

Severe Mid-Face Retrognathism following
BiPAP use in a Patient with Muscular
Dystrophy

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Abstract

Aim: To describe severe facial disfigurement in a patient with familial Progressive Muscular Dystrophy (PMD) treated with a Bi-level Positive Airway Pressure (BiPAP) device.

Study design and methods: A 41-year-old female with PMD was treated with BiPAP from the age of 21 in order to improve sleep-disordered breathing and nighttime hypoventilation.

Results: Severe mid face retrognathism was noted with a reverse over jet between upper and lower incisors of 12 millimeters in centric relation.

Conclusion: We present a rare case of severe facial disfigurement secondary to orthopedic forces from a BiPAP device in a patient with familial PMD. The simple addition of a forehead or chin support may minimize these changes.

Introduction

Progressive Muscular Dystrophy (PMD) is a neuromuscular disease of muscle weakness and wasting. The most common form is Duchene muscular dystrophy, an x-linked recessive condition associated with a mutation in the dystrophic gene. Respiratory muscles become weaker, causing an ineffective cough and decreased ventilation, which may lead to respiratory insufficiency and eventually to morbidity and mortality. Patients with PMD have an increased risk of sleep-disordered breathing, including central and Obstructive Sleep Apnea (OSA), and hypoxemia.

OSA is a disorder characterized by frequent repetitive events of the upper airway blockage during sleep, it is associated with significant adverse health-related outcomes and therefore treatment is crucial, particularly in patients with respiratory muscle weakness.

The treatment of the pulmonary complications associated with PMD with noninvasive ventilatory support may improve quality of life and reduce morbidity and mortality. Continuous or Bi-Level Positive Airway Pressure (CPAP or BiPAP) generators or mechanical ventilators have been used successfully in the treatment of sleep-disordered breathing and night-time hypoventilation in patients with neuromuscular disorders [1,2]. This is a kind of pump device connected to a mask that increases air pressure continuously in the upper airways to prevent their collapse during sleep. As PMD progresses patients enter a state of constant hypoventilation, and require 24-hour positive pressure ventilation. An invasive ventilator placed via endotracheal or tracheostomy tube may become necessary. In less severe cases, non-invasive delivery via a face/nasal mask or mouthpiece can be used [3]. We present a severe, non-age dependent craniofacial disfigurement in a patient with PMD due to long-term use of a BiPAP mask.

Case Presentation

A 41-years old female paralyzed and confined to a wheelchair due to a mild Duchene like form of Familial PMD presented to the clinic. The patient could breathe spontaneously, eat, speak and move one finger, enabling her to control the wheelchair. Her presenting complaint was difficulty eating due to changes in her bite and facial esthetics. From the age of 21 the patient used a BiPAP like ventilatory machine with a nasal mask during sleep (especially at the beginning) and as time progress also during the day hours, the patient couldn't remember when she started using the mask also during the day. Upon extra and oral examination, severe mid face, retrognathism (Figure 1) was noted with a reverse over jet between upper and lower incisors of 12 millimeters in centric relation (Figure 2). The severe reverse over jet interfered with chewing and had a negative esthetic impact. No treatment was suggested of done to minimize or repair this problem. Pictures were taken following patient consent.

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Figure 1: Reverse over jet between upper and lower incisors due to the position of the BiPAP mask on the mid-face area.



Figure 2: Mid-face retrognathism.

Discussion

Positive airway pressure pumps are the first line of treatment for OSA in adults, and have specific indications for children such as craniofacial anomalies, obesity, and neuromuscular weakness [4]. These devices may cause facial deformities, especially when used before growth is complete [5]. Permanent changes to the facial bones from CPAP\ BiPAP are rare in healthy full-grown individuals. One report examined this phenomenon in adults and found minor statistically significant changes [6]. PMD patients particularly those

with Duchene, have a distinct anatomical dental structure with an extensive open bite and wide dental arches, but these are not the changes we are referring to in this presentation. We believe that the severe maxillary disfigurement noted, was caused by the orthopedic forces applied by the nasal mask, causing a “concave face”. Her muscle weakness may have enabled relatively greater and more direct forces of the mask on the bone, causing severe structural changes even though BiPAP treatment began at the age of 21. This malformation could also result from either discontinuing of normal growth or due to an active retrognathic force. Even though, several factors associate with the syndrome of PMD, could have further influence patient facial characteristics, among them: treatment continued also during waking hours (far longer than average normal sleep time), patient’s natural oro-motor function and mouth breathing. These factors cannot be ignored and may have contribute to facial deformity. A large-scale study including patients with severe muscle weakness, aim to compare facial characteristics of CPAP users to non-CPAP users is indicated.

Conclusion

Patients with severe muscle weakness may be at risk for significant craniofacial disfigurement caused by direct orthopedic forces of the BiPAP or CPAP masks on the maxillary bone. The simple addition of a forehead or chin support or a regular full-face mask is recommended instead of a nasal mask to spread the orthopedic force more evenly and reduce maxillary deformity.

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