Disease/Medical Condition

MUSCULAR DYSTROPHY

College of Dental Hygienists of Ontario
Protecting your health and your smile

Date of Publication: Dec. 20, 2017

(also known as "MD"; includes "Duchenne muscular dystrophy" [DMD], "Becker muscular dystrophy" [BMD], "oculopharyngeal muscular dystrophy [OPMD], "fascioscapulohumeral muscular dystrophy" [FSHMD][1], "limb-girdle muscular dystrophy" [LGMD], "myotonic dystrophy" [DM][2], "Emery-Dreifuss muscular dystrophy" [EDMD], and congenital muscular dystrophy [CMD][3])

Note: *Unless otherwise specified, this fact sheet primarily addresses the two most common types of muscular dystrophy; namely, Duchenne and Becker type.*

Is the initiation of non-invasive dental hygiene procedures* contra-indicated? No.

- **Is medical consult advised?** No, assuming patient/client is already under medical care for muscular dystrophy.

Is the initiation of invasive dental hygiene procedures contra-indicated?** Possibly, but not typically.

- **Is medical consult advised?** ..... See above.
- **Is medical clearance required?** ..... Yes,
  - if patient/client is being treated with medications associated with immunosuppression +/- increased risk of infection (i.e., corticosteroids, such as deflazacort or prednisone, which are used to preserve muscle strength in Duchenne and some other forms of muscular dystrophy), or
  - if general anesthetic agents are contemplated (due to elevated risk of adverse reactions, some of which may be life-threatening)[4], or
  - if bisphosphonate-related osteonecrosis of the jaw (BRONJ) exists or is suspected[5].
- **Is antibiotic prophylaxis required?**..... No, not typically (although extended use of corticosteroids may warrant consideration of antibiotic prophylaxis).
- **Is postponing treatment advised?** ..... Possibly, but not typically (depends on medical clearance for patient/client taking corticosteroids, as well as whether patient/client can be sufficiently cooperative for examination and treatment to take place, as well as presence/absence of oral conditions — such as tooth fractures — that may need to be addressed prior to dental hygiene treatment).

Oral management implications

- Ambulatory patients/clients with MD may be prone to falls and injuries. Therefore, appropriate precautions should be taken in the dental hygiene office.
- By 10 years of age, patients/clients with Duchenne MD are usually unable to walk and are confined to a wheelchair. Patients/clients with myotonic dystrophy are wheelchair-bound within 20 years of onset (which is usually the second to fifth decade of life). Thus, the dental hygienist should ensure familiarity with wheelchair protocols.
- A warm towel may be useful to alleviate muscle cramping or tightness while the patient/client is in the dental chair.

1 Fascioscapulohumeral muscular dystrophy is subdivided into FSHMD type 1 and FSHMD type 2, which have different genetic causes but generally similar signs/symptoms.
2 Myotonic dystrophy type 1 (DM1) is also known as Steinert's disease. Myotonic dystrophy type 2 (DM2) is also known as proximal myotonic myopathy (PROMM).
3 Congenital muscular dystrophy comprises a group of muscular dystrophies, which includes Fukuyama CMD, Walker-Warburg Syndrome, and Muscle-Eye-Brain Disease.
4 Depolarizing muscle relaxants (e.g., succinylcholine, also known as suxamethonium) are contraindicated in many persons with MD due to risk of potentially life-threatening hyperkalemia (elevated blood potassium). Furthermore, certain inhaled anaesthesic agents (e.g., desflurane, enflurane, isoflurane, haloflurane, and sevoflurane) should be avoided if possible in many patients/clients with MD due to risks including rhabdomyolysis (breakdown of skeletal muscle tissue, which may lead to kidney damage) and hyperkalemia. Some studies have found an increased risk of malignant hyperthermia in patients/clients with DMD or BMD following general anaesthesia. With close monitoring, local anaesthetic agents and nitrous oxide are safe for most patients/clients with Duchenne and other forms of muscular dystrophy.
5 To counteract osteoporosis resulting from lack of physical activity and concomitant steroid therapy for management of MD, bisphosphonate agents (e.g., aldrenonate) may be employed.

cont'd on next page...

