Swallowing and Secretion Management in Neuromuscular Disease

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KEYWORDS

• Neuromuscular disease (NMD) • Dysphagia • Sialorrhea • Amyotrophic lateral sclerosis • Muscular dystrophy • Myasthenia gravis

KEY POINTS

- Dysphagia associated with neuromuscular diseases reflects underlying disease patterns, such as severity, natural course, progression, and intervention options.
- The clinical swallowing examination provides information regarding underlying neuromuscular function leading to dysphagia symptoms, potential benefit from and timing for further instrumental studies, prognosis for improvements in swallowing function and potential to benefit from intervention.
- Supplemental instrumental assessment is needed when the clinical examination is inadequate to determine aspects of swallowing that will impact outcome.
- Dysphagia intervention aims to reduce risk for medical complications, including pneumonia and respiratory failure, and improve participation with life activities involving intake of food and quality of life.

INTRODUCTION

Neuromuscular disease (NMD) frequently leads to difficulty swallowing (dysphagia) and managing secretions (sialorrhea and/or excessive thick mucus). The act of swallowing is complex and involves at least 3 phases (oral, pharyngeal, and esophageal). Swallowing requires adequate strength and coordination of the bulbar musculature and production of saliva, as well as intact sensory, esophageal, respiratory, and cortical functions. Swallowing function is also important for saliva management, because humans typically generate more than one-half liter of saliva per day[1] and unconsciously swallow approximately every 1 to 3 minutes.[2,3] Bulbar and respiratory muscle weakness, often associated with NMDs, disrupt the ability to swallow safely and efficiently. Dysphagia may lead to deleterious medical complications, such as malnutrition,[4] dehydration,[5] aspiration pneumonia, and other pulmonary sequelae,[6] such as pulmonary

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fibrosis, localized lung inflammation, or abscess. In many NMDs, the ultimate cause of death is respiratory failure, which is often preceded by aspiration pneumonia.[7–9] Not surprisingly, dysphagia in individuals with NMD is also associated with increased health care costs, and mortality.[10] For many, dysphagia may also interfere with participation in communicative events involving food, such as conversation around the dinner table with family and other social events, and lead to reduced enjoyment of eating and quality of life.[11] This, in turn, may lead to depression and/or social isolation.[12] The identification and management of dysphagia can help to mitigate these complications and consequences. This review provides an overview of dysphagia associated with NMD in adults, along with a concise review of swallowing assessment and intervention options.

DYSPHAGIA ASSOCIATED WITH NEUROMUSCULAR DISEASE: AN OVERVIEW

Specific patterns of dysphagia associated with NMD reflect the underlying disease. Therefore, these patterns vary depending on several factors, including the characteristics of the disorder; the severity, natural course, or stage of progression; and the intervention options. For instance, dysphagia in some NMDs, such as Duchenne muscular dystrophy, progresses relatively slowly, whereas others, such as amyotrophic lateral sclerosis (ALS), often progress rapidly. Furthermore, dysphagia in myasthenic patients tends to fluctuate and is typically reversible with adequate treatment of the disease. In addition, the pattern of swallowing-related impairments reflects the pattern of neuromuscular weakness. For instance, reduced speed,[13] aspiration (including silent aspiration),[14] coughing and choking,[15] upper esophageal sphincter (UES) hypertonicity[16] and laryngospasm[17] in individuals with ALS may be related to the combination of spastic and flaccid weakness associated with ALS. However, in Duchenne muscular dystrophy, a pattern of difficulty with chewing and oropharyngeal transport of solid foods, as well as pharyngeal residue without aspiration is more common,[18] and is likely due to flaccid weakness of jaw, tongue,[19] and pharyngeal constrictor musculature. Many NMDs occur in individuals of advancing age. For instance, peak age of onset is typically between 55 and 75 years for ALS,[20] and 70 years for inclusion body myositis.[21] In these cases, the impact of the NMD is overlaid upon age-related changes to swallowing, such as those caused by the loss of dentition, presbylarynx or presbyesophagus.[22] For these reasons, the overall prevalence and incidence of dysphagia in individuals with NMD is challenging to determine. **Table 1** outlines the literature describing the prevalence of dysphagia in selected NMDs.

Respiratory muscle weakness is associated with NMDs, leading to restrictive lung disease and impairment of the ability to cough, which contributes to the risk for dysphagia, aspiration pneumonia, and other pulmonary complications. Discoordination of breathing and swallowing may also occur.[30] When respiratory impairments are combined with bulbar impairments, the impact on swallowing and pulmonary defenses may be synergic, even lethal.[31] For instance, because the ability to cough requires the active coordination of respiratory musculature and intrinsic laryngeal muscles, the combination of bulbar and respiratory muscle impairment may render the ability to cough or use compensatory strategies for cough completely ineffective at a time when the protective defense of cough is needed to guard the lungs from aspiration.

Excessive saliva or drooling frequently co-occurs with dysphagia and is a manifestation of swallowing difficulty and clearance of normal secretions, rather than an increase in salivary flow.[32] Indeed, increasing salivary flow is a treatment challenge—even pharmacologically—whereas a decreased frequency of automatic saliva swallowing has been observed in individuals with dysphagia.[3] In addition, individuals with dysphagia owing to

Table 1
Prevalence of dysphagia in selected neuromuscular diseases

Examples	Prevalence
Inclusion body myositis	65%[23] to ≤80%[24]
Duchenne muscular dystrophy (muscle level)	Unknown; ≤30% report dysphagia[25] >95% based on MBSS findings[26]—greater with advancing age
Myasthenia gravis (neuromuscular junction disease)	30% as an early symptom[27] Up to two-thirds of individuals with myasthenia gravis[28]
Amyotrophic lateral sclerosis (motor neuron disease)	95%–98% bulbar onset[29] 35%–73% spinal onset[29]

Abbreviations: CNS, central nervous system; MBSS, modified barium swallow study.
Data from Refs.[23,26–29]