Neurological and Neuromuscular Disorders
Dental Hygiene Care for Special Patients

Introduction
Physiologically, the sympathetic and parasympathetic nervous systems, and their links with the human musculature, are intricate and complicated. Neurological and neuromuscular conditions and disorders, such as Multiple Sclerosis and Muscular Dystrophy, pose oral care challenges for both clients and for the dental hygienists who provide comprehensive care for this population. Due to the complex nature of these conditions, and in order to provide optimal levels of dental hygiene care, it is important for dental hygienists to understand the various signs and symptoms of neurological and neuromuscular conditions as well as the associated limitations these disorders place on the affected clients. An increased understanding of neurological and neuromuscular disorders can lead to a collaborative approach between the dental hygienist and the client, thus optimizing their oral health outcomes.

LEARNING OBJECTIVES
Upon completion of this course, the dental professional will be able to:
1. Discuss the general pathophysiology related to neuromuscular disorders.
2. Recognize the signs and symptoms of potential neuromuscular disorders.
3. Make ongoing evaluations of a client’s physical and mental complications for individualized care planning.
4. Discuss knowledgeably the various aspects of the specific neurological and/or neuromuscular disorder with the client.

NEUROLOGICAL AND NEUROMUSCULAR DISORDERS
The neuromuscular system is a fine balance of electrical impulses from the brain, moving down the spinal cord, and the action of chemical transmitters to the muscle fibers working cohesively to produce movement. Disorders affecting these systems fall into two descriptive categories: neurological and neuromuscular. A plethora of conditions affecting neuromuscular function exists; however, for the purpose of this review, the following four conditions will be discussed: Multiple Sclerosis (MS), Amyotrophic Lateral Sclerosis (ALS), Myasthenia Gravis (MG), and Myotonic Muscular Dystrophy (DM). Knowledge and understanding of these disorders is critical to the dental hygiene professional in order to optimize dental hygiene assessment, treatment planning and the delivery of care.

Neurological Disorders
Neurological disorders stem from pathology of the brain, spinal cord, and nerves. As a result of neurological dysfunction, the signaling between the nervous system and muscles is disrupted. This leads to malfunctions affecting voluntary and involuntary movement, including walking, speaking, swallowing, breathing, and cognitive functions such as learning and memory. Some examples of neurological disorders are: Alzheimer’s, Parkinson’s, migraine headaches, and Multiple Sclerosis.

Multiple Sclerosis
Multiple Sclerosis (MS) is a neurological condition of unknown etiology affecting sensory and motor nerve transmission. It affects more females than males at a ratio of 3:2, with an average age of onset between 20 and 45 years. The disease causes damage to the myelin sheaths around the axons within the brain and spinal cord. Due to the loss of myelin sheathing and exposure of axon fibers, inflammation results, leading to interference with nerve impulse transmission. The range of symptoms for MS is varied but includes: muscle weakness, dizziness, balance problems, bladder and bowel dysfunction, changes in personality, depression, and cognitive impairment. Visual disturbances, facial pain, and trigeminal neuralgia are common early symptoms, as well as numbness or paresthesia of the extremities. In addition, people affected by MS complain of muscle spasticity, tremors, spasms, and fatigue, which may limit daily activities. Dysphagia (difficulty with swallowing), and dysarthria (difficulty with speech) can also be complications of MS, particularly in the advanced stages. Trigeminal neuralgia, a painful condition caused by inflammation of the trigeminal nerve (fifth cranial nerve), is present in up to 32% of those individuals afflicted with MS. It is important for dental hygienists to be cognizant of the pain experienced with trigeminal neuralgia, which can be triggered by even the lightest, most gentle touch. In addition, some individuals may find tooth brushing to be very painful due to their trigeminal neuralgia, resulting in poor oral hygiene. Bilateral facial palsy may occur in up to 25% of MS cases, but should not be confused with Bell’s palsy, a temporary form of facial paralysis of the sixth cranial nerve affecting only one side of the face. Between 20 and 50% of individuals living with MS report experiencing chronic facial pain, presenting challenges for clinicians in interpreting and diagnosing their pain. The clinician must try to determine whether the pain is due to a dental infection or disease, or due to MS-related symptoms. Chronic pain from MS may present as paraesthesia, burning, throbbing.
or shooting pain, or may be revealed as a painful reaction to a typically non-painful touch.

