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Partial glossectomy in a patient carrier of Beckwith–Wiedemann syndrome: Presentation of a case☆

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ABSTRACT

The Beckwith–Wiedemann syndrome is a congenital disorder with diverse clinical manifestations, among which the most prevalent are, macroglossia (97%), gigantism (88%), and abdominal wall defects (80%). Orthodontically, most patients present with anterior open bite and Angle Class III malocclusion. Macroglossia can cause cosmetic problems and functional abnormalities associated with speech, mastication, swallowing and breathing, with potential obstruction of the upper airways and decreased stability of orthodontal-surgical treatment. In order to avoid episodes like this, a partial glossectomy is necessary in some patients. This article looks at the diagnosis and treatment of macroglossia, while presenting the case of a patient with Beckwith–Wiedemann syndrome who underwent surgery by partial glossectomy using the technique advocated by Obwergeser et al. in 1964, and 3 years postoperatively showed good cosmetic and functional results.

Glosectomía parcial en paciente portador del síndrome de Beckwith-Wiedemann: relato del caso

Palabras clave:
Glosectomía
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Síndrome de Beckwith-Wiedemann

RESUMEN

El síndrome de Beckwith-Wiedemann es una alteración congénita con diversas manifestaciones clínicas, de entre las cuales las más prevalentes son la macroglosia (97%), el gigantismo (88%) y los defectos de la pared abdominal (80%). Ortodónticamente, la mayoría de los pacientes presentan mordida abierta anterior y relación de clase III de Angle. La macroglosia puede causar problemas estéticos y anomalías funcionales relacionadas con el habla, la masticación, fonación, deglución y respiración, con potencial de obstrucción de las vías respiratorias superiores y disminución de la estabilidad del tratamiento ortoquirúrgico.

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Con el fin de evitar episodios como este, es necesaria la realización de una glosectomía parcial en algunos pacientes. El presente trabajo realiza consideraciones con relación al diagnóstico y tratamiento de la macroglosia y relata el caso clínico de un paciente portador del síndrome de Beckwith-Wiedemann que fue intervenido por medio de glosectomía parcial, utilizando la técnica preconizada por Obwergeser et al. (1964) y que en un postoperatorio de 3 años presentó resultados cosméticos y funcionales satisfactorios.

Introduction

The *Beckwith–Wiedemann* syndrome, first described by Beckwith[1] and Wiedemann,[2] is a rare congenital disease that involves an overgrowth disorder and affects one in 14,200 newborns.[3] Its origin may be hereditary (15%) or originate in genetic alterations in the chromosome 11p15.5 region (75–80%).[4]

The main characteristics of the Beckwith–Wiedemann syndrome are: macroglossia (97%), gigantism (88%) and defects of the abdominal wall such as omphalocele, umbilical hernia and diastasis recti (80%), followed by abnormal creases in the ear lobes (76%), hypoglycaemia (63%), facial nevus flammeus (62%), kidney abnormalities (59%), haemihypertrophy (24%), congenital heart defects (6.5%), intestinal problems (5%), neoplasia (4%), mental retardation (4%), polydactyly (3%) and labial-palatal fissure (2.5%).[3]

Macroglossia is defined as a tongue that in the resting position protrudes beyond the teeth towards the alveolar ridge. It has a multifactorial aetiology and it is classified in true and relative macroglossia. The relative macroglossia occurs when the tongue has a normal size, but the intraoral space is insufficient and the true one is observed due to an increase in its volume.[5]

Among the characteristics of the *Beckwith–Wiedemann* syndrome, macroglossia represents 97% of the incidence, and this condition may cause a functional and aesthetic abnormality with speech, mastication, deglutition and suction difficulty.[6] There is also the risk of upper airways obstruction during childhood and adolescence, of development of obstructive sleep apnoea syndrome and even of decrease in the stability of the orthosurgical treatment.[3,7,8]

As for the maxillofacial morphology, some specific findings may be found in patients carrying the *Beckwith–Wiedemann* syndrome, such as bimaxillary protrusion, mandibular protrusion, goniac angle and increased mandibular body. Orthodontically, most patients present anterior open bite, negative *overjet*, Angle Class III relationship and vestibular inclination of the anterior elements. These characteristics depend on the patient's degree of macroglossia.[9]

Many of those patients present important functional problems or may be candidate for orthognathic surgery and they need partial glossectomy for the oral functions dependent on the tongue to be restored and to obtain improvement of the foresee ability of the bone movements, especially all those that involve the jaw.

Case description

Patient receiving H.V.P.D., 16 years, presented with main complaint of phonation difficulty. During the physical examination, patient showed elevated height, facial pattern III, with severe maxillary hypoplasia, dental crowding, Angle Class III relationship, anterior open bite, bifid uvula and macroglossia (Fig. 1A–C). At the data collection during anamnesis, it was also revealed that, at birth, patient had an abdominal wall defect that was later corrected. All these findings contributed to the formation of the *Beckwith–Wiedemann* syndrome diagnosis.

Patient was included in the orthosurgical treatment, and deglutition and phonation problems and respiratory difficulties, with a strong correlation with macroglossia, were observed. Therefore, for this case, a surgical procedure of partial glossectomy was scheduled, before the orthognathic surgery procedure. The technique used was the one proposed by Obwegeser et al.,[10] in which a resection of the central segment of the tongue and the tongue apex was performed, which achieved a decrease in its dimensions in the anteroposterior and transversal sense, to lessen the speech-related problems (Fig. 1D–H).

For appropriate transoperatory bleeding control and ease of execution, the surgery was performed under general anaesthesia with orotracheal intubation, due to the presence of nasal concha hypertrophy that prevented the passing of the nasotracheal tube. To perform the incisions, anatomical landmarks were established in the lateral and anterior extremities of the tongue with 2.0 cotton thread and an *Allis* tissue grasping clamp was used as auxiliary in the dissection. After stabilisation, incision demarcation with blue methylene and also infiltrations of local anaesthetic (lidocaine 2% with epinephrine 1:200,000) were performed. The removal of the demarcated segment was performed with an electrosurgical dissector (Colorado needle, Stryker Corporation®) in the apex-base direction, preserving the lateral edges and minimising the risk of lesions in the lingual nerve branches. The segment was removed and careful haemostasis was performed to minimise the risks of lingual haematoma and postoperative bleeding. After bleeding control, the segments were thoroughly approximated with resorbable thread (Vicryl 3.0-ETHICON) from the deepest to the superficial portion in the base-apex direction. In the immediate postoperative period, it was already possible to verify the anterior and transversal decrease of the tongue without airway involvement, and without any complication of haemorrhagic or infectious nature. Ten days after surgery, patient started phonoaudiologic therapy to assist in the recovery of tongue mobility and phonation.