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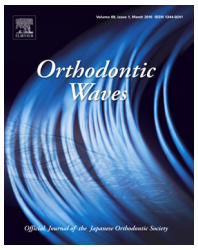

Short communication

Early dentofacial orthopedic treatment of a patient with maxillary hypoplasia and congenital central hypoventilation syndrome

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ABSTRACT

Congenital central hypoventilation syndrome (CCHS) is a respiratory disorder characterized by compromised central and peripheral chemoreflexes that lead to hypoventilation during sleep. All patients with CCHS require lifelong ventilatory support during sleep; however, nasal bilevel positive airway pressure (BiPAP) therapy can cause maxillary hypoplasia. In the present case, the patient had been diagnosed with CCHS and was initiated on BiPAP therapy at the age of two months. The patient presented to our orthodontic department with severe midface hypoplasia along with an anterior dental crossbite at the age of five years. An orthodontic appliance (facemask) was prescribed to assist forward growth of the nasomaxillary complex. The patient was instructed to wear the facemask together with the BiPAP appliance while sleeping and for as long as possible when awake. Orthopedic treatment for at least one year resulted in favorable forward growth of the maxilla and reduced facial concavity.

1. Introduction

Congenital central hypoventilation syndrome (CCHS), also called Ondine's curse, is a respiratory disorder that can be fatal if left untreated. CCHS is characterized by compromised central and peripheral chemoreflexes in the absence of primary neuromuscular, lung, cardiac, or metabolic diseases that lead to hypoventilation during sleep [1]. All patients with CCHS require lifelong ventilatory support during sleep [2,3]. One modality for home mechanical-assisted ventilation is bilevel positive airway pressure (BiPAP) therapy via a nasal mask. A counteracting force against the positive pressure exerted by BiPAP is necessary to maintain the mask in position and to

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prevent leaks, and previous studies have described midface hypoplasia as a complication of noninvasive ventilation in children [4,5].

The reported incidence of CCHS is approximately 1 in 200,000 live births [6]. Since the incidence of the CCHS is low, the orthodontic treatment of midface hypoplasia when it occurs as a complication of noninvasive ventilation in CCHS patients has yet to be standardized. Generally, maxillary hypoplasia can be orthopedically corrected with an orthopedic facemask [7,8]. In this report, we present a case of a child with CCHS who developed maxillary hypoplasia secondary to nasal BiPAP therapy. Her orthopedic treatment outcomes after wearing the facemask together with the BiPAP appliance are discussed.

2. Case report

The patient presented to our orthodontic and dentofacial orthopedic department with an anterior dental crossbite at the age of five years. She had a significant concave-type soft tissue facial profile (Fig. 1). Lateral cephalometric radiograph revealed a skeletal Class III due to maxillary hypoplasia and mandibular protrusion (Fig. 1 and Table 1), and an intraoral examination revealed an anterior dental crossbite. The overjet and overbite were −10.0 mm and +1.5 mm, respectively (Fig. 2).

The patient had been diagnosed with CCHS in infancy. Examinations included ultrasonography, magnetic resonance

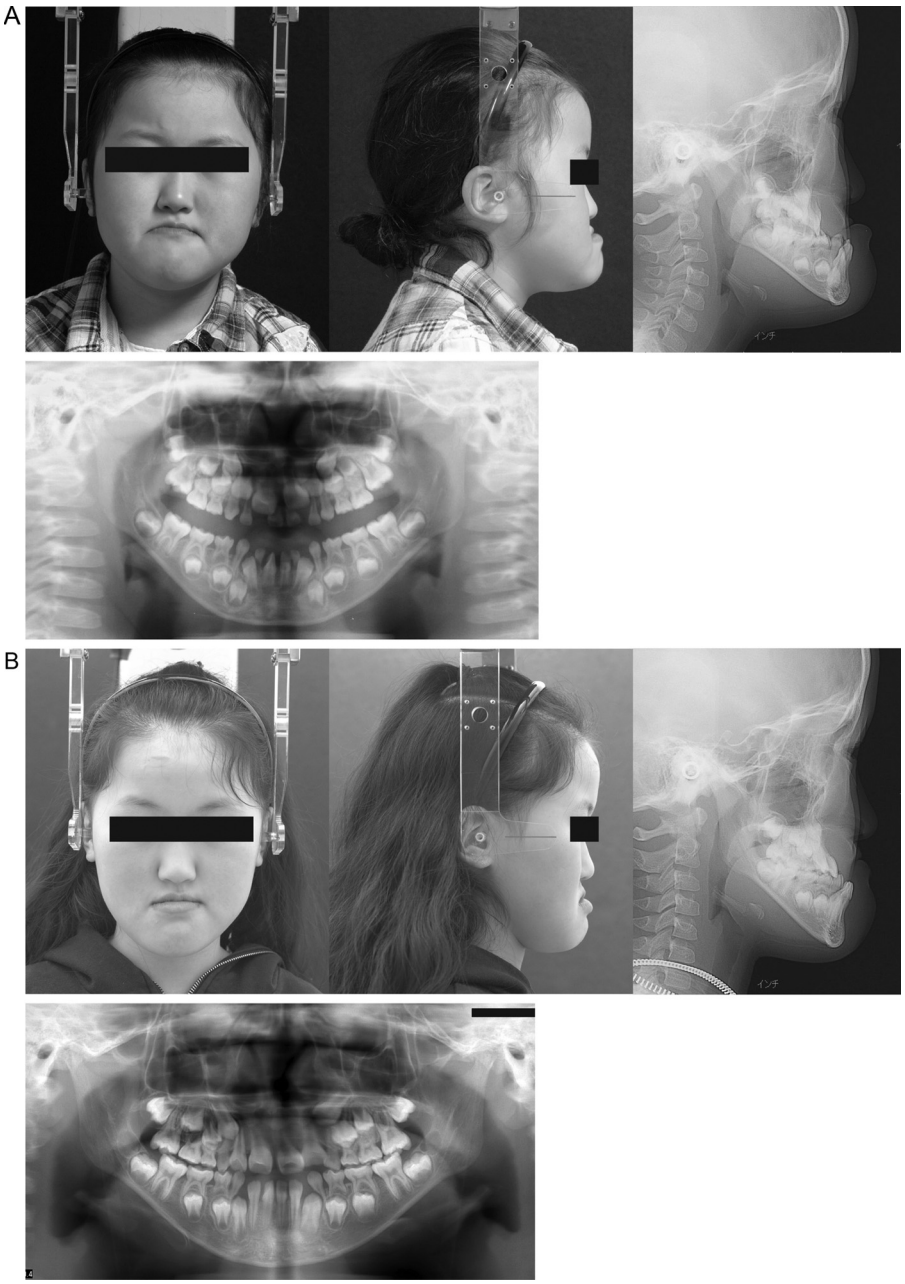

Fig. 1 – Facial photographs and radiographs: (A) pre-treatment and (B) after 1 year of orthodontic treatment.