Mobility challenges, including problems with gait, affect the activities of daily living for the client with MS. Depending on the severity of the condition, people living with MS may require a cane or other mobility aids such as walkers and wheelchairs. Dental hygienists must consider mobility as a potentially limiting factor for access to professional oral health care. As the severity of the disease increases, so do the physical barriers regarding access to care. When mobility is severely restricted and the individual loses the ability to bear weight, additional challenges may arise due to the client’s inability to transfer from the wheelchair to the treatment chair without mechanical assistance. In such cases, individuals must be treated in the wheelchair, or treated in special practice settings equipped with mechanical lifting devices. Some wheelchairs have a reclining feature, allowing the clinician easier access to the oral cavity while the client remains in his/her wheelchair. Even with a reclining feature, access and visibility may be limited for the dental hygienist in addition to the obvious ergonomic challenges.

Manual dexterity is another challenge for individuals living with MS, making tooth brushing and flossing difficult. Of the people living with MS, approximately 25% are unable to independently execute personal oral self-care, and almost one third of individuals need to adapt to using their non-dominant hand in order to perform daily oral self-care tasks. With advanced stages of disease, oral health care providers must be aware that the client’s ability to verbalize his/her needs may be impaired due to dysarthria. In addition, if dysphagia becomes significant, choking and aspiration risks are increased, thus contraindicating the use of ultrasonic debridement equipment. Individuals may require a feeding tube for nutrition when dysphagia is substantial and may have increased levels of dental calculus due to the lack of mastication, therefore creating an even greater need for professional dental hygiene care.

Neuromuscular Disorders

Neuromuscular disorders, as compared and contrasted to neurological disorders, affect nerves controlling voluntary muscles such as the muscles of the arms and legs, while neurological disorders can affect both voluntary muscles and involuntary muscles, as well as brain neurons. The numerous neuromuscular disorders can be subcategorized by pathophysiology and for the purpose of this review, examples of neuromuscular conditions categorized as motor neuron, peripheral neuropathy, and myopathy will be highlighted.

Motor Neuron Diseases

Motor neuron diseases affect essential muscle activity; including walking, breathing, and swallowing. Messages sent from the brain are disrupted due to nerve damage, with gradual muscle weakening and wasting, and muscle twitching resulting from these signal disruptions. Motor neuron diseases are more common in males than females, and typically do not affect individuals until after 40 years of age. Sporadic, or non-inherited motor neuron diseases, have unknown causes but may have environmental or viral factors associated with them. Amyotrophic lateral sclerosis is one of the most common motor neuron diseases and will be discussed in the following section.

Amyotrophic Lateral Sclerosis

Amyotrophic Lateral Sclerosis (ALS), also referred to as Lou Gehrig’s disease, is a neurodegenerative disorder of unknown etiology; however some genetic factors have been identified with this disease. While there have been no consistent associations between environmental factors and ALS, tobacco use has been associated with its development in some cases. Progressive muscular paralysis due to degeneration of motor neurons in the primary motor cortex, brain stem, and spinal cord are all characteristics of ALS. The distribution of ALS diagnoses is 5-10% familial, exhibiting a Mendelian genetic inheritance pattern, while in the vast majority of cases, 90%, it is a sporadic disease without a familial link. The age of onset for sporadic ALS is between 55 and 65 years, accounting for about one third of all ALS cases, with a male to female ratio of approximately 1.5 to 1. Approximately two thirds of all ALS cases are of the classical or spinal form, affecting the upper and lower motor neurons.

ALS can have a ‘limb’ onset (i.e., affecting the arms and legs) or a ‘bulbar’ onset (i.e., affecting swallowing ability). With limb onset, the main symptoms are related to muscle weakness and wasting, with foci being muscles of the shoulders, forearms and hands, as well as the proximal thigh and distal foot muscles. Upper limb weakness presents difficulties for the individual facing tasks associated with both gross motor function such as hair washing, and fine motor skills such as holding and manipulating a pen. Abnormal muscle tone in limb onset ALS may present as fasciculations (involuntary muscle twitching), and spasticity (increased stiffness and uncontrolled or sudden jerking movements).