MUSCULAR DYSTROPHY

(also known as "MD"; includes "Duchenne muscular dystrophy" [DMD], "Becker muscular dystrophy" [BMD], "oculopharyngeal muscular dystrophy" [OPMD], "fascioscapulohumeral muscular dystrophy" [FSHMD], "limb-girdle muscular dystrophy" [LGMD], "myotonic dystrophy" [DM], "Emery-Dreifuss muscular dystrophy" [EDMD], and congenital muscular dystrophy [CMD])

Oral management implications *(cont'd)*

- Adjustments to the dental chair may be required to accommodate physical deformities (e.g., scoliosis or swan neck) and/or muscle weakness. Special cushions and lumbar/cervical rolls (which the patient/client may be able to provide) can be helpful.
- For the patient/client who has difficulty keeping his/her mouth open for a long time, a rubber bite block may prove useful for dental hygiene procedures. A tongue retractor can be used to reduce unwanted tongue movement. A rubber dam will reduce inhalation of foreign substances while the patient/client is in the dental chair.
- Loss of strength in the hand and wrist muscles can impede ability to grasp and use a toothbrush and dental floss. In fascioscapulohumeral MD, muscle weakness in the shoulder area can affect the patient/client's ability to lift objects such as a toothbrush. Thus, brushing of the teeth and other oral hygiene activities may require the assistance of a caregiver. Referral to an occupational therapist may also be helpful for assistive device selection and technique modification.
- The dental hygienist should be alert for signs/symptoms of bisphosphonate-related osteonecrosis of the jaw in patients/clients receiving bisphosphonate treatment.
- Anaesthetic agents should be used cautiously, if at all, in patients/clients with neuromuscular disorders. Inhaled anaesthetics may have an adverse effect on an already compromised heart. Any anaesthetic agent that affects the muscles will also affect muscles used to breathe.
- Because many patients/clients with MD have difficulty chewing and dysphagia, referral to a registered dietitian should be considered to assess the patient/client's nutritional status and feeding abilities and to provide recommendations.
- Preventive dental treatment should be introduced to children with DMD at the earliest age possible. This includes regular prophylaxis, topical fluoride, and instruction on daily oral hygiene and home care (including proper diet), as well as restoration of hypoplastic and carious defects. However, lack of cooperation often makes dental treatment difficult in young patients/clients with DMD.
- Depending on the muscle groups affected by muscular dystrophy, modifications may need to be made to tooth brushing and other oral hygiene activities.

Oral manifestations

- Facial muscle weakness occurs, to varying degrees, in Duchenne, Becker, fascioscapulohumeral, oculopharyngeal, myotonic, and occasionally Emery-Dreifuss forms of MD. In FSHMD, inability to purse the lips occurs.
- The masticatory muscles are particularly affected in fascioscapulohumeral, oculopharyngeal, Duchenne, and myotonic forms of MD. Additionally, the masseter muscles may be affected in myotonic MD.
- Around the oral cavity, muscular atrophy occurs in DMD, often masked by overgrowth of adipose and connective tissues. This process results in hypertrophy (real and false) of the masseter muscles, orbicularis oris, and muscles of the tongue and lips. As hypotonia of the orbicularis oris worsens, patients/clients develop the habit of mouth breathing.
- Dysphagia is characteristic of oculopharyngeal MD, but swallowing difficulties also occur in other types, including DMD and BMD.
- Tongue weakness and atrophy occurs in OPMD. An enlarged, hypotonic tongue occurs in DMD.
- Malocclusion and temporomandibular joint (TMJ) disorder occur in mytonic dystrophy due to muscular weakness. Associated are difficulties in chewing and turning the head.
- Forward curvature of the neck ("swan neck") occurs in myotonic dystrophy due to weakness of the sternocleidomastoid

cont'd on next page...

