty's significant motor impairment, these changes represent a noticeable difference when compared to his skills a few months ago. Marty's team is continually asking whether his inconsistent lack of response is due to FTD progression, changes in motivation, hearing, or some other communication breakdown.

Marty has reduced visual acuity and wears glasses during all of his waking hours, for both distance and reading. They often slide down the bridge of his nose and must be adjusted by a caregiver. Marty also reports light sensitivity and intermittent double vision, requiring modifications to font sizes, contrast, and brightness of the SGD screen. He has bilateral hearing loss that is corrected with hearing aids, which he wears consistently with support from his care providers.

Marty is known to have an upbeat personality and has had many years to adjust to his diagnosis and the changes in his physical function. In the past, he has welcomed information and advice from his medical team and has taken a proactive approach to symptom management and decisions about his care. However, he is wary of making changes to his current SGD or access method and is not always interested in trying new things. It is critical to continually evaluate the input and value of changes or

suggestions made by every team member (including family, clinicians and care team), and to honor Marty's choices.

- What is the PALS' **familiarity with and previous use of technology** and assistive technology?
- What **communication platforms** (face-to-face, social media, texting, email, etc.) does the PALS use now and plan to use in the future?
- What are the PALS' **message needs** now and what might they be in the future?
- What are the PALS' **positioning needs** and what equipment are they using to address these needs now and for the future?
- What **access methods** were used in the past and present, and when/how are they changed?

Marty is a retired information technology professional with a longstanding interest in computers and technological innovations. Before his diagnosis, he used computers daily for work, communication, entertainment, social interaction, shopping, managing finances, and other purposes. Currently, he is primarily interested in using his speech-generating device for face-to-face communication and email. He frequently uses both generic and customized prestored messages for social interaction and directing his care, but also requires access to a keyboard for novel messages. At one point, Marty used his speech-generating device to type an eight-page caregiver manual to direct his care and communicate his preferences. In addition to communication, he would like to use his speech-generating device to

access the Internet and control his TV. For many years, Marty has used yes/no responses and partner-assisted scanning as additional communication methods.

Marty spends his time either lying in bed with his head elevated, or in a tilted and reclined position in his power wheelchair. He is unable to drive the chair, which is fitted with attendant controls. His speech-generating device is mounted to his hospital bed on an arm that can swing aside or be removed entirely to provide care providers with easy access to Marty, and to other medical equipment when needed. One proximity switch is positioned near his chin on a flexible arm, while another is near his left thumb, attached to a custom-made foam arm support. A second speech generating device mount is attached to Marty's wheelchair, though he often prefers to use partner-assisted scanning when seated in the chair to avoid the inconvenience of moving and positioning the speech generating device and switches.

Marty received his first speech-generating device soon after his ALS diagnosis, and accessed it using direct selection on the touch screen. After his tracheostomy and significant decline in motor function, he was no longer able to activate the touch screen, and accessed the speech-generating device with a USB joystick. As his motor function continued to decline, he trialed both eye tracking and single-switch scanning. Marty experienced inconsistent accuracy with eye tracking, apparently related to both his glasses, and reduced ocular motility. He preferred single-switch scanning with hand movements, and used this method reliably for several years, with periodic changes to the switch (e.g., mechanical switch to proximity switch) and its positioning.

After several years of successful switch scanning, Marty's switch activation became increasingly unreliable, and his speech-language pathologist initiated trials of both eye tracking and a BrainFingers™ switch. BrainFingers™ activation was inconsistent, and this was ruled out as an access method. Eye tracking was also inconsistent, but was both faster and more reliable than switch scanning with the thumb switch. Marty's speech-language pathologist customized an eye tracking page set to accommodate his reduced tracking accuracy. He continued to prefer switch scanning, but began using eye tracking as a backup access method at times when he had difficulty activating the switch.

II. Environment-Focused Considerations

- Who are the members of the PALS' **social circle and care team**? Consider level of familiarity with communication partners for each member of team, level of support for communication provided by team members including communication advocacy, presence, or lack of communication partner training for members of PALS circle, and other social factors contributing to communication from members of social circle.
- Consider the **impact of the physical environment on access technologies** (e.g., light sources and reflective surfaces for eye gaze, electrical "noise" for brain-based signals, and insufficient space for all DME, difficult environment for attending to competing sensory sources to use speech-generating device).

- Consider the **technical support** available for AAC technologies through manufacturers and other sources and the competence of members of the PALS' social circle or care team to program, set up, or troubleshoot assistive equipment.

Marty currently lives in an adult foster home specializing in caring for people using mechanical ventilation. His wife, daughter, and extended family members and friends visit him often. His care team includes the foster home nursing staff, a home health speech-language pathologist with expertise in AAC, and other home health care providers. His family, friends, speech-language pathologist, and the foster home staff are well versed in interpreting Marty's unaided communication signals (eye movements for yes/no responses, as well as facial expressions) and in repositioning his speech generating devices and switches as needed. The foster home staff often helps facilitate interaction between Marty and less familiar communication partners.

