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Case Report

Management of Obstructive Sleep Apnoea in a Patient with Pierre Robin Sequence by Using Distraction Osteogenesis

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***Abstract*—Miss A is a 23-year-old Malay female diagnosed with a Pierre Robin sequence (PRS) associated with severe obstructive sleep apnoea (OSA) and feeding problem because of a severe micrognathic mandible. She required a continuous positive airway pressure (CPAP) device to sleep at night. The mandible was successfully treated with distraction osteogenesis. Her OSA improved, and her feeding problem was alleviated. Thus, she no longer needs a CPAP device.**

***Keywords* - Pierre Robin Sequence; Distraction Osteogenesis; Obstructive Sleep Apnoea**

I. INTRODUCTION

The Pierre Robin sequence (PRS) is a description of a complex condition involving a cleft palate, micrognathia and airway obstruction [1]. It can occur without a cleft palate [2] and in combination with other syndromes, such as hemifacial microsomia and velocardiofacial syndrome [3]. Patients with PRS have two common problems: airway obstruction and feeding difficulties. Her treatment is divided into nonsurgical and surgical methods. Nonsurgical techniques include prone positioning, suction and drinking plates [4] and a nasopharyngeal airway [5]. Some surgical modalities have tracheostomy, orthognathic surgery and distraction osteogenesis [6]. Obstructive sleep apnoea (OSA) is due to the obstruction of the upper airway during sleep. OSA causes hypersomnolence, hypertension, cardiac arrhythmia and myocardial infarction. Distraction osteogenesis involves a bone formation in a stretched fractured callus bone. An appliance attached to the osteotomised fragments is pulled apart subsequently. This technique has shown acceptable efficacy, safety and feasibility [7].

II. CASE REPORT

A 24-year-old girl was referred from the Department of Otorhinolaryngology for mandibular advancement. Examination revealed a convex facial profile with micrognathic mandible, which caused limitation in mouth opening, thereby leading to feeding problems. Pretreatment extra and intra-oral photographs, orthopantomogram (OPG) and lateral cephalogram were investigated (Fig. 1). Since the age of 16 years, she experienced multiple episodes of breathing cessation during sleep, leading to frequent waking at night. A polysomnography test in 2007 confirmed severe OSA with an apnoea: hypoapnoea index (AHI) of 144. Thus,

she was placed on a continuous positive airway pressure (CPAP) device at night.

Two surgical approaches were chosen to address her three main problems: OSA, aesthetics and limited mouth opening. The first one was applied to correct her micrognathic mandible, and the subsequent orthognathic surgery was intended to improve her mastication and appearance. Her mandible was osteotomised under general anaesthesia at the retromolar area, and an intraoral distractor device was placed bilaterally. After 7 days (latency period), the distractor was activated twice daily to achieve a 1 mm advancement per day (Fig. 2). The total bone length was 12 mm. After the activation phase was completed, the distractors were maintained in place for 2 months for bone consolidation. The distractors were removed under general anaesthesia. A post-distraction polysomnogram test showed that the AHI was 41.1. Her AHI is still within the severe range, but she no longer depends on her CPAP to date. Her mouth opening also dramatically improved from 18 mm to 35 mm (Fig. 3). The patient was satisfied with the treatment outcome and thus refused to undergo orthognathic surgery, which was planned to improve her mastication and appearance.

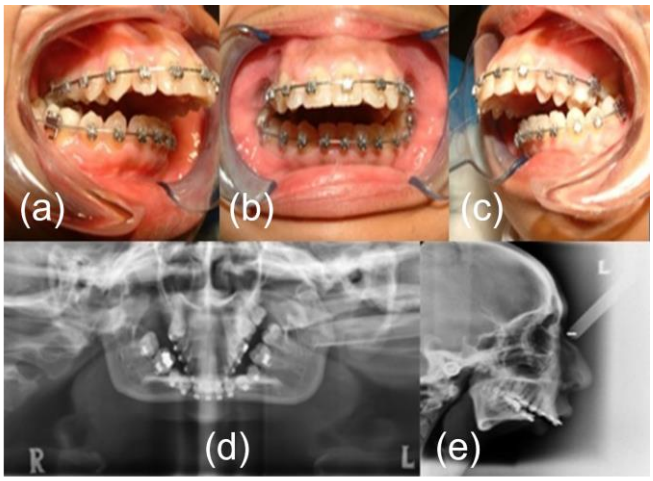

Fig. 1. Pre-treatment photograph of occlusion. a) Right lateral view. b) Anterior view. c) Left lateral view. d) Lateral cephalogram. e) OPG.

III. DISCUSSION

Micrognathia causes the tongue base to be displaced in the oral pharynx in a condition known as glossoptosis. In turn, glossoptosis causes severe airway obstruction and feeding difficulties. Infants with micrognathia may experience a decrease in weight gain and failure to thrive because of feeding difficulty. The management of micrognathia ranges from conservative approaches (such as prone positioning) to aggressive counterparts (such as head braces, nasopharyngeal intubation, glossopexy, tracheostomy and distraction osteogenesis) [8]. These patients likely grow normally later. Contrary to this notion, Pruzansky S [9] reported no significant catch-up growth.