MUSCULAR DYSTROPHY

(also known as "MD"; includes "Duchenne muscular dystrophy" [DMD], "Becker muscular dystrophy" [BMD], "oculopharyngeal muscular dystrophy" [OPMD], "fascioscapulohumeral muscular dystrophy" [FSHMD], "limb-girdle muscular dystrophy" [LGMD], "myotonic dystrophy" [DM], "Emery-Dreifuss muscular dystrophy" [EDMD], and congenital muscular dystrophy [CMD])

Oral manifestations *(cont'd)*

- Cross-bite and open bite malocclusion are frequently seen in DMD. As the patient/client ages, the open bite and increase in the angle between the maxilla and base of the mandible become increasingly pronounced. Flattening of the palate can also be seen, as can transverse widening of the alveolar arch.
- Drooling occurs in patients/clients with OPMD and some other forms of MD. This results from difficulty swallowing liquids, including saliva. Excess saliva, often accompanied by thick mucus, can cause choking and disrupt sleep.
- Reduced salivary secretion is seen in myotonic dystrophy type 1. Xerostomia also occurs in patients/clients with MD secondary to anticholinergic medications used to decrease salivary, bronchial, and gastric secretions, as well as secondary to certain antidepressants (e.g., amitriptyline) used to decrease saliva production and improve sleep pattern.
- Plaque and calculus formation, gingivitis, caries, and/or tooth loss occur at elevated rates in some forms of MD, likely related to reduced motor ability and other complicating factors.
- Dental erosion, thermal sensitivity, and pain can result from gastroesophageal reflux (GERD), which is commonly encountered in children with DMD.
- Disturbances in tooth form, number, and eruption occur in some patients/clients with Duchenne MD. Anterior and posterior open bites are also common, associated with lip incompetence, mouth breathing, macroglossia, and tongue thrusting. Dental arches tend to be hyperbolic, with posterior teeth being displaced buccally as a result of imbalance between lingual and facial musculature.
- Patients/clients who are tube-fed[6] are prone to low caries, rapid accumulation of calculus, GERD, and oral hypersensitivity.

Related signs and symptoms

- Muscular dystrophies are a group of genetically based neuromuscular disorders characterized by muscle necrosis and progressive muscle weakness. Over time, muscle weakness reduces mobility and makes activities of daily living difficult. Different types of MD affect specific muscle groups, have a specific age when signs/symptoms occur, vary in severity (i.e., mild to severe), and are caused by defects in different genes[7]. Currently there is no cure, although gene replacement therapy offers hope for future years[8].
- MD may run in a family (i.e., inheritable genetic defect), or the affected person might be the first family member to have the condition (i.e., new onset genetic mutation). Males are overall affected more than females, because of X-chromosome linked recessive forms of MD. In autosomal dominant forms of MD[9], males and females are equally affected.

6 The most common ongoing feeding tube for patients/clients with neuromuscular disorders is a percutaneous endoscopic gastrostomy (PEG) tube, which enables direct delivery of nutritional formulas to the stomach.

7 The disease is mostly caused by mutations in genes that code for proteins responsible for maintaining muscle fibre integrity for functional muscle contraction and relaxation.

8 In childhood-onset forms of muscular dystrophy, management includes early intervention services to improve a child's development. More generally, patients/clients with MD interact with one or more of the following: neurologist, rehabilitation specialist, physiotherapist, occupational therapist, respiratory therapist, clinical geneticist, paediatrician, primary care physician, dietitian, speech language pathologist, and surgeon (e.g., for scoliosis treatment). In DMD and BMD, specific therapeutic modalities include: corticosteroid treatment to maintain muscle strength as long as possible (particularly for DMD); stretching and other exercises; braces and splints; assistive devices such as wheelchairs, computer technology, and lifting devices; and surgery to prolong walking.

9 Duchenne, Becker, Emery-Dreifuss, and some forms of limb-girdle muscular dystrophies are X-linked, meaning that there is defective gene in the X-chromosome. Because males have XY sex chromosomes, the defective gene (in this case defectively coding for a protein necessary for proper muscle function) on the single X-chromosome manifests as muscular dystrophy. Because females have XX sex chromosomes, the second normal X chromosome usually results in enough protein production and no overt signs/symptoms of MD. Because a female can carry an X-linked gene mutation/defect and not be affected, she is referred to as a carrier who risks passing the same mutation on to her children.

cont'd on next page...

