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PATIENTS WITH DUCHENNE MUSCULAR DYSTROPHY IN A DENTAL SETTING - A REVIEW ARTICLE

Dentistry

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KEYWORDS

INTRODUCTION

Muscular dystrophies are a collection of inherited degenerative disorders impacting the skeletal muscle structure, leading to a range of clinical symptoms such as advancing dystrophic alterations in muscle tissue. Over time, these conditions systematically break down muscle cells, replacing them with connective tissue, leading to a gradual decline in muscle strength and functionality. Common features of muscular dystrophies involve muscle wasting in various regions like the axial, facial, and limbs. Additionally, degenerative changes can extend to other muscle groups such as those in the heart, respiratory system, swallowing, and eyelids. [1] DMD is an exceedingly rare disease, with a 2020 meta-analysis reporting the birth prevalence of DMD as 19.8 per 100,000 live male births.[2] The occlusal force of DMD patients decreases to 8.1 18.9 kg, as compared to 52.2 kg in healthy subjects. [3]

Degeneration of the perioral muscles causes sagittal contraction and lateral expansion of the dental arch, protrusion of the maxillary incisors, retrusion of the mandibular incisors, lateral mandibular expansion beyond the maxilla, and abnormal facial morphology due to excessive vertical growth. [4]

ORAL MANIFESTATIONS

DMD is an inherited myopathy, in which a genetic mutation is observed in the Xp locus, in the short arm of the X chromosome. DMD is an X-linked disease caused by the absence of a protein named dystrophin, which is found more frequently in skeletal muscles and neurons in specific regions of the central nervous system.

The initial DMD symptoms appear in childhood (usually around 5 years of age) with difficulty in walking. The most prominent symptoms include severe progressive muscle weakness in the pelvic, shoulder, upper, and lower limbs. [5]

Duchenne muscular dystrophy (DMD) is a severe X-linked recessive disorder primarily affecting males, characterized by progressive muscle degeneration and weakness due to mutations in the dystrophin gene. While the primary focus of DMD is on skeletal muscle deterioration, it also presents significant dental manifestations.

Research has shown that individuals with DMD often experience various dental abnormalities, including malocclusion, delayed eruption of permanent teeth, and abnormal tooth morphology.[6] The weakening of perioral muscles can lead to sagittal contraction and lateral expansion of the dental arch, protrusion of the maxillary incisors, retrusion of the mandibular incisors, and lateral mandibular expansion beyond the maxilla.[7]

Moreover, DMD-related muscle weakness can result in poor oral hygiene practices, predisposing individuals to dental caries and periodontal disease.[8] It is crucial for dental professionals to be aware of these manifestations and collaborate with multidisciplinary teams to provide comprehensive care for individuals with DMD.

CLINICAL INDICATIONS

The initial clinical indications usually manifest between the ages of 2 to 6 years, as the child's physical capabilities start to deviate from those of their peers. These early signs commonly include delayed onset of walking, challenges in running or ascending stairs, and an increased frequency of falls.

A characteristic observation during this stage is known as *Gowers' sign*, which describes the necessity for a patient to utilize their hands and arms to ascend from a squatting position due to weakness in the hip and thigh muscles. Over time, muscle strength progressively declines, often necessitating the use of a wheelchair around the age of 10. By the early teenage years, cardiac and respiratory muscles become affected, culminating in potential complications such as cardiac or respiratory failure, which may ultimately lead to fatality. [9]

Children with DMD often exhibit delayed motor milestones, such as delayed walking, and may experience difficulties with running or climbing stairs. As the disease progresses, muscle weakness becomes more pronounced, leading to an inability to walk independently, usually by early adolescence. Additionally, DMD can affect respiratory and cardiac muscles, resulting in symptoms such as shortness of breath, respiratory insufficiency, and cardiomyopathy. [10,11]

LOCAL ANESTHESIA IN PATIENTS WITH DMD

Local anesthesia is a crucial component of dental procedures, providing pain control and facilitating patient comfort. However, in patients with Duchenne muscular dystrophy (DMD), careful consideration must be given to the administration and management of local anesthetics due to potential complications arising from the underlying muscle weakness and associated comorbidities.

Patients with DMD may present with compromised respiratory function, cardiac involvement, and musculoskeletal abnormalities, which can impact their tolerance and response to local anesthesia. Therefore, an in-depth preoperative assessment is essential to evaluate the patient's medical history, current medications, and overall health status.

