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Taste and speech following surgical tongue reduction in children with Beckwith–Wiedemann syndrome

Saskia M. Maas [a, b, 1], Daniel J. Kadouch [c, 1, 2], Anne-Claire C.M. Masselink [d, 3], Chantal M.A.M. Van Der Horst [c, *]

[a] Academic Medical Center, Department of Pediatrics, Amsterdam, The Netherlands
[b] Academic Medical Center, Department of Clinical Genetics, Amsterdam, The Netherlands
[c] Academic Medical Center, Department of Plastic Reconstructive and Hand Surgery, Amsterdam, The Netherlands
[d] Academic Medical Center, Department of Audiology, Amsterdam, The Netherlands

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ABSTRACT

Beckwith–Wiedemann syndrome (BWS) is an overgrowth disorder in which macroglossia is one of the main signs. We investigated the long-term outcome of tongue surgery reduction (TRS) on taste and speech in patients with BWS who were more than 5 years of age and had undergone surgical anterior wedge resection of the tongue. A questionnaire was used to assess medical history and to determine some aspects of speech, taste, psychological well-being, and degree of satisfaction with regard to TRS and tongue mobility. Speech sound error pattern and degree of intelligibility were measured by a speech therapist, and taste was assessed using a validated test.

The degree of both intelligibility and satisfaction with the surgery was high. There were some speech errors; especially the interdental 's', addental 't', and addental 'd' were more noticed. We conclude that anterior wedge resection is an effective technique to treat macroglossia in children with BWS, and that it has no long-term consequences for intelligibility and taste perception and only limited consequences for speech.

1. Introduction

Beckwith–Wiedemann syndrome (BWS) is a well-known overgrowth disorder (Beckwith, 1963; Wiedemann, 1964) with an estimated incidence of 1 in 12,000 to 1 in 13,700 live births (Engstrom et al., 1988; Wiedemann, 1997; Weksberg et al., 2001; Cohen, 2005). The most characteristic features are prenatal or postnatal overgrowth, anterior wall defects, and macroglossia. Other common symptoms are neonatal hypoglycemia, organomegaly, facial naevus flammeus, ear creases or helical ear pits, and hemihypertrophy. The clinical presentation is very variable, and diagnosis is made using criteria by either Elliott et al. (1994), de Baun and Tucker (1998). BWS shows etiologic heterogeneity explained by abnormal methylation of one or both imprinted growth regulatory genes *H19* and *LIT1* on chromosome 11p15 (Weksberg et al., 2001). Affected individuals have an increased risk of developing embryonal neoplasms such as Wilms tumor or hepatoblastoma (Bliek et al., 2001; Cooper et al., 2005; Rump et al., 2005; Brioude et al., 2013).

The major sign of BWS is macroglossia, which is seen in up to 94% of patients. As a consequence of the large tongue, various oral functions may be affected, including breathing, swallowing, chewing, and speech. Moreover, macroglossia has esthetic implications (open mouth appearance, widened interdental spaces, mandibular prognatism) that may cause marked negative consequences in regard to body image and psychological well-being, and a reduced quality of life (McManamy and Barnett, 1985; Elliott et al., 1994; Engel et al., 2000; Van Lierde et al., 2010; Brioude et al., 2013).

Tongue reduction surgery (TRS) aims to reduce the size of the tongue, while maintaining normal shape and function. Ideally, the reduction should result in a tongue that remains behind the lower

* Corresponding author. Department of Plastic, Reconstructive and Hand Surgery, Academic Medical Center, P.O. Box 22660, 1100 DD, Amsterdam, The Netherlands. Tel.: +31 20 5662974; fax: +31 20 6917549.
E-mail address: c.m.vanderhorst@amc.uva.nl (C.M.A.M. Van Der Horst).
[1] These authors contributed equally to this work.
[2] Present address: Academic Medical Center, Amsterdam, Department of Dermatology, The Netherlands.
[3] Present address: University Medical Center, Utrecht, Department of plastic surgery, The Netherlands.

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dental arch, yet can wet the lips on protrusion (Heggie et al., 2013). TRS should also improve speech intelligibility, articulation, and growth of the jaw (Van Lierde et al., 2010). Our own clinical experience (Kadouch et al., 2012) indicated that the anterior wedge procedure is the most effective technique to treat macroglossia in BWS. Long-term functional outcome studies of TRS procedures are sparse and show various results (Kadouch et al., 2012), while patients and parents have expressed concerns about the long-term outcomes, especially with respect to taste (Niki et al., 2000; Matsune et al., 2006).

The aim of the present multidisciplinary study is to evaluate long-term outcomes with regard to taste and speech in BWS patients after surgical tongue reduction for their macroglossia.

