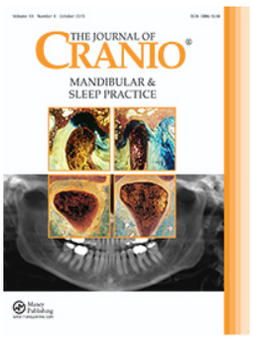

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CASE REPORT



# Maxillary deformity following CPAP treatment in myasthenia gravis

Yaron Haviv DMD PhD[a], Shirley Leibovitz DMD MA[b], Galit Almoznino DMD MSc MHA[a,c], Yair Sharav DMD MS[a] and Uri Zilberman DMD PhD[b]

[a]Department of Oral Medicine, Sedation & Maxillofacial Imaging, Hebrew University Hadassah School of Dental Medicine, Jerusalem, Israel; [b]Department of Pediatric Dentistry, Barzilai Medical Center, Ashkelon, Israel; [c]Department of Oral Medicine, Oral and Maxillofacial Center, Medical Corps, Israel Defense Forces, Tel-Hashomer, Israel

**ABSTRACT**

**Objective:** Patients with Myasthenia gravis (MG) are characterized by muscle weakness that may cause obstructive sleep apnea (OSA). The use of a continuous positive airway pressure (CPAP) device is often needed in order to maintain free breathing during the night and sometimes even during the day.

**Clinical Presentation:** A 29-year-old MG patient is presented who used a CPAP continuously since the age of 12. Tight fitting of a nasal mask applied enough force to cause severe maxillary deformity.

**Clinical Relevance:** Masks with additional forehead and chin support or a regular full face mask are recommended for patients with muscle weakness, in order to spread forces more evenly.

**KEYWORDS**

Myasthenia gravis; obstructive sleep apnea; CPAP; maxillary deformity

## Introduction

Myasthenia gravis (MG) is an autoimmune disorder of neuromuscular transmission, caused by the destruction of acetylcholine receptors, leading to fluctuating degrees of muscle weakness and fatigability [1]. Facial characteristics of MG patients may include episodes of "open-lock," jaw drop, elongated face, open bite, nasal regurgitation of liquids, and hyper-nasal speech. Mandibular prognathism with Class III malocclusion was observed [2], even though Class I and Class II were also identified [3]. In up to 36% of MG patients, muscle weakness can cause obstructive sleep apnea (OSA), a disorder characterized by frequent repetitive events of the upper airway blockage during sleep [4]. OSA is associated with significant adverse health-related outcomes [5], and therefore treatment is crucial, particularly in patients with respiratory muscle weakness, such as MG patients. The gold standard of OSA treatment is continuous positive airway pressure (CPAP), a pump device connected to a mask that increases air pressure continuously in the upper airways to prevent their collapse during sleep [6]. CPAP use may successfully improve morning symptoms as well as quality of life in MG patients [4,7]. However, CPAP may exert a constant orthopedic restraining force on the mid-face. The authors describe a patient with MG, displaying a craniofacial deformity due to nasal CPAP use.

## Case presentation

A 29-year-old male attended the Department of Oral Medicine for dental treatment and orthodontic consultation. The patient was diagnosed with generalized MG, was partly paralyzed and confined to a wheelchair. The patient used CPAP during sleep and while resting for approximately 12 h a day since he was 12 years old. Tight fitting of the nasal mask applied enough force to cause severe maxillary deformity (Figure 1).

A lateral digital cephalometric X-ray was taken in centric occlusion in the upright position, keeping the Frankfort horizontal plane parallel to the floor. Following data exportation to the computer, Eastman analysis (cephalometric standard for Caucasians) was performed for orthodontic diagnosis (Figure 2). The results of the analysis are shown in Table 1. The findings revealed a Class III relationship (ANB = −1.12°) with a Class III skeletal problem, Wits appraisal = −2.8 mm (A method of assessing sagittal or A-P jaw relationship, which is independent of apical base relationship to cranial landmarks). Other findings revealed: upper incisors protruded forward (UI-PAL. Plane = 140.22°), lower incisors also protruded (Li to MAND = 101.38°), protruded incisors relationship (Inter-incisal Angle = 96.18°), upper face height too high (N-NAS = 53%), lower face height too low (ASN-Gn = 47.4%), and horizontal growth tendency (FH