With bulbar onset, ALS presents with dysarthria and dysphagia. Dysphagia leads to increased risk of aspiration, dehydration and malnutrition. The soft palate may be weakened, contributing to dysarthria and causing a nasal quality of the voice, along with a weakened tongue causing slow, slurred speech. Limb symptoms may also present concurrently with bulbar symptoms. Sialorrhea (excessive drooling) is present in most ALS cases due to swallowing...
difficulties in combination with mild bilateral facial weakness affecting the lower part of the face. In addition to the above ALS symptoms, there are ‘pseudobulbar’ symptoms of lability (spontaneous emotional changes) and excessive yawning. Emotional lability consisting of pathological laughing or crying is seen in approximately 50% of all cases. It is important for dental hygienists to understand that emotional lability is not a mood disorder, but rather an effect of the underlying ALS pathology. Other signs and symptoms of ALS that may impact dental hygiene care are the brisk jaw jerks associated with spasticity. Furthermore, the gag reflex is preserved or heightened throughout the disease’s progression. Fasciculations, or wasting of the tongue, as well as slowed tongue movements due to spasticity, are other features of ALS that the oral health care provider should be aware of as they create clinical management challenges affecting intraoral access can increase the risk of inadvertent aspiration.

Other common symptoms of ALS include fatigue and reduced capacity for exercise, eventually leading to the need for assistance with activities of daily living. Individuals with ALS experience chronic pain in up to 73% of cases and it is not uncommon for dementia to be present as well. Approximately half of individuals diagnosed with ALS will die within three years of the disease onset, with respiratory failure and pulmonary complications being the typical causes of death.

Peripheral Neuropathy

Over one hundred types of peripheral neuropathy have been identified, each with their own set of symptoms. All peripheral neuropathies consist of damage to the peripheral nervous system. The peripheral nervous system is responsible for transmitting information from the brain and spinal cord to the rest of the body. The types of symptoms experienced are dependent upon the diagnosis, and vary from temporary numbness to muscle weakness, burning pain to organ dysfunction. For the purpose of this review, Myasthenia Gravis has been highlighted as one of the peripheral neuropathies.

Myasthenia Gravis

Myasthenia Gravis (MG) is an uncommon acquired autoimmune disorder that can occur at any age, but has a peak age of onset for females between the teen years and the 30s, and between the age of 50 and 70 for males. More females are diagnosed with MG than males, and the disorder is characterized by weakness of the skeletal muscles and fatigue with exertion. MG is also associated with other autoimmune conditions such as Grave’s disease (a thyroid condition), diabetes, rheumatoid arthritis, and lupus. A distinct characteristic of MG is a fluctuating skeletal muscle weakness that can vary from day-to-day, hour-to-hour, and muscle-to-muscle. MG only affects voluntary muscles; while other involuntary muscles, such as the heart, remain unaffected.

With the number of different muscles that can be affected, there is a wide variation in clinical presentation of MG. Ocular symptoms are common at onset of MG. Individuals may experience ptosis (eyelid drooping) due to weakness of the levator palpebrae muscle, and diplopia (blurred vision) due to weakness of the extraocular muscles. A characteristic of MG is Cogan’s lid twitch, an upper eyelid twitch that occurs after looking down for a few seconds and then looking straight again. Moreover, many persons with MG report that bright lights are bothersome, making the eye muscles weak. From a dental hygiene perspective, problems tolerating bright lights are important to remember due to the bright overhead treatment lights or light sources attached to loupes. Taking care to keep the operatory light directed away from the client’s eyes as much as possible, and providing dark protective eyewear can help to minimize the negative effects of the bright sources of light required for the provision of dental hygiene care.

With MG disease progression, facial, including the orbicularis oris, and masticatory muscle weakness will become more apparent leading to dysphagia, dysarthria, and eventually to the appearance of an expressionless face. When an affected individual laughs, the appearance of a ‘myasthenic sneer’ is created. The sneering appearance is due to the activation of the levator muscles that raise the upper lip exposing the canines without the corners of the mouth being drawn up and out. Individuals experiencing these effects are unable to whistle, effectively use a straw, blow up a balloon, or expectorate. Tongue weakness is another symptom of MG. The classic ‘myasthenic’ tongue has a distinctive triple longitudinal furrow and is flaccid and atrophied, creating ongoing choking risks as the tongue is unable to move food effectively. Like ALS, the soft palate muscles may be weakened producing a nasal quality to the voice, and nasal regurgitation. Individuals affected with MG may have difficulty with chewing due to masseter muscle weakness, and in severe cases the jaw may need to be manipulated by hand to facilitate chewing as well as finger support to stay closed.