When administering local anesthesia in patients with DMD, practitioners should consider utilizing techniques that minimize systemic absorption and reduce the risk of adverse effects. This may involve using lower doses of anesthetic agents, choosing anesthetic formulations with shorter durations of action, and employing alternative routes of administration, such as intraosseous or intraoral techniques, to minimize tissue trauma and maximize efficacy. [12]

Despite the challenges associated with administering local anesthesia in patients with DMD, effective pain management remains essential for ensuring a positive dental experience and maintaining oral health. By employing tailored approaches and adhering to best practices, dental practitioners can mitigate risks and provide quality care for individuals with DMD undergoing dental procedures. [13]

TREATMENT MODALITIES

Duchenne muscular dystrophy (DMD) is a progressive genetic disorder with no known cure, but various treatment modalities aim to manage symptoms, improve quality of life, and slow disease progression. Multidisciplinary care involving neurologists, physical therapists, respiratory therapists and orthopedic specialists is essential in addressing the needs of individuals with DMD.

One of the cornerstones of DMD management is corticosteroid therapy, typically with prednisone or deflazacort. Corticosteroids have been shown to prolong ambulation, preserve muscle function, and delay the onset of respiratory and cardiac complications in DMD patients. However, they are associated with adverse effects such as weight gain, osteoporosis, and behavioral changes necessitating careful monitoring and dose adjustment.[14]

Physical therapy and exercise play a crucial role in maintaining mobility, preventing contractures, and preserving muscle function in DMD patients. Regular stretching exercises, range-of-motion activities, and low-impact aerobic exercises can help optimize muscle function and delay functional decline.

Respiratory management is another vital aspect of DMD care, as respiratory muscle weakness can lead to respiratory insufficiency and pneumonia. Non-invasive ventilation (NIV) strategies, such as bilevel positive airway pressure (BiPAP) or continuous positive airway pressure (CPAP), may be used to support breathing and improve oxygenation in DMD patients with respiratory compromise.[11]

Cardiac monitoring and intervention are crucial in DMD due to the risk of cardiomyopathy and heart failure. Regular cardiac evaluations, including electrocardiograms (ECGs) and echocardiograms, are recommended to detect early signs of cardiac dysfunction. Pharmacological interventions, such as angiotensin-converting enzyme (ACE) inhibitors and beta-blockers, may be prescribed to manage cardiac symptoms and delay disease progression.[15]

Difficulties Faced While Treating Patients With DMD

Treating patients with Duchenne muscular dystrophy (DMD) poses numerous challenges due to the complex nature of the disease and its progressive course. One significant difficulty is the multi-system involvement seen in DMD, which requires a multidisciplinary approach involving various healthcare professionals to address the diverse needs of patients.

Muscle weakness and degeneration in DMD present obstacles in administering certain treatments and procedures. For example, difficulties may arise in achieving adequate muscle relaxation during surgery or dental procedures due to the weakened state of the muscles, making anesthesia management challenging.

Managing the respiratory complications of DMD also presents significant challenges. Respiratory muscle weakness can lead to respiratory insufficiency, recurrent respiratory infections, and eventual respiratory failure. Implementing effective respiratory support measures, such as non-invasive ventilation (NIV) or cough-assist devices, requires close monitoring and specialized training to optimize outcomes and prevent respiratory complications.[14,15]

Furthermore, cardiac involvement in DMD, including the development of cardiomyopathy and heart failure, adds another layer of complexity to patient care.[16]

COMPLICATIONS DURING TREATMENT

One of the most prominent complications in treating patients with DMD is the risk of respiratory insufficiency and failure. Respiratory muscle weakness, particularly involving the diaphragm, can lead to decreased lung function, impaired coughing ability, and an increased susceptibility to respiratory infections. As the disease progresses, individuals with DMD may require respiratory support, such as non-invasive ventilation or mechanical ventilation, to maintain adequate oxygenation and ventilation.[17]

Cardiac complications are another significant concern in DMD management. Cardiomyopathy, characterized by progressive deterioration of cardiac function and muscle weakness, commonly develops in individuals with DMD. As a result, patients are at increased risk of arrhythmias, heart failure, and sudden cardiac death. Regular cardiac monitoring, including electrocardiograms (ECGs) and echocardiograms, is essential to detect early signs of cardiac dysfunction and initiate appropriate interventions.[11]

Psychosocial complications also play a significant role in DMD management. The progressive nature of the disease, combined with physical limitations and healthcare demands, can lead to emotional distress, anxiety, and depression in patients and their families.[14]

In conclusion, treating patients with DMD involves addressing a wide range of complications across multiple organ systems. A multidisciplinary approach, encompassing medical, rehabilitative, and psychosocial interventions, is essential in optimizing care and improving outcomes for individuals living with DMD. Ongoing research and advancements in treatment strategies are crucial in addressing these complications and enhancing the quality of life for patients with DMD.

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