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Mini Review

A Mini-Review on Respiratory Complications of Duchenne Muscular Dystrophy

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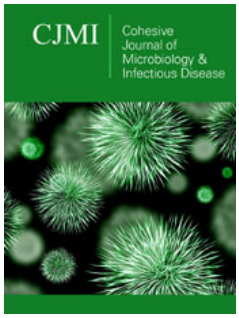

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Abstract

Duchenne muscular dystrophy (DMD) is a rare, severe, progressive genetic disorder causing disability and premature death. Mutations in the DMD gene encoding the dystrophin protein leads to the dystrophinopathies DMD. It affects approximately 1 in 3500 to 5000 male births worldwide. Respiratory complications, such as pneumonia and respiratory failure, are the leading cause of morbidity and mortality for patients with DMD. In this mini review, we discuss respiratory care of the patient with DMD.

Keywords: Duchenne muscular dystrophy; Pneumonia; Infections; COVID-19

Introduction

Duchenne muscular dystrophy, a rare X-linked disorder, is caused by a genetic mutation that prevents the body from producing dystrophin [1], a protein that enables muscles to work properly. It is one of the most common types of muscular dystrophy. DMD symptom onset is in early childhood, usually between ages 3 and 5 years. Over time, children with Duchenne will have difficulty walking and breathing, then lead to disability, dependence, and premature death. The disease primarily affects boys, but in rare cases, it also can affect girls. The prevalence of DMD is approximately 1 in 3500 to 5000 male births worldwide [2]. Muscle weakness is the principal symptom of DMD and worsens over time, first affecting the proximal muscles and later affecting the distal limb muscles. Patients with DMD progressively lose the ability to perform activities independently and often require a wheelchair by their early teens. As the disease progresses, life-threatening heart and respiratory conditions can occur [3]. In general, patients succumb to the disease in their 20s or 30s [3]; however, disease severity and life expectancy can vary. DMD is caused by mutations in the DMD gene, which encodes the protein product called dystrophin. DMD gene is one of the largest of the identified human genes, spanning 2.4Mb of a genomic sequence and corresponding to about 0.1% of the total human genome [4]. The gene consists of 79 exons encoding a 14,000 bp messenger RNA transcript that is translated into dystrophin [5]. The most common mutation responsible for DMD is a deletion spanning one or multiple exons. Such deletions account for 60-70% of all DMD cases. Point mutations are responsible for around 26% of DMD cases. Exonic duplications account for 10 to 15% of all DMD cases [6]. Mutations in the DMD gene disrupt the protein's reading frame causing premature stop codons, leaving little or no functional dystrophin protein produced in cells. Respiratory complications, such as pneumonia and respiratory failure, are the leading cause of morbidity and mortality for DMD patients. Respiratory muscle weakness is found in all individuals with DMD, although it may develop at varying rates [7]. Due to weakened cough, individuals with Duchenne are at a higher risk for pneumonia. With progressive loss of muscle strength, and it may be more difficult to remove this mucus (secretions) from lungs. The inability to remove mucus from lungs puts DMD patients at risk for pneumonias or other respiratory infections.

To prevent respiratory illnesses, individuals with DMD should receive immunization with the inactivated influenza vaccine yearly and pneumococcal vaccines as advised by your

primary care provider [8,9]. Oral hygiene is important to prevent bacteria in the mouth that can cause chest infections as well. There are also several ways to assist your cough, both manually and with a machine, help DMD patients to clear secretions from lungs. During acute respiratory illnesses, antibiotic therapy should be added for individuals who have 3 of the following 5 clinical signs of pneumonia: fever, elevated white blood cell count or C-reactive protein level, sputum production, a pulmonary infiltrate on the chest radiograph, or hypoxemia or respiratory distress [10]. In the late stages of DMD, individuals require assisted ventilation to prolong their survival. The novel coronavirus (COVID-19) outbreak has caused a global pandemic. As individuals with Duchenne are immunocompromised (if on steroids) and have respiratory problems, this may put them at a higher risk of developing complications from COVID-19 if infected. Patients with DMD and their families should follow current national, state, and local guidelines for people at risk for serious illness from COVID-19 [11,12]. DMD patients should continue their current treatments, and specifically should not discontinue existing medications, unless approved by their treating neurologist or neuromuscular specialist. All decisions regarding treatment should be individualized and jointly considered between the patient, family, and providers [13].

Conclusion

Respiratory complications, such as pneumonia and respiratory failure, are the leading cause of morbidity and mortality for DMD patients. Managing respiratory health in individuals with DMD is vital to improve patient quality of life and prolong survival.

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