**CONTACT** Yaron Haviv yaron.haviv@gmail.com

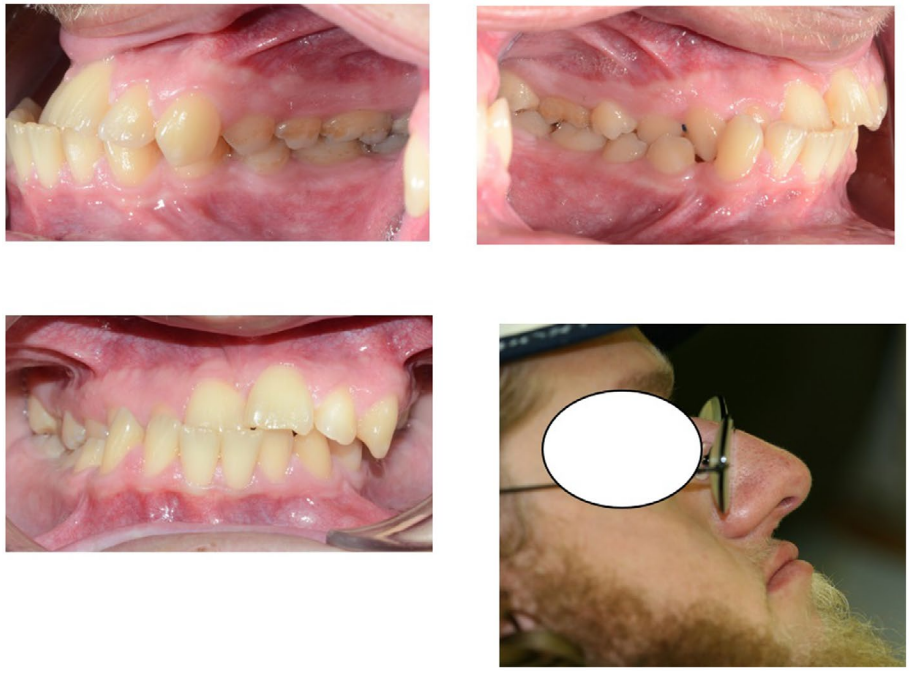

**Figure 1.** Intra oral and extra oral images.

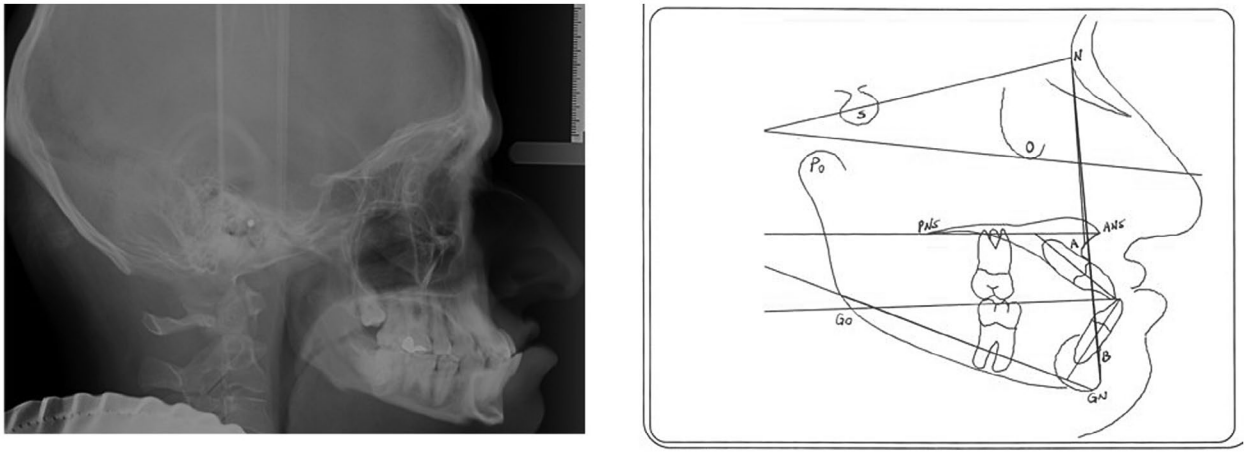


**Figure 2.** Cephalometric X- ray (left) and analysis (right).

**Table 1.** Eastman Analysis.

| Descriptor | Type | Mean | SD | Patient | Graph | Comment |
|---|---|---|---|---|---|---|
| SNA | Degrees | 82 | 2.0 | 80.71 | –(*)+ | |
| SNB | Degrees | 80 | 2.0 | 81.83 | –(*)+ | |
| ANB | Degrees | 2 | 2.0 | –1.12 | –(*\|)+ | Class III relationship |
| WITS | mm | 0 | 1.0 | –2.8 | –(* \|)+ | Class III Skeletal problem |
| Upper incisor – palate plane | Degrees | 110 | 5.0 | 140.33 | –(\| *)+ | Upper incisor inclined |
| Lower incisor to mandible | Degrees | 90 | 3.7 | 101.38 | –(\| *)+ | Lower incisor too protruded |
| Inter incisal Angle (/1 to 1/) | Degrees | 131 | 13.0 | 96.18 | –(* \|)+ | Protruded Incisor relationship |
| PL-mandibular Plane | Degrees | 25 | 3.0 | 22.09 | –(*)+ | |
| Upper face height (N-ANS) | % | 45 | 3.0 | 53.0 | –(\| *)+ | Upper face height too large |
| Lower face height (ANS-Gn) | % | 55 | 3.0 | 47.4 | –(* \|)+ | |
| Ratio | % | 55 | 3.0 | 47.4 | –(* \|)+ | Lower face height too small |
| Li-APOG | mm | 1 | 2.0 | 8.33 | –(\| *)+ | Lower incisor inclined forward |
| Lower lip to E-LINE | mm | –2 | 2.0 | –1.13 | –(*)+ | |
| Y axis (FH-to S-Gn) | Degrees | 59.4 | 3.8 | 44.62 | –(* \|)+ | Horizontal growth tendency |

Notes: WITS measures the disharmony between the AP degree, by drawing perpendicular lines connecting A point and B point to the Occlusal plane (AO and BO). The greater the WITS reading, the greater the jaw discrepancy.

SD = Standard deviation; SNA = sella-nasion-A point angle; SNB = sella-nasion-B point angle; ANB = A point-nasion-B point angle; ANS = anterior nasal spine; Gn = Gnathion; FH = Frankfort Horizontal.

to S-Gn = 59.4°). Soft tissue analysis revealed a retruded upper lip (Upper lip to E-line = −10.66 mm).

## Discussion

CPAP is considered the first line treatment for OSA in adults [6]. The majority of children with OSA are successfully treated with adenoidectomy and tonsillectomy [8]; however, when these procedures are not recommended and other alternatives are not available, CPAP can also be used in children, particularly in children with craniofacial anomalies, obesity, and neuromuscular weakness [9]. Significant permanent face and teeth modifications caused by long-term use of CPAP in healthy adult patients are usually very rare and minor [10], unlike the frequent reports describing facial deformities in young children treated by CPAP [11,12].

The current patient had a relatively severe form of MG, which was the main reason for CPAP use. Eventually, this caused a major dentoskeletal, as well as soft tissue maxillary deformity. The upper lip was retruded with both skeletal and dental Class III, and reduced lower face height and protruded upper and lower incisors were presented. These were very disturbing to the patient in terms of esthetics and eating comfort.

