

Types muscular dystrophy and management strategies



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Muscular Dystrophy

Abstract

Muscular dystrophy (MD) is comprised of a group of genetic diseases that cause muscle degeneration, progressive weakness and permanent shortening of tendons and muscles. The muscles affected will vary between the different types. MD's most common form in children, Duchenne MD, affects approximately 1 in every 3,500 to 5,000 boys in the United States. Most forms of MDs are inherited. There is no cure for any type of MD but there are some things that can help a person with MD live a fuller life.

Medical needs and lifestyle accommodations will vary depending on the age and degree of disability. Medical care can include a variety of specialists depending on the body systems that are affected. Physical, occupational, speech therapy and respiratory therapy are common interventions. There will be special considerations and self-care instructions for these patients because of progressive muscle weakness. As a dental professional it will be my goal to provide a welcoming environment and to ensure my patient has a positive experience.

Muscular Dystrophy (MD) is a group inherited diseases that cause necrotic degeneration of the muscle fibers. There are nine major groups of MD. The disease is characterized by progressive muscle weakness and degeneration of the skeletal muscles. The skeletal muscles that are affected vary depending on the type and pattern of the affected muscles. Some types also affect the cardiac muscle (Muscular Dystrophy: Hope through Research, 2018). The progression of the disease varies depending on the age at onset

and severity of the particular type of MD. Some forms occur during childhood while others occur as an adult. All types are characterized as a progressive disease because the muscles continue to degenerate. The cause of MD is a genetic mutation that can either be passed from a parent or can be a spontaneous mutation. The mutations can be autosomal dominant, autosomal recessive and X-linked (Naseem, 2016). The defective gene prevents the body from making a protein that builds and maintains muscles. Duchenne Muscular dystrophy usually isn't caused by a familial defect, typically it is a spontaneous mutation. The gene that becomes mutated in this disease is larger than most so this makes it more likely to be mutated. Dystrophin complex is a group of proteins in the muscle fiber membranes that helps to keep muscles strong. When the membrane is damaged from a mutation, the muscles leak creatine phosphokinase (CPK). This causes the muscles to acquire extra calcium and this damages the muscle fibers. The fibers eventually die which is the cause of the progressive weakness and muscle degeneration. The (CPK) level is the most important blood test to do when myopathy is being investigated (Naseem, 2016). The tendons reflex and muscle strength are affected and the lost muscle is replaced by connective tissue and fat (Muscular Dystrophy: Hope through research, 2018).

Types of Muscular Dystrophy

Duchenne MD is the most common form of MD and makes up around 50% of MD (Muscular Dystrophy: Hope through Research, 2018). This is also the most common type that affects male children. The prevalence is around 1 in 3,500 births (Naseem, 2016). This type is usually diagnosed when the child

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begins to walk. Weakness begins in the thigh muscles and pelvis and then spreads to the biceps and triceps. The child starts walking with a waddling gait. The calf muscles become enlarged from fat and connective tissue replacing muscle mass. This type also affects the respiratory system by causing lung weakness, breathing difficulty and swallowing problems. As the disease progresses, the diaphragm will weaken. Cognitive decline affects some children with this form of the disease. This type affects the orofacial muscles and can lead to dysphagia, so it is important for dental hygienists to be familiar with this form. The cardiac muscle will have abnormalities in the muscle fibers that cause cardiomyopathy. With this form of MD, most children are unable to walk by the time they are in their early teens. Adolescents with this form of MD typically die in their late teens or early twenties unless they have intensive multidisciplinary care. With this care, some can survive to their thirties or forties (Muscular Dystrophy: Hope through research, 2018).

Becker MD is a form that is typically diagnosed around the age of eleven but can be as late as the mid-twenties. This form's progression is more varied than the Duchenne MD. The progression ranges from some not being able to walk when they are teenagers to walking well into their thirties. Some never have to use a wheel chair at all. Becker MD doesn't always have cardiac involvement and cognitive impairment is less common. Some of the first signs are walking on the toes, falling frequently and having a difficult time rising from the floor. As with Duchenne MD, the calf muscles also become enlarged (Muscular Dystrophy: Hope through Research, 2018).

