

Managing Neck Weakness in Clients With Neuromuscular Disease

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Occupational therapy practitioners face a unique challenge when treating persons with a neuromuscular disease such as amyotrophic lateral sclerosis (ALS), particularly when these individuals lose head control. Invariably fatal, ALS is a progressive disorder of the nervous system that results in weakness of the skeletal muscles, including those responsible for speech, swallowing, and respiration.

Neck supports that immobilize weakened neck muscles are usually rigid and uncomfortable and inhibit speech and swallowing. We decided to investigate a variety of prefabricated and custom-made devices used to manage neck weakness in clients with ALS that might also prove helpful to persons with other neuromuscular diseases. Many of our clients exhibit a loss of head control even though they may continue to ambulate independently. These individuals present the greatest challenge for the occupational therapy practitioner because they must rely on external supports besides seating systems or positioning techniques.

Persons who have to live with their chin resting on their chest and their eyes fixed on the floor have a poor quality of life and limited social contacts. This flexed neck posture may interfere with their breathing by compromising their airway. Weakness results in an unstable upper body, which in turn causes fatigue, discomfort, and pain. Neck collars and braces are designed to immobilize the cervical spine, but most persons with ALS and other neuromuscular diseases require only support of their weakened neck muscles.

Neck supports often fail to position clients' heads that are not upright due to postural problems, such as rounded shoulders, forward head, and spinal malalignment. These deviations, along with inappropriate seating systems, contribute to neck discomfort and shoulder contractures.

CASE STUDY

One example of a person with complex and multiple problems in the dependent stage of ALS is Mrs. S., whom we worked with at our Muscular Dystrophy Association-ALS Clinic. She exhibited severe dysarthria, quadriplegia, and diffuse muscle atrophy. A pressure ulcer defaced her right ear due to an ill-fitting soft collar that she wore when sitting in her low-back wheelchair. Despite the collar, her neck remained fixed in 55° of forward flexion, 20° of right lateral flexion, and 28° of right rotation as measured by the passive cervical range of motion (CROM) instrument. She had no muscle strength in her cervical spine, including the upper quarter. Her shoulders were rounded forward approximately 2 inches. Her low, sling-back wheelchair contributed to her neck and shoulder dysfunction and reinforced her spinal malalignment (see Figure 1).

Our preorthotic management consisted of attempting to correct Mrs. S.'s deformities. We instructed her husband in how to assist with CROM exercises and upper-chest stretching to increase her mobility. A vertical roll placed along her

upper spine provided passive scapular abduction, and a lumbar support improved her posture. We fitted Mrs. S. with a Newport Neck Collar, which straightened her head and provided more jaw mobility than more rigid supports would.

Upon her return visit, Mrs. S.'s husband reported that the Newport Neck Collar required frequent adjustments and "buried" Mrs. S.'s chin. Her head orientation remained in right lateral/forward flexion and rotation. To bring her neck upright, we combined a Philadelphia Collar back with the Newport Neck Collar front. We recommended home therapy, with emphasis on family and caregiver education.

Three weeks later, Mrs. S.'s husband reported that the caregiver had



Figure 1. Mrs. S.'s inadequate seating system contributed to neck and shoulder problems.

instituted a program of range of motion and positioning. Mrs. S. indicated that her neck felt better and that she was now sleeping through the night. Her CROM had increased 5° to 10° and her ear ulcer had healed. However, the Newport-Philadelphia configuration provided inadequate support. We fitted Mrs. S. with the complete Philadelphia Collar, trimming the areas around her ear and under her chin, bringing her head up to 45°.

The modified Philadelphia Collar, along with positioning, exercise, and good family support, enabled Mrs. S. to enjoy an improved quality of life. She remained on a basic maintenance program, and we followed up with her at regular intervals.

SEATING SYSTEMS

The seating system is key to achieving proper positioning for individuals with ALS. Recommending a seating system is complicated by the fact that the person's support needs change as his or her muscles continue to weaken. Therefore, the occupational therapy practitioner must

learn to anticipate the person's future needs to avoid costly seating replacements. We have found that a lightweight chair with a head support and a reclining high back is best for balancing the head in the correct position. Additional recommendations include a firm cushion, a rigid back or insert, and a lumbar support. Removable elevating leg rests, a lap tray, and padded desk arms or arm troughs may also be desirable.

NECK SUPPORTS

We have found that our clients primarily use neck supports (i.e., soft collars, Philadelphia Collars) when riding in a car, working at a computer, walking, reading, bathing, and watching television.

Otherwise, they rely on recliner or wheelchair headrests, limiting the use of external supports to 1 to 2 hours a day, if at all.

Factors that influence their choice of support include cost, their mobility, and their access to assistance. Before recommending a collar, the occupational therapy practitioner should consider the cir-

cumstances in which it will be used and the client's muscle strength, means of locomotion, and financial circumstances.

Questions to ask when selecting a collar include the following: (a) Does the collar provide adequate support without severely restricting the person's neck rotation and peripheral vision? (b) How will the person don and doff the collar if a caregiver is unavailable? (c) Is the support cosmetically acceptable? (d) Does the support cause undue perspiration or friction?

Our clients with ALS have used the following with varying degrees of success (see Table 1):

- *Headmaster Collar*: Made in Canada, this collar is lightweight and open. Although it supports under the chin, the Headmaster is less obtrusive than other collars. Of the collars on the market, this is currently our favorite.
- *Soft Collars*: These inexpensive collars are readily available and can be used when minimal support is needed for a brief time, such as while riding in a car.

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