Dysphagia, resulting from weakness of the tongue and posterior pharyngeal muscles, and muscle fatigue associated with chewing and swallowing is common and one of the most serious symptoms of MG. Another serious symptom of MG is difficulty breathing due to the airway becoming obstructed by secretions that the individual cannot clear away because the muscles that produce a cough are too weak. Because of facial muscle weakness and the resulting lack of facial expression, the affected person may be in crisis, but not appear to be distressed. Signs and symptoms of airway distress include restlessness and rapid shallow breathing. For clinicians, an awareness of such
signs of distress are key points to remember when treating a client with MG in order to maintain client safety. In addition, MG muscle weakness causes difficulties for self-care (such as hair washing and shaving) and oral self-care (such as tooth brushing). Furthermore, difficulties walking long distances, walking up stairs, and the use of a wheelchair, may pose obstacles in accessing care for oral hygiene services. Certain factors have been identified in increasing muscle weakness for individuals with MG including: exertion, hot temperatures, infections, stress, and certain drugs including ester-type local anesthetics. When administering a local anesthetic, it is important that the client remain in the dental chair until they are able to swallow without difficulty and they no longer need suction assistance to clear secretions. For stress or anxiety reduction, use of nitrous oxide sedation is an acceptable option for these clients. In terms of oral self-care, the use of a powered toothbrush is helpful. Regardless of the type of toothbrush used, the handle may require a modification in order for the individual to be able to manage the device effectively while reducing muscle fatigue.

Myopathy

Myopathies are primary, inherited neuromuscular disorders characterized by muscle necrosis and progressive degenerative skeletal muscle. Myopathies fall into two broad categories: acquired, as with muscle cramps, and inherited, as with muscular dystrophy.

Myotonic Muscular Dystrophy

Myotonic muscular dystrophy (DM) is an autosomal dominant, multisystemic genetic disease that encompasses a variety of impairments including the muscular, ocular, respiratory, cardiac, endocrine, and central nervous systems. Myotonic dystrophy type 1 (DM1) is the most common type affecting adults, with a prevalence rate of 2.1 to 14.3 per 100,000 people. The age of onset varies greatly, from birth to 60 years of age and beyond. The clinical manifestations of DM1 include ptosis (drooping eyelids), atrophy of the small muscles of the hand and forearm extensor muscles, and weakness of facial muscles. Common signs and symptoms include: myotonia (muscle rigidity), cardiomyopathy (abnormal functioning of the cardiac muscle), cataracts, and endocrinopathy. Cataracts are the most common finding in those individuals with asymptomatic or late onset DM1, and in such late onset cases, myotonia and muscle weakness are rarely present. However, for most individuals with DM1, muscle atrophy affecting curvature of the neck, coined ‘swan neck.’ DM does not always demonstrate myotonia, or limb or facial muscle weakness. Typical symptoms may include diabetes, cognitive deficits, excessive daytime sleepiness, irritable bowel syndrome, respiratory failure, and cardiac conduction (electrical impulse) abnormalities.

With DM, there is a progressive loss of muscle strength, and weakness of facial and anterior neck muscles. As deterioration progresses, individuals typically rely on wheelchairs for mobility and often have difficulty carrying out daily activities. Excessive daytime sleepiness is common and can be debilitating or disabling for DM1. Myotonias are common and can affect grip and other muscles such as the tongue and facial muscles, causing challenges with speaking, chewing and swallowing. Along with myotonias, it should also be noted that muscle pain is a common symptom of DM. Sudden cardiac death, in all age groups, may occur due to affected cardiac muscles. Furthermore, involvement of the central nervous system may occur including cognitive impairment, dementia, and changes in personality. Chest infections and diaphragm weakness contribute greatly to respiratory failure, with many individuals become severely disabled by the fifth or sixth decade of life.

For clients with DM, dental hygienists should be aware that there is a high prevalence of malocclusion, including anterior open bite, lateral cross bite, and excessive inter-occlusal distance, possibly caused by reduced muscle function. Due to their inability to maintain adequate personal oral hygiene, individuals with DM tend to have fewer teeth, higher plaque scores and caries experience, and greater periodontal pocket depths compared to healthy cohorts. Further considerations related to weakness of the muscles of mastication and facial expression with DM include potential difficulties with mastication, swallowing, respiration, and expectoration.
limitations during self-care management, strategies for individual client care could include: modified handles on manual toothbrushes, use of powered toothbrushes, and when determined necessary, assistance with daily oral care. In addition, any modified oral care techniques that have been recommended should be periodically re-evaluated as disease progression continues. New strategies and modifications should be suggested in order to meet the evolving challenges and specific needs of the client.

