Mobility-Assistive Technology in Progressive Neuromuscular Disease

Wendy Lin, MD[a], Aaron Pierce, MS, ATP[b],
Andrew J. Skalsky, MD[c,d,*], Craig M. McDonald, MD[e]

KEYWORDS

• Neuromuscular disease • Mobility • Environmental control • Seating • Positioning

KEY POINTS

- Children as young as 24 months can learn to safely operate a power wheelchair.
- Power-assist wheelchairs are useful for nonambulatory patients with neuromuscular disease (NMD) with mild upper limb weakness.
- The NMD multidisciplinary team needs to consider the specific neuromuscular disease and plan for appropriate progression affecting the need for modifiable mobility devices.
- Proper seating and positioning is critical to maximize function and the optimal use of mobility-assistive technologies.
- Electronic control systems are available to integrate with environmental control systems and computer access.

INTRODUCTION

Mobility-assistive technology is essential for maintaining the function of individuals with severe or progressive neuromuscular disease (NMD). The types of devices required change as the disease progresses. In the pediatric population, despite disease progression, there can still be developmental progress in a variety of cognitive and social domains. Without proper accommodations for goal-directed independent mobility whereby children can explore and affect their environment, there can be delays in cognitive and social development caused by mobility impairment.[1] These devices need to be able to accommodate and adapt to the individual's needs.

[a] Department of Pediatrics, University of California San Diego, San Diego, CA, USA; [b] Independent Consultant, 429 Durian Street, Vista, San Diego, CA 92083, USA; [c] Pediatric Rehabilitation Program, University of California San Diego School of Medicine, San Diego, CA, USA; [d] Pediatric Rehabilitation Medicine, Rady Children's Hospital, 3020 Children's Way, MC 5096, San Diego, CA 92123, USA; [e] Department of Physical Medicine and Rehabilitation, University of California Davis School of Medicine, 4860 "Y" Street, Sacramento, CA 95817, USA
* Corresponding author. Pediatric Rehabilitation Medicine, 3020 Children's Way MC 5096, San Diego, CA 92123.
E-mail address: askalsky@rchsd.org

Phys Med Rehabil Clin N Am 23 (2012) 885–894
http://dx.doi.org/10.1016/j.pmr.2012.08.007 **pmr.theclinics.com**
1047-9651/12/$ – see front matter

As more specialized functions are required, the financial cost also increases. It is important for the clinician, therapist, family, and patient to be aware of what durable medical equipment coverage is available as well as the qualification for such equipment based on medical necessity and documentation requirements imposed by their specific funding source. Coverage, qualification based on medical necessity, and documentation requirements vary by payer source and region. It is also important to consider charitable resources that can provide assistance when needed.

Required considerations for the proper evaluation of mobility-assistive technology include a physical, visual, and cognitive assessment. The evaluation frequency needs to be completed based on the functional status of the patient and the rate and degree of disease progression. Physical, occupational, and speech therapists can be involved to provide insights regarding the interface between the patient and the device. These insights may influence the decision to choose a particular product rather than another. Overall, there needs to be a patient-centered team approach when addressing the needs of this patient population to determine what type of device is most appropriate.

MOBILITY-ASSISTIVE TECHNOLOGY IN THE PEDIATRIC NMD POPULATION

Mobility-assistive technology has multiple roles. Besides providing home and community accessibility, it plays a role in the child's overall development. These devices facilitate learning and social development. These devices also need to be tailored to the child's developmental needs. Failure to provide access to the appropriate devices limits children's ability to achieve their full potential.[1]

Butler[2] published a case series of 8 children mostly with spinal muscular atrophy (SMA) type II. The youngest child who was able to achieve safe and independent power mobility was 24 months old. One child began learning the task of operating a powered mobility device at 20 months of age. These children were all of normal intelligence.[1] Common diagnoses considered for early power mobility include SMA type II, congenital muscular dystrophies, congenital myopathies, congenital myasthenic syndromes, and Charcot-Marie-Tooth (CMT) type 3 or Dejerine-Sottas disease. Butler and colleagues[2] subsequently studied the effects of powered mobility on the development of young children with locomotor disability. They showed that learning powered mobility at 24 months had benefits on the frequency of self-initiated interaction with objects, spatial exploration, and communication with caregivers.

MANUAL WHEELCHAIRS

Patients with milder or more slowly progressive NMD can often use a manual wheelchair as their primary mode of mobility. Decreased exercise tolerance either caused by primary muscle or cardiopulmonary effects of the disease needs to be taken into consideration in determining what type of manual wheelchair would be most appropriate.

Manual wheelchairs can be divided into independent versus dependent devices. Independent manual wheelchairs (eg, lightweight or ultralightweight manual wheelchairs) can be used early in the disease to provide some level of energy conservation for community mobility for patients who may be able to ambulate only household distances.[3] Dependent manual wheelchairs (eg, tilt-in-space manual wheelchairs) may become necessary as the disease progresses and patients are no longer able to perform any type of independent mobility or cognitive dysfunction limits their ability to use forms of independent mobility. Dependent manual wheelchairs are pushed by the caregiver. Dependent-type wheelchairs, although heavier, can better accommodate additional devices such as adaptive communication devices, ventilators, and