



Contents lists available at ScienceDirect

Journal of Oral and Maxillofacial Surgery, Medicine, and Pathology

journal homepage: www.elsevier.com/locate/jomsmmp

Case report

Efficacy and optimal timing of tongue reduction surgery in three patients with Beckwith–Wiedemann syndrome

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ARTICLE INFO

Article history:

Received 21 November 2016

Received in revised form 20 February 2017

Accepted 22 March 2017

Available online 20 April 2017

Keywords:

Beckwith–Wiedemann syndrome

Macroglossia

Tongue reduction

ABSTRACT

Beckwith–Wiedemann syndrome (BWS) is a rare congenital overgrowth disorder. The most common manifestations are exomphalos, macroglossia, and gigantism. Macroglossia has possibility of multiple finding such as functional, cosmetic or psychological issues with the patient and their families. The treatment for macroglossia is tongue reduction surgery in general. Besides airway or feeding problem, the indication and of tongue reduction surgery is controversial. The necessity of surgery depends on each patient with multiple clinical finding by macroglossia, and the age of patients has to be considered for risk management of surgery. We report three cases with BWS who were considered efficacy and optimal timing of tongue reduction surgery and had positive outcome. It is concluded that the capable timing for tongue surgery to mitigate macroglossia should be between 2 to 3 years of age.

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1. Introduction

Beckwith–Wiedemann syndrome (BWS) is an overgrowth disorder, initially described by Beckwith in 1963 and Wiedemann in 1964, and has an estimated incidence of 1 in 12,000 to 1 in 13,700 live births [1–3]. The most common manifestations of BWS are exomphalos, macroglossia, and gigantism [4]. Therefore, it has also been referred to as EMG syndrome. Other features commonly found include distinct facial characteristics, such as ear lobe creases, posterior helical ear pits, facial nares flares, neonatal hypoglycaemia, organomegaly, renal abnormalities and hemihypertrophy [1,5]. In BWS 80–99% of these patients have macroglossia, which is defined as a resting tongue that protrudes beyond the teeth or alveolar ridge [6–8]. Depending upon the degree of severity, macroglossia can lead to complications involving feeding and respiration in infancy. Later, macroglossia can impede speech articulation and lead to malocclusion as the growth of the mandible is guided, at least in part, by the size of the tongue. The purpose of this report is to present the authors' series of BWS patients who have undergone surgical

reduction of the tongue with consideration of efficacy and optimal timing for surgery.

2. Case report

2.1. Case 1

The patient was diagnosed clinically with BWS at birth. He was re-evaluated for an umbilical hernia by a paediatrician and was referred to the oral and maxillofacial department in Kameda medical Centre for an opinion regarding macroglossia related to BWS. His parents reported that the child was not able to close his mouth and that he had adequate sleep only in a lateral or sitting position due to sleep-disorder breathing with heavy snoring (Fig. 1). Other associated factors included mandibular protraction, sialorrhea, phonation difficulties, and protruded tongue (Figs. 2 and 3). His occlusion consisted of an open bite with the mandibular anterior teeth at a labial inclination.

2.2. Case 2

The patient was diagnosed clinically with BWS during the thirty-third week of embryonic life. She had done an umbilicoplasty and was referred to our department. She displayed mandibular protraction (Fig. 4) and tongue protrusion over the teeth arch resulting in an inability to fully occlude and phonation difficulties. The patient's anterior dentition had a labial splay and the mandibular arch was

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Fig. 1. The aspect of supine position at sleeping.



Fig. 3. Preoperative picture showing relationship between tongue and dentition (A: front view, B: side view).



Fig. 2. Preoperative picture showing macroglossia (A: rest position, B: protrusive position).

wider than the maxillary arch, resulting in a posterior bilateral cross-bite.

2.3. Case 3

The patient had an excessively long umbilical cord, macroglossia, gigantism, and was diagnosed BWS at birth. The department of pediatric surgery referred him to the NICU to manage his air-

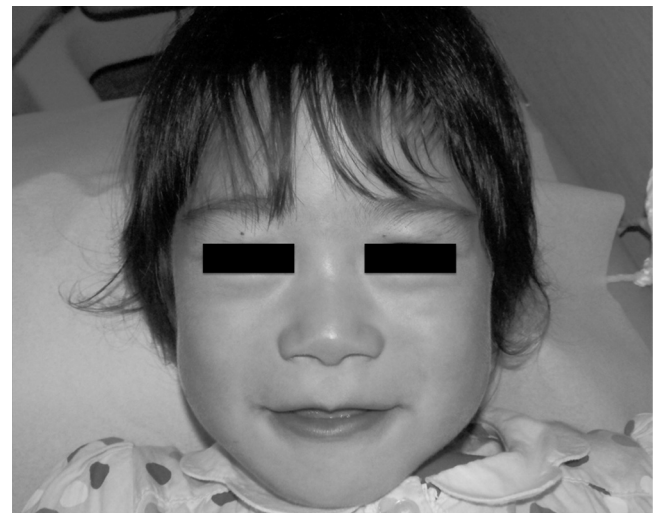


Fig. 4. Preoperative picture showing mandibular protraction.

way due to the macroglossia, which caused an airway obstruction. Other associated factors included mandibular protraction, sialorrhea, phonation difficulties, and protruded tongue. His occlusion consisted of an open bite (Fig. 5).

Case 1 underwent a tongue reduction at two years of age. Case 2 and 3 underwent a tongue reduction at two years and ten months of age, respectively. For the surgical procedure, a keyhole type reduction of the tongue was performed to enable a satisfactory position within the oral cavity at rest under general anaesthesia. All the cases

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