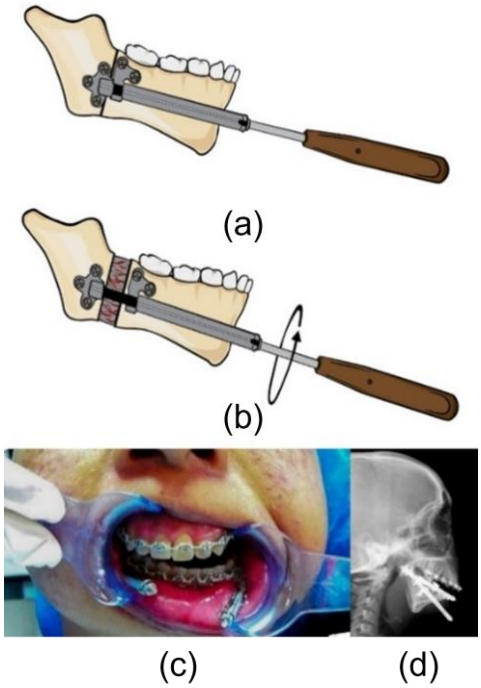

Fig. 2 a) Placement of the distractor on the osteotomised mandible. b) Formation of a callus following distraction activation. One revolution led to 0.5 mm advancement. c) Intraoral activation port position d) Position of the distractors on the patient's mandible (lateral cephalometric radiograph).

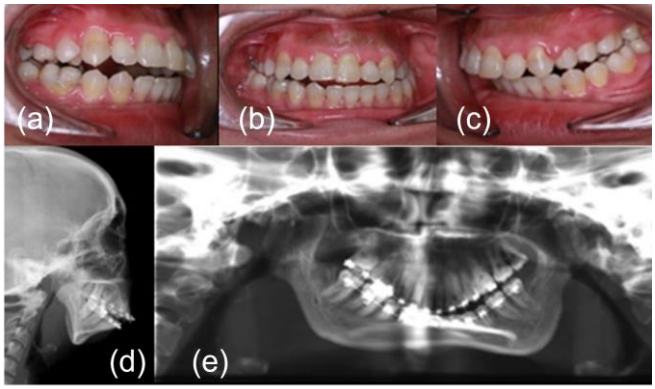

Fig. 3 Post-treatment photograph of occlusion. a) Right lateral view. b) Anterior view. c) Left lateral view. d) Lateral cephalogram. e) OPG

Several authors reported only partial catch-up growth later [12,13]. This outcome is in accordance with our patient's convex profile prior to the commencement of the treatment. Airway obstruction occurs when pharyngeal muscles collapse and block the upper airway during sleep. This situation leads to the partial (hypopneas) or complete stop (apnoea) of breathing that lasts between 10 s or longer. Blood oxygen saturation decreases up to 40 percent or more in severe cases. Patients usually increase their negative intrathoracic pressure to overcome their collapsed airways. This occurrence leads to sleep fragmentation and hypersomnolence.

The severity of OSA must be established to make an appropriate treatment decision. The most common method involves laboratory polysomnography. Patients with OSA should be comprehensively evaluated, including the assessment of sleepiness severity by using an Epworth Sleepiness Scale [14]. OSA management may include a CPAP device, oral appliances, surgery, behavioural changes, over-the-counter remedies and positional therapy. The use of CPAP devices is the standard treatment option for mild, moderate and severe OSA.

Our patient has been using a CPAP device since 1998. The device keeps her airway open, thereby preventing breath pauses and causing the restoration of normal oxygen levels. Persons with severe anatomic deformity may benefit from surgery. This surgery may be as simple as soft tissue removal from intraoral structures (e.g. the soft palate, uvula, tonsils or adenoid) or as complex as craniofacial bone adjustments (e.g. orthognathic surgery or distraction ontogenesis). Her CT scan revealed that our patient had a very small mandible, which made conventional orthognathic surgery almost impossible. Thus, we decided to correct her severe hypoplastic mandible through distraction osteogenesis.

Distraction osteogenesis offers less tissue manipulation, minimal blood loss and lower risk of relapse compared with those of orthognathic surgery. Airway obstruction is relieved when the tongue base is carried anteriorly via its muscular attachment to the distracted mandible [15]. In distraction osteogenesis, a screw-driven device is attached to the newly osteotomised bone fragments. As these fragments are pulled apart, a new bone is generated in the newly formed stretched callus. Then, 3 years after mandibular lengthening by distraction osteogenesis, the AHI of our patient improved from 133 preoperatively to 41.1.

Our patient is no longer using a CPAP device at night and has stopped experiencing daytime hypersomnolence. Her weight dramatically increased from 40 kg to 65 kg post-distraction.

IV. CONCLUSION

The management of obstructive sleep apnoea remains a challenging task for clinicians. In comparison with mandibular lengthening in infants, distraction osteogenesis in adults has not gained significant acceptance. This situation may be due to the extended treatment time relative to conventional orthognathic surgery. Our patient showed an improvement in her respiratory and feeding problem after distraction osteogenesis.

Therefore, distraction osteogenesis is a proven treatment method for improving obstructive sleep apnoea because of micrognathia.

CONSENT TO PARTICIPATE

Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

CONFLICT OF INTERESTS

The authors declare that there is no conflict of interest.

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