Marty spends much of the day lying in a hospital bed or seated in his power wheelchair in his private room. The room is small and must accommodate an array of medical equipment such as a medicine cabinet, ventilator, enteral feeding pump, charting station for caregiving team, and suctioning machine, while allowing adequate space for a Hoyer lift for transfers. Marty's speech generating device-mounting system must be low-profile and easy to swing aside or remove entirely to facilitate daily care activities. Reflective surfaces, including switch mounts and IV poles, have at times had to be moved from behind his bed or modified to reduce

interference during use of eye gaze access. His room is dimly lit given his light sensitivity.

Speech generating device technical support is available by phone or e-mail from the device manufacturer. Marty also has a device company representative that lives locally and is available for in-person trainings and technical support as needed. Marty's home-health speech-language pathologist visits him regularly. She has extensive experience and expertise in AAC, and often works with individuals with complex access needs. The foster home staff is committed to maintaining Marty's communication access and has received training in positioning and troubleshooting the speech-generating device, mount, and switches. Laminated signs are positioned around his room to communicate his yes/no response ("I look left for 'yes' and right for 'no').

III. Clinician-Focused Skills and Competences

- Does the clinician have competence in **disease-specific knowledge** about ALS to provide disease education and counseling regarding both AAC and care structures and systems (e.g., palliative care, hospice, team discussion, and support groups)?
- Does the clinician have proficient knowledge of the **use, programming, and modification of AAC technology**? What training or mentorship is required to gain proficiency?
- Does the clinician have **knowledge of health care systems** (e.g., insurance funding, alternative funding sources, loaner equipment, and timing of funding) in order to obtain

equipment and appropriate related support services ideally within an interdisciplinary care team and model)?

Soon after his diagnosis, Marty began receiving care through an ALS Association Certified Treatment Center of Excellence. He received education about AAC options even before his speech had changed, and his speech and communication were monitored at clinic visits every three months. When he began to experience dysarthria, he met with a speech-language pathologist employed by the ALS Association, who completed an AAC evaluation, pursued insurance funding for a speech-generating device, and provided support with training and device programming. After his tracheostomy and changes in his physical function and access needs, Marty worked with a home health speech-language pathologist who specializes in AAC evaluation and treatment.

Both speech-language pathologists were knowledgeable about ALS, AAC (including the latest technology for speech generating devices and alternative access), and the systems involved in obtaining and funding AAC equipment. Both reported gaining expertise in these areas through self-study (including textbooks, journal articles, and speech generating device manufacturer websites), conference workshops/trainings, guidance from mentor speech-language pathologists, support from peer speech-language pathologists (including via message boards, listservs, and AAC interest groups on social media), and on-the-job experience working with PALS and AAC technologies and techniques.

Discussion of Case Study

Finding optimal access technologies is a challenge that constantly changes within the decision-making framework. The overarching question that remains for Marty and his clinicians is, “How will Marty access technology today?” This question is daunting in the face of a progressive disease, and as Marty is functionally progressing into a locked-in state. Every AAC clinician who works with individuals with changing health conditions, either progressive diseases such as ALS or improving conditions such as traumatic brain injury has been faced with this question many times. The authors of this chapter recommend using the patient’s aspirations and goals in an attempt to improve quality of life as they define it. Access requirements of current assistive technology options are often difficult, if not impossible, for patients to meet. All providers should: (1) strive to try to do their best with available information and technology, (2) promote the use of new options, and (3) center care around patient needs and priorities. Even with optimal effort, answers can be elusive. Sometimes there are only more questions. Other times our best option is to consult the literature or other providers who may know the answer. Sometimes, questions simply remain unanswered.

Be creative. Try your best. Strive to give your patient every option. Try to answer questions that you and they have. Do not be afraid to feel like a novice clinician. Contribute to research efforts and research teams to help develop new options. Advocate with assistive technology companies for the needs of your patients. Together, providers can promote the health and well-being of people with late stage ALS.

Conclusions

People with late stage ALS often have difficulty using access technologies that reliably and efficiently meet their complex communication and computer access needs. There are many contributing factors to this difficulty, including disease factors (e.g., symptoms, presentation, progression, trajectory), environmental factors (e.g., positioning limitations, interference of other equipment), equipment factors (e.g., limited technological advancement to deal with unreliable user input), and treating clinician skills and competences. Given the complex interaction of contributing variables to AAC access challenges in people with late stage ALS, it can often be challenging for clinicians working with this population to identify possible solutions.

Although there is no clear answer to access challenges for this population, clinicians may address this difficulty in several ways. They can stay informed on disease-specific contributions to late stage ALS and the current state of access technologies that are available. Second, clinicians can increase their comfort and competence in operating access technologies through training and consultation with related AT professionals (e.g., ALS associations, local AT resources/groups, online AT communities). Finally, they can increase their understanding of health care systems in order to provide appropriate equipment and intervention.

The ultimate hope for the future of access technologies as it applies to this clinical population is to make technology more adaptable, accurate, and efficient for many people with late stage ALS through additional research and development. Future technological advancements will hopefully improve not only the user's experience with the technology, but also the clinician's ability to find the most optimal solution when faced with complex access challenges. These necessary advancements will, no doubt, improve the overall ability for the user to access essential communication tools, thereby improving quality of life in late stages of disease.

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