The patient started using nasal CPAP at age 12, an age at which 90% of facial development is completed [13]. Therefore, the expected impact of the CPAP treatment on skeletal development is most likely minor. The deformity could also be attributed to the lack of sufficient muscle resistance due to his disease, which may have caused more direct and effective force on facial structures.

This malformation could also result from either a discontinuation of normal growth or be due to an active retrognathic force. Using a ventilator machine or CPAP in patients with muscle weakness and difficulty breathing during the night is extremely important and usually cannot be avoided. Furthermore, treatment may also continue during waking hours, as was the case in the current patient, who used CPAP for 12 h a day, which is far longer than average normal sleep time. The nasal mask had probably been utilized because it is somewhat less cumbersome than a full-face mask, and it probably contributed to his deformity by creating a force concentrated on a smaller area. No doubt several factors associated with the syndrome of MG and muscle weakness could have further influence on patient facial characteristics, including: patient's natural oro-motor function, mouth breathing, and low tongue posture, which contribute to posterior crossbite and lack of lip seal that may lead to a lack of nasomaxillary development [2]. These characteristics are familiar but not totally confirmed [3]. Also, the presence of OSA and mouth breathing may be the cause of several facial features of children regardless of CPAP use, even though this connection was not totally proven in means of chronological order or cause and effect relation [14–18]. The authors believe that these primary factors cannot be ignored and may contribute to facial deformity. A large-scale study including patients with muscle weakness, like MG, which aims to compare facial characteristics of CPAP users to non-CPAP users, is indicated to clear this point.

## Conclusion

A wise treatment approach combining orthodontics and CPAP engineering can lead to reduced facial disfigurement later in life. Thus, a mask with a forehead or chin support or a regular full face mask is recommended instead of a nasal mask for patients with muscle weakness in order to spread the orthopedic force more evenly and reduce maxillary deformity.

## Contributors

YH made substantial contribution to the study's conception, acquisition and interpretation of the data, drafted the submitted article to be published. SL, GA, and YS helped with the interpretation of the data and wrote the draft. UZ analyzed the orthodontic data and approved the draft.

## Disclosure statement

The authors deny any conflicts of interest.

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