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ALS Best Practice and Documentation Examples

Introduction

- ALS, or amyotrophic lateral sclerosis, is a progressive neurodegenerative disease that affects nerve cells in the brain and spinal cord
- From the ALS Association (n.d.):
  - For about 90% of all cases, there's no known family history of the disease or presence of a genetic mutation linked to ALS.
  - For 5-10% of all cases, there's a known family history of the disease. This is often called familial ALS.
- Types of ALS
  - Spinal onset ALS
    - Approximately 70% of individuals present with spinal onset ALS (Wijesekera & Leigh, 2009)
    - These individuals present with muscle weakness and atrophy in
    - Nearly 85% of individuals with spinal-onset ALS exhibit bulbar changes as the disease progresses
  - Bulbar onset ALS
    - Approximately 30% of individuals present with bulbar onset ALS (Wijesekera & Leigh, 2009)
    - These individuals present with bulbar changes which affects speech (dysarthria) and swallowing (dysphagia)
- ALS patients typically present with a mixed dysarthria (spastic-flaccid) characterized by (Duffy, 2019):
  - Impaired articulation
  - Slow laborious speech
  - Imprecise consonant production
  - Marked hypernasality
  - Harshness
  - Strained/strangled voice
  - Disrupted prosody
  - Monopitch, monoloudness

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Three-Phase Intervention Model (Ball, Beukelman, & Bardach, 2007)

- Through the course of the disease, people with ALS (pALS) may progress to the point where they have no functional speech and require AAC as an alternative form of communication (Ball et al., 2004)
  - Approximately 80% of pALS use some form of AAC across their lifespan (Ball et al., 2004; Brownlee & Palovcak, 2007)
  - Continued technological advances provide pALS access to eye-tracking technology for communicative independence (Ball et al., 2010; Beukelman et al., 2011)

- When providing treatment for patients with ALS, it is recommended SLPs use the three-phase intervention model by Ball, Beukelman, and Bardach (2007)
  - Early Phase: Monitor, Prepare, Support
    - Occurs during the period from the patient's initial diagnosis of ALS through referral for an AAC assessment
    - Clinicians at this time:
      - Monitor for speech changes
      - Screen for cognitive decline
      - Voice banking for future AAC use
  - Middle Phase: Assess, Recommend, Implement
    - Occurs from time of referral for AAC assessment until AAC supports are selected, purchased, and initial training is completed
    - The pALS participates in an AAC evaluation assessing for both current AND future AAC needs that will meet their motor and cognitive abilities as the disease progresses
    - When to refer for an AAC evaluation?
      - Speaking rate reduction precedes reduction in speech intelligibility and is the standard for determining when to refer for an AAC assessment (Ball et al., 2002)
  - Late Phase: Adapt and Accommodate
    - Occurs during the time after the initial AAC intervention until the individual's death
    - During this phase, clinicians provide AAC modifications as needed as the disease progresses
    - Typically, pALS require the use of eye gaze at this stage

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Documentation Examples for Medical Necessity

- Case History Example
  - Patient was diagnosed with amyotrophic lateral sclerosis (ALS), a rapidly progressive neurological disease that rarely impacts cognitive function. Due to severe speech impairments impacting effective communication, the patient was referred for a comprehensive augmentative communication evaluation by their treating neurologist.
  - Patient presents with severe dysarthria making verbal communication ineffective. Low-tech strategies and devices will not be effective devices due to impaired upper extremity mobility and strength and the wide variety of messages that the patient needs to convey including communicating medical needs to medical personnel and caregivers.
  - ALS is a life-long disease and speech will continue to deteriorate, therefore prognosis for improved verbal communication is poor. It is difficult to meet patient's daily communication needs using natural modes of communication and it is necessary to evaluate and utilize a speech generating device (SGD) as means for the patient to communicate medical needs, request assistance in emergency situations, express personal wants and needs, and to socialize and interact with family.

- Motor/Access Examples
  - Example 1:
    - Access to the SGD requires use of the eye gaze accessory. Given the patient's upper extremity weakness, the eye gaze accessory is a more efficient and productive form of access for her communication needs. She demonstrates adequate oculomotor and head control to utilize the eye gaze accessory as an access method. Of note, the patient is not able to consistently and reliably use her upper extremities, lower extremities, head or neck to successfully and consistently activate SGD and the required accessories to communicate. This is secondary to profound weakness in both her arms and hands. Furthermore, the repetitive and consistent movements necessary to appropriately access speech via SGD with switch access, head pointer, and/or direct selection leads to excessive muscle fatigue given the ALS diagnosis. Over time, patient's disease will continue to progress. The patient fatigues in minutes.

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- Example 2:
    - Patient is currently unable to sustain use of hands/arms for activities of daily living throughout the course of the day secondary to increased weakness reported by patient and caregiver as well as observation during session. Head mouse is not a viable option as the repetitive movements required result in over exertion of the muscles and lead to increased muscle fatigue/weakness. A head mouse was further ruled out due to the patient's inability to perform the constant and repetitive up-down, left-right head movements required to effectively use this as an access method. In addition, switch access was also considered but ultimately ruled out as a viable option secondary to patient's inability to consistently utilize upper and lower extremities for repetitive movements. For this reason, medical necessity indicated that eye gaze access was the only access method that could be considered without causing further deterioration of motor skills. The patient possesses the physical abilities to effectively use an SGD and required accessories to communicate.

• Functional Communication Example:

- Currently, the patient's speech is not effective in meeting their communication needs across different contacts or with novel listeners. Low-tech and no tech communicative strategies and techniques are not effective or functional in meeting their communication needs, as explained below. Due to upper extremity weakness due to ALS, writing and sign language are not viable options. These communication methods rely on adequate upper extremity strength and range of motion, do not provide adequate means of communication for emergency situations, and do not permit communication via telephone. The patient previously utilized applications to support effective communication including text to speech (TTS) applications and a boogie board; however, the disease continued to progress making the use of TTS applications impossible. The voice amplifier is no longer effective due to severe dysarthria, dyspnea with verbal communication, and need for noninvasive volume ventilation support. Further, the low-tech communication aids such as communication symbols and communication boards are not adequate to fulfill daily communication needs, as they are limited by the symbols provided in the book or

board. Low-tech options cannot be used for communication over the telephone as they are non-verbal and cannot be used to summon help in an emergency situation. The patient needs to communicate a variety of messages with several communication partners in a number of different settings, and this system is not sufficient for this type of communication. Therefore, low-tech and no-tech options are not suitable to meet current communication needs and were eliminated from consideration. The patient demonstrates the need to communicate all messages including letters, single words, phrases, and sentences using the SGD. This improves efficiency and reduces excessive fatigue in communication of novel sentences and complex topics. The patient requires an SGD in order to be the most natural communicator possible. Given that the SGD has the ability to store a large number of pre-programmed messages, the patient is able to communicate efficiently and quickly without having to spell out phrases. Rate enhancement techniques such as word prediction also allow him to communicate with improved speed.

Conclusion

- Technology continues to advance providing pALS the ability to communicate independently about simple wants and needs as well as socialize with family and friends. Additionally, it allows pALS to communicate about end-of-life decisions allowing them to maintain autonomy.
- Due to the variability of progression of the disease it is incredibly important that SLPs consider AAC recommendations for the user that meets their current needs as well as their future needs.
- An AAC system that can accommodate the progression of an individual's fine/gross motor impairment is best practice!

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