Congenital MD affects both sexes and symptoms become apparent by the age of 2. This type varies between people and may be mild to severe. The first signs are noticed when the child doesn't meet milestones for their age. Some children affected by this type of MD never learn to walk. Vision, speech and cognition may be affected. Some children may experience seizures. This type of MD symptoms are greatly varied between people with some children dying while still in infancy and others living until into adulthood with only slight disabilities (Muscular Dystrophy: Hope through research, 2018).

Distal MD is a less severe and slower progressing form of MD. It affects men and women and typically becomes evident between the ages of 40-60 years old. It affects fewer muscles than most other types of MD, mostly the lower arms, legs and feet, cardiac and respiratory muscles. Fine motor hand movement is affected (Muscular Dystrophy: Hope through research, 2018).

Emery-Dreifuss MD causes progressive weakness that manifests symmetrically. Muscle contractures appear in the spine, neck, knees and elbows. Some muscles such as the elbows and spine may become locked and rigid. Almost every person with this type of MD has cardiac problems (Muscular Dystrophy: Hope through research, 2018).

Facioscapulohumeral MD weakness initially begins in the face, shoulder and arms. Muscles around the mouth and eyes tend to be the first affected. Dysphagia and chewing difficulties are early signs of the disease. The onset can be anywhere from childhood to 40 years of age. Most people live a normal lifespan but have disabilities which are severe. Hearing problems and

curvature of the spine are common with this type of MD (Muscular Dystrophy: Hope through research, 2018).

Limb-Girdle MD affects both males and females and usually becomes evident anywhere from childhood to the teen years. Most people have severe disabilities 20 years after the initial onset. It affects the muscles in the hip, shoulders, legs and neck. Some forms are rapidly progressing while others progress slowly. Cognition is usually not affected. Some people develop cardiomyopathy and respiratory problems (Muscular Dystrophy: Hope through research, 2018).

Myotonic MD (Steinert's Disease) has a unique feature called myotonia. This is the sudden contraction of a muscle followed by the inability to relax the muscle. This slowly progressing form can show up anywhere from childhood until the age of 30. Facial muscles are the first to be affected followed by the forearms. This type affects many body systems including the cardiovascular, endocrine, nervous and gastrointestinal tract. Vision can also be affected (Muscular Dystrophy: Hope through research, 2018).

Oculopharyngeal MD occurs in both sexes beginning in the forties. It begins with drooping eyelids, and then progresses to the pharyngeal muscles and the tongue. Weakness in the shoulders and neck muscles soon becomes apparent. This type of MD is important for hygienists to be familiar with because it involves oral muscles (Muscular Dystrophy: Hope through Research, 2018).

There is no cure for any type of MD but there are some things that can be one to help people with the disease live a fuller life. Medical needs and <https://assignbuster.com/types-muscular-dystrophy-and-management-strategies/>

lifestyle accommodations will vary depending on the age and degree of disability. Medical care can include a variety of specialists depending on the body systems that are affected. Pulmonologists, cardiologist and neurologists are commonly seen. Physical, occupational, speech and respiratory therapy are common interventions. Some commonly prescribed medications for MD are corticosteroids, anticonvulsants, immunosuppressants, cardiac and antibiotics to treat respiratory infections (Muscular Dystrophy: Hope through Research, 2018). Corticosteroids are standard therapy but they can have serious side effects such as bone demineralization, hypertension and growth retardation. A peer reviewed study published in 2016 assessed the efficacy of some nutraceuticals for the treatment of MD. CoQ10 was found to reduce reactive oxygen species and prevent calcium accumulation in the muscles and taurine significantly improved grip strength (Woodman2016). This study concluded that these supplements could have significant potential as complimentary therapy against counteracting chronic inflammation or oxidative stress (Woodman2016). More studies should be done to compare the effects against of nutraceuticals compared to corticosteroids.