2. Material and methods

The present study is a retrospective observational clinical follow-up to assess long-term outcomes after TRS. From July 2011, all 18 patients with BWS (older than 5 years of age) who had undergone surgical tongue reduction at the Academic Medical Center in Amsterdam were invited to participate to the study. We obtained written informed consent from all study participants and/or their parents/caregivers. The study was approved by The Medical Ethical Committee of the AMC, University of Amsterdam (METC 2011_018#C201124.

2.1. Study population

All patients fulfilled the criteria by either Elliott et al. (1994), de Baun et al. (1998). The clinical diagnosis was molecularly confirmed in all study participants except two. BWS patients who underwent an anterior wedge resection of the tongue at our institution between 1990 and 2009 were allowed to participate in the study. Patients were excluded if they had a serious mental impairment, a history or presence of a medical condition that may influence speech (such as cleft palate and hearing problems), and/or a history or presence of a medical condition that might influence taste perception.

2.2. Study outcomes

A questionnaire was used to assess medical history, as well as historical aspects of speech, taste and psychological well-being. To evaluate the degree of satisfaction with regard to TRS, both the patients and/or parents and plastic surgeon. (C.M.A.M. van der Horst) were asked to give a score on a scale from 1 (not satisfied at all) to 10 (extremely satisfied) on a questionnaire.

Participants underwent the following tests that were performed in the same order:

2.2.1. Taste test

Taste assessment was performed by a validated test developed by our institute. Sweet, salt, sour, and bitter solutions were tested. After making small circular movements on the tongue for 1–2 s, the patient was asked what taste was perceived on the region tested (Van der Horst et al., 2010). Differences in outcomes of the taste test between the study and case control group were described by differences in (correct) taste perception (yes = 1; no = 0).

2.2.2. Speech assessment

Differences in outcomes of speech between the study and case control group were evaluated and scored by the same experienced speech pathologist (A.C. Masselink.). Sampling took place in a sound-protected room and was videotaped. Each patient's oral motor, speech, language, and swallowing proficiencies were assessed. Swallowing motions were assessed by observing swallowing saliva and consumption of water. Tongue mobility was assessed by observing each subject's ability to maneuver the tongue tip within and outside the oral cavity (touch nose, chin, lick lips, lick teeth, move from right to left side of the mouth). The speech sound error pattern was assessed with a standardized articulation test for words and sentences, spontaneous speech, and speech in conversation. Speech errors that were scored were the interdental and addental S, T, and D. The degree of intelligibility was tested on a scale from 1 to 5 according to the NVSCA standard as described (Dutch Society of Clefts and Craniofacial abnormalities).

3. Results

3.1. Study population

Demographic, genetic, and BWS-related health data are described in Table 1. In total, 18 patients with macroglossia as part of BWS underwent TRS at our institution between 1990 and 2009. Ten of them fulfilled the selection criteria and were willing to participate.

Almost all subjects kept their tongue a substantial time out of their mouth (75–100%) before TRS surgery and only two after surgery. Tongue protrusion was reported as normal. Eight of the 10 patients could wet their lips on protrusion of the tongue, but of those, five only barely could. Tongue mobility tests and satisfaction with regard to TRS and speech development are summarized in Table 2. All parents would opt for surgery again. Three children were old enough to answer the question about satisfaction after the TRS; two gave the surgery a 10 and one a 7 (on a scale of 1–10).

3.2. Study outcomes

3.2.1. Taste test

Two patients still had abnormal eating or drinking habits after surgery, such as drinking at an angle from a cup or ability to eat only small pieces. No children mentioned having tasting problems. The outcomes of the taste test are listed in Table 3.

3.2.2. Speech assessment

The score of degree of intelligibility is described in Table 2. Normal speech was seen in three children who underwent operation children, and in 6 children speech was judged to be different from other children but intelligible and not leading to comments. Only in one instance was speech really different from that of other children, leading to comment. Speech errors were especially the interdental ‘s’ and addental ‘t’ and addental ‘d’.

4. Discussion

The present results indicate that the anterior wedge resection is an effective treatment for macroglossia in children with BWS, has no major long-term sequelae for speech and taste, and that patients and parents experience a high degree of satisfaction with results, which is mirrored by the opinion of their treating plastic surgeon.

The impact of partial glossectomy on overall speech intelligibility or articulation in children with BWS has been studied by several authors. van Lierde et al. published a comprehensive review of these articles (Van Lierde et al., 2010). Most reports mention an improvement of intelligibility and articulation of phonetic placement of the tongue, but are difficult to compare because of different assessment methods and different surgical techniques. However, Tomlinson et al. reported that patients are unlikely to have