MUSCULAR DYSTROPHY

(also known as "MD"; includes "Duchenne muscular dystrophy" [DMD], "Becker muscular dystrophy" [BMD], "oculopharyngeal muscular dystrophy" [OPMD], "fascioscapulohumeral muscular dystrophy" [FSHMD], "limb-girdle muscular dystrophy" [LGMD], "myotonic dystrophy" [DM], "Emery-Dreifuss muscular dystrophy" [EDMD], and congenital muscular dystrophy [CMD])

Related signs and symptoms *(cont'd)*

- MD is rare, with poor data on how many persons are affected. Duchenne MD has an incidence of 1 in 3,500 births, while for Becker MD incidence is 1:20,000. The estimated combined prevalence of Duchenne and Becker muscular dystrophies[10] is 1 in every 7,250 males aged 5 to 24 years. In Canada, it is estimated that about 800 males between 0 and 24 years have Duchenne muscular dystrophy, which is the most severe form of MD.
- In Duchenne MD, muscle weakness in males typically begins before 5 years of age, whereas for Becker MD onset usually begins between 7 and 12 years. Muscle weakness in both types usually manifests first in the pelvis and femoral (thigh) regions, followed by the upper arms. The brain, throat, heart, diaphragm, chest muscles, stomach, intestines, and spine[11] may also be affected.
- In DMD, delays in motor development include crawling, sitting, standing, walking, running, and climbing stairs[12]. There is a tendency to fall and problems with joints locking in one position. Hypertrophy of the calf muscles is seen in children with DMD and BMD, coupled with a tendency to walk on the toes and lean backwards to maintain balance.
- Mild mental disability is observed in many persons with DMD, as well as in some persons with myotonic dystrophy.
- Myotonic dystrophy — the most common form of adult-onset muscular dystrophy[13] — is characterized by difficulty in relaxing the muscles after they have been contracted (i.e., myotonia). Cataracts are also common. While myotonic dystrophy type 1 affects 1 in 8,000 persons worldwide, prevalence of this autosomal dominant disorder is much higher in the Saguenay-Lac St. Jean (SLSJ) area of Quebec (1:500).
- Unlike most forms of muscular dystrophy that tend to manifest in childhood, adolescence, or early adulthood, oculopharyngeal MD typically manifests between 30 and 50 years of age. Persons of French-Canadian or Jewish ancestry are most frequently affected, with prevalence in the French-Canadian population of Quebec being 1 in 1,000 people. Signs/symptoms include progressive ptosis (drooping eyelids), diplopia (double vision), proximal muscle weakness, change in voice, and cachexia[14] resulting from dysphagia.
- Moderate to severe hearing loss can occur in young children with fascioscapulohumeral MD.
- Weakened bones (including osteoporosis) are a corollary of muscle weakness, lack of movement, and use of steroid medications. Risk of fracture is elevated, especially in the legs and spine in DMD. Scoliosis may cause breathing difficulties.
- Both Duchenne and Becker MD affect muscles in the heart, with progressive weakness as the patient/client ages. This cardiomyopathy may result in heart failure and/or arrhythmias such as atrial fibrillation.
- As patients/clients with DMD or BMD age, the muscles that support breathing weaken. Signs/symptoms include shortness of breath, fatigue, headaches, and difficulty sleeping. Mechanical breathing assistance may eventually be required for nighttime and daytime use.
- As DMD and BMD progress, the lungs weaken, elevating risk of pulmonary infections such as pneumonia. Manual and mechanical assistance may be required for coughing when the patient/client has a respiratory tract infection.
- Diet, nutrition, and digestion issues are common. These include being underweight or overweight, gastroesophageal reflux, and constipation. Swallowing problems can lead to unhealthy eating habits.

10 Becker MD is a milder and less progressive variation of Duchenne MD. Both conditions are caused by a mutation of the dystrophin gene, which causes a deficiency of dystrophin protein.
11 Scoliosis (abnormal lateral curvature of the spine) can result from muscle weakness in DMD and BMD.
12 Diagnosis of MD is based on clinical presentation, usually combined with tests such as electromyography (EMG, which measures electrical activity in the muscle), serum creatine kinase, muscle biopsy, genetic testing, electrocardiogram (ECG), and echocardiogram.
13 DM1 has both congenital (i.e., present at birth) and adult-onset forms. The latter tends to manifest signs/symptoms between 10 and 40 years of age. DM2 is a milder and more rare form of myotonic dystrophy.
14 Cachexia, or wasting syndrome, is characterized by weigh loss, muscle atrophy, weakness, fatigue and loss of appetite.

cont'd on next page...