Ensuring that safe professional oral care is provided requires a working knowledge of neurological and neuromuscular conditions. Health and safety issues include risk of falls due to compromised ability to walk, risk of aspiration, and the need for appropriate use of local anesthetics. Potential side effects from local anesthesia include increased impairment of the swallowing reflex with mandibular nerve block injections.18 In order to facilitate a safe dental hygiene treatment experience, consultation with general medical practitioners or specialists is recommended.

Generally, for neurological and neuromuscular disorders with the common symptoms of fatigue, weakness and the potential for distress, it is helpful to establish an open and friendly rapport with the client to reduce or eliminate emotional stress, and to adequately and respectfully assess and meet their specific needs.18 Allowing the client to rest before beginning the procedure may also be beneficial, along with scheduling multiple, short early morning appointments when the client has greater muscle strength.18 Keeping the client upright or only slightly reclined during the appointment will also help to avoid closing the throat, thereby reducing fluid regurgitation.18 Use of a mouth prop may help to reduce masticatory muscle stress; however it is important to ensure that the jaw is not being overstretched.18

Removing and adapting to physical barriers, such as ensuring wheelchair accessibility, and working while the client remains seated in a wheelchair, should be considered fundamental and reflect the right of all individual’s to receive the highest standard of professional oral healthcare possible. Complex and severe cases may require special care practice settings.

While the preceding discussion reflects a mere sampling of neuromuscular and neurological conditions, dental hygienists should be aware of the general and specific health and care needs of each client. A thorough review of the client’s medical history is crucial to obtaining relevant and adequate information in order to provide the best, safest, and most empathetic oral healthcare possible. Individualized treatment adaptations and considerations can serve to reduce client anxiety, and increase confidence in the dental hygiene practitioner’s knowledge and expertise, culminating in a positive experience and improved outcomes for both the client and the dental hygienist.

References are available in the online version of this issue at www.cdha.org

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1. Neurological disorders have symptoms affecting:
   a. voluntary and involuntary movement and cognitive functioning
   b. involuntary movements
   c. voluntary movements
   d. cognitive functioning and voluntary movements

2. Which of the following statements is TRUE of Multiple Sclerosis?
   a. it has unknown etiology and affects more females than males
   b. it is caused by a virus and affects more males than females
   c. the average age of onset is birth to 16 years
   d. it causes damage to the myelin sheaths in the brain and spinal cord
   e. both a and d

3. Trigeminal neuralgia, which occurs in 32% of Multiple Sclerosis clients, can result in:
   a. pain with bright lights
   b. pain with walking
   c. pain with toothbrushing
   d. pain with shoulder and arm movements

4. Motor neuron diseases including Amyotrophic Lateral Sclerosis (ALS):
   a. affect essential muscle activity; including walking, breathing, and swallowing
   b. affect more males than females
   c. usually have an onset at 40 years of age or older
   d. all of the above

5. Most of the cases of Amyotrophic Lateral Sclerosis (ALS) have a strong familial or genetic link.
   a. True  b. False

6. Difficulty swallowing, (dysphagia) a common condition in neurological as well as neuromuscular disorders, can lead to which of the following?
   a. increased risk of aspiration
   b. cardiac arrhythmia
   c. bilateral facial muscle weakness
   d. increased pain

7. Myasthenia Gravis (MG) is an autoimmune disorder and is characterized by weakness of the skeletal muscles and fatigue with exertion. MG is also associated with other autoimmune conditions such as:
   a. Parkinson’s disease and Alzheimer’s disease
   b. Grave’s disease and diabetes
   c. Sjögren’s syndrome and Parkinson’s disease
   d. Nicotine and alcohol addiction

8. Ocular symptoms are common at the onset of which neuromuscular disorder?
   a. Multiple Sclerosis  b. Myasthenia Gravis
   c. Amyotrophic Lateral Sclerosis  d. Myotonic Muscular Dystrophy

9. Forward curvature of the neck, “swan neck”, caused by atrophy and weakness of the sternocleidomastoid muscle is a symptom found in:
   a. Multiple Sclerosis
   b. Myasthenia Gravis
   c. Amyotrophic Lateral Sclerosis
   d. Myotonic Muscular Dystrophy

10. Special dental hygiene treatment considerations for neurological and neuromuscular disorder clients include:
    a. more upright client positioning
    b. careful selection of local anesthesia types and techniques
    c. individualized modifications for personal oral hygiene strategies
    d. optimal appointment scheduling
    e. all of the above

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