Most people will need some degree of help with activities of daily living depending on how far the disease has progressed. Some people may have a full time caregiver and others may have part time help or even a service dog. Wheelchairs, orthotic devices, lifts, bathroom aids and other adaptive devices help to maintain as much independence as possible. Children with cognitive decline may qualify for an individualized education plan (IEP) through the school system that will help them get extra help and

accommodations (Special Needs Fact Sheet: Muscular Dystrophy, 2015).

Special adaptive devices such as keyboards and large diameter writing utensils are helpful for children in school.

When a patient with muscular dystrophy comes to the dental office, there will be special considerations. If the patient is taking immunosuppression medications, medical clearance from their physician may be needed (Balasubramaniam, 2008). Shorter appointments may be the best option for these patients because they tend to get fatigued easily. Some aspects such as assessments and communication may take longer than usual so a series of short appointments can be a good option. It is best to ask the patient what suits their needs. The office should be easy to navigate, free from barriers and should always accommodate those in the wheelchair. The patient shouldn't ever feel rushed, or that their disability is bothersome. If the office has the ability to treat the patient in their wheelchair, the patient should be asked what they prefer. Dental chair adjustments and support cushions may be helpful. Certain types of MD affect the facial and neck muscles so this will particularly affect the way dental care is given. A bite block may be an essential part of treatment for those that are unable to keep their mouth open and to reduce stress on the muscles. The patient should be told that they can postpone treatment at any time during the appointment if they need to. Patients may need frequent position changes may help with patient comfort. Four handed dental hygiene may be helpful for efficiency and safety of the patient. Adequate suctioning is very important because of dysphagia and the possibility of aspiration. Rubber dams may be useful to avoid inhalation of foreign objects during treatment. A referral to a nutritionist may

be appropriate if the patient isn't getting proper nutrition because of dysphagia and muscle weakness.

Oral health education should be in depth and tailored to the degree of muscle weakness. If the patient has a caregiver that assists in their oral care regimen, they should present if possible. The hygienist should teach the caregiver techniques as well. Modifications may need to be made because of muscle weakness in the hand and wrist muscles. Patients may benefit from using a power toothbrush because of the large diameter handle and increased biofilm removal. Some models feature a texture on the handle that enhances the grip. Water flossers should be recommended because they are easier to manipulate and the flushing action of the water will benefit this patient. A dentifrice with fluoride should be used if the patient is able to expectorate adequately. If they cannot, a home fluoride regimen should be implemented. Many oral manifestations associated with MD relate to a decrease in muscle function. An increase in calculus and biofilm accumulation is often seen because of weakness of the tongue muscles (Symons, 2002). Food remaining in the vestibule is another consequence of weakness of the tongue muscle. A high prevalence of malocclusion and class III relationships are seen in MD (More-Verdebout). Disturbances in tooth form, number and eruption of the second premolars were observed in 39% of patients (Symons, 2002). Mouth breathing, macroglossia, tongue thrusts, cross bites and anterior and posterior open bites are common. Both arches are widened posteriorly with the lower arch wider than the upper which results in frequent posterior crossbite (Morel-Verdebout). Posterior teeth tend to be buccally displaced. Patients who are taking anticonvulsants may

experience gingival hyperplasia. A bite splint may need to be fabricated as a result of the high incidence of temporomandibular joint disease.

The dental hygiene care plan should include frequent recall intervals due to the high possibility of biofilm and calculus accumulation. Intensive oral hygiene education should be personalized for the disability and areas that need special attention. This should be done frequently because this disease is progressive so modifications may be needed more often. Nutritional counseling to reduce cariogenic choices should be included in counseling.

I have become more aware of the special needs and adaptations needed to treat a patient with muscular dystrophy. A basic human need is to feel safe and have a sense of belonging, I believe this is especially true for those with disabilities. I enjoy building a rapport with all of my patients but I think it is especially important with special needs patients. The hygienist may be the only personal interaction they have that day other than a caregiver. Some patients may be fearful of going to the dentist and others may anticipate the simple opportunity to talk and interact. We should be able to recognize not only the dental needs of the patient but also the psychological ones. One of our goals is to make their dental experience positive so they will want to return. A patient may not like going to the dentist but your demeanor and empathy just may be the deciding factor on whether they have positive or negative feelings about going to the dentist.

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