MUSCULAR DYSTROPHY

(also known as "MD"; includes "Duchenne muscular dystrophy" [DMD], "Becker muscular dystrophy" [BMD], "oculopharyngeal muscular dystrophy" [OPMD], "fascioscapulohumeral muscular dystrophy" [FSHMD], "limb-girdle muscular dystrophy" [LGMD], "myotonic dystrophy" [DM], "Emery-Dreifuss muscular dystrophy" [EDMD], and congenital muscular dystrophy [CMD])

Related signs and symptoms *(cont'd)*

- Seizures occur in some forms of congenital muscular dystrophy.
- Edema of the lower legs and feet can result from reduced mobility of the lower limbs. This swelling may be accompanied by a burning sensation, and, if left untreated, can lead to permanent damage to the veins, valves, and skin.
- Muscle cramps, twitches, and spasms may cause inconvenience and pain.
- Mental and emotional health challenges (including depression and anxiety arise from the condition itself, interaction with others, and/or medications (e.g., corticosteroids) used to treat health issues.
- Reduced life expectancy is common in many forms of muscular dystrophy, typically resulting from pulmonary infections, respiratory failure, or cardiac causes. About 40% of males with Duchenne or Becker forms of MD (combined) die before 25 years of age.
- While the clinical course of Emery-Dreifuss MD is generally benign, sudden death is a frequent occurrence due to severe cardiomyopathy with conduction defects resulting in cardiac dysrhythmias.

References and sources of more detailed information

- Balasubraniam R, Sollecito TP, Stoopler ET. Oral health considerations in muscular dystrophies. *Spec Care Dentist*. 2008 Nov-Dec;28(6):243-53. doi: 10.1111/j.1754-4505.2008.00047.x. http://www.academia.edu/22294717/Oral_health_considerations_in_muscular_dystrophies
- Engvall M. On oral health in children and adults with myotonic dystrophy. *Swec Dent J Suppl*. 2010;(203):1-51. https://www.ncbi.nlm.nih.gov/pubmed/20514921
- RDH Magazine http://www.rdhmag.com/articles/print/volume-33/issue-4/features/neuromuscular-disorders-affect-the-oral-cavity.html
- Symons AL, Townsend GC, Hughes TE. Dental characteristics of patients with Duchenne muscular dystrophy. *ASDC J Dent Child*. 2002 Sep-Dec;69(3):277-83, 234. https://www.ncbi.nlm.nih.gov/pubmed/12613312
- Mielnik-Blaszczak M, Malgorzata B. Duchenne muscular dystrophy — a dental healthcare program. *Spec Care Dentist*. 2007;27 (1):23-25. http://www.parentprojectmd.org/site/DocServer/DMD_A_dental_care_program.pdf
- Romitti PA, Zhu Y, Puzhankara S, et al. Prevalence of Duchenne and Becker Muscular Dystrophies in the United States. *Pediatrics*. 2015;135(3):513-521. doi:10.1542/peds.2014-2044. https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4477633/
- Muscular Dystrophy Canada http://www.muscle.ca
- Canadian Neuromuscular Disease Network http://www.muscle.ca/wp-content/uploads/2012/11/CNDR_DMD_in_Canada_Report.pdf
- Centers for Disease Control and Prevention https://www.cdc.gov/ncbddd/musculardystrophy/facts.html
- Parent Project Muscular Dystrophy http://www.parentprojectmd.org/site/PageServer?pagename=Care_surgery

*Includes oral hygiene instruction, fitting a mouth guard, taking an impression, etc.

**Ontario Regulation 501/07 made under the *Dental Hygiene Act, 1991*. Invasive dental hygiene procedures are scaling teeth and root planing, including curetting surrounding tissue.

Date: October 25, 2017