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

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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

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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

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Table 1
Patient demographic and clinical data regarding Beckwith–Wiedemann syndrome (BWS)–related health (N = 10).

Characteristics	Patients
Gender	
Male/female	3/7
Genetic subtype of BWS	
Aberrant imprinting Lit1	5
Aberrant imprinting of H19	2
UPD	2
No aberration found	1
Age at time of operation	9 months 4.5 years (mean 21 months)
Mean age at evaluation (range)	9.7 years (5.6–17.9 years)
Breathing problems at birth	6
Having frequent colds	6
Removal of tonsils	6
Snoring	5 (all less after tongue reduction surgery)
Drooling	Preoperatively 8, postoperatively 2
Tongue hanging out of mouth	Preoperatively 7, postoperatively 2
Percentage of time hanging out of mouth	9: 75–100% (preoperatively) 1: 50–75% (preoperatively)

Table 2
Reported clinical outcome regarding tongue mobility and speech (N = 10).

Characteristics	Patients
Surgery	
Should you do the surgery again?	10
Satisfaction parents[a]	8 (n = 1); 9 (n = 3), 10 (n = 5), NA (n = 1)
Satisfaction plastic surgeon[a]	7 (n = 2), 8 (n = 4), 9 (n = 4)
Satisfied about mobility tongue	6 (after surgery)
Can bite an apple	8
Normal protrusion tongue	9
Can put the lips together	10
Can lick the lips with the tongue	8
Can lick the teeth with the tongue	9
Can move the tongue to tip of nose	6
Can move the tongue to chin	10
Can move the tongue to left corner of mouth	8
Can move the tongue to right corner of mouth	10
Is able to click with tongue	8
Satisfied about speech development	9
Speech therapy for macroglossia	8
Length of speech therapy	6: <3 years 1: 3–6 years 1: >6 years
Phonemic analysis	
Interdental 's'	5
Addental 's'	1
Interdental 't'	3
Addental 't'	4
Interdental 'd'	1
Addental 'd'	6
Degree of intelligibility[b]	3 times: 1 6 times: 2 1 time: 3

1 = Speech is normal and intelligible.
2 = Speech is different from other children but there are no comments and speech is intelligible.
3 = Speech is different form other children and there are comments but speech is intelligible.
4 = Speech is only intelligible with effort.
5 = Speech is not intelligible.
[a] Score satisfaction on surgery: 0–10 (0 = completely unsatisfied, 10 = completely satisfied), NA = no answer.
[b] Range of speech intelligibility.

completely normal tongue function and appearance as adults (Tomlinson et al., 2007).

In our study, we evaluated tongue mobility, overall intelligibility, articulation, and taste function in 10 children with BWS who underwent TRS. We found that clinical symptoms related to the macroglossia, such as persistent drooling, breathing problems, and disturbed speech, diminished after surgery. Most of the children had good mobility of the tongue, for example, while biting an apple or moving the tongue to the nose, chin, and corners. Most were content about their tongue mobility. All children had speech therapy, and mostly for the same period of time. All patients would undergo surgery again and were very satisfied with the result; the same was true for parents and the plastic surgeon.

4.1. Taste

Parents of patients in whom a tongue reduction is indicated do not have to worry whether TRS will influence the ability to taste

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Table 3
Number of children who gave the right answer in taste test (N = 10).

		Taste			
		Sweet	Salt	Acid	Bitter
Tip of tongue	TRS	8	7	5	5
Right side	TRS	7	8	5	6
Left side	TRS	9	9	6	6
Back of the tongue	TRS	9	8	5	5

TRS = tongue reduction surgery (N = 10). All children expressed taste on every spot of the tongue, but not all could not name the taste adequately. The same results were seen in macroglossia children without tongue reduction (unpublished data).

well. Taste is derived from the interaction of paired cranial nerves (facial, glossopharyngeal, and vagal) (Moore, 1992). The tongue and palate are both involved in taste. As illustrated by Davalbhakta and Lamberty, traditional teaching of taste bud distribution involves tongue mapping with a predominance of sweet sensing buds concentration in the anterior region (Davalbhakta and Lamberty, 2000). The tip of the tongue had been identified as the most sensitive part for taste (Kruchinsky, 2006). Later it was reported that these previously accepted tongue maps are incorrect, and that all four tastes are perceived on all loci with taste receptors (Linden, 1993). Breslin stated that the receptor expression zones are heavily overlapping in most regions of the mouth (Breslin, 2013).

There have been only a few reports that have assessed sensory changes after TRS. Matsune et al. tested taste in four girls with BWS after TRS, and showed that taste thresholds for salty and bitter tastes in the central region and for salty taste on the left side were significantly higher than the values for normal Japanese children. The authors did not report the method that they used for the tongue surgery (Matsune et al., 2006). Niki et al. reported that tastes for acidity and bitterness were in the central region of the tip of the tongue in a patient with BWS and declined after reduction of the tongue (Niki et al., 2000). Both studies were very small and difficult to compare. In a recent study by Tomita et al., 24 patients who underwent a total or subtotal glossectomy and surgical reconstruction were evaluated for residual taste sensation with graded filter paper test for all four tastes (sweet, salt, sour, and bitter). Patients with more than half the tongue base remaining had good taste sensation, whereas those with less than one-third residual tissue had impaired taste function. These patients have more tongue tissue removed than TRS patients (Tomita et al., 2014).

In our study, none of the parents of patients complained of problems with tasting. Kadouch et al. have demonstrated that the anterior wedge technique is a simple, effective, and safe technique for the surgical treatment of BWS patients with macroglossia (Kadouch et al., 2012). The results of the taste test in our study group showed that taste perception was not disturbed in children with BWS who had undergone TRS.

4.2. Speech

Like other studies, speech intelligibility and articulation sounded good, and almost all subjects were content with their speech. Speech outcome after tongue surgery may affect sounds made primarily by the tip of the tongue against the upper or lower teeth or the anterior palate. Subjects seem to benefit from tongue reduction in mobility and functionality of the tongue in speech, chewing, and control of motor function of the mouth. These results are comparable to the results described by Van Borsel et al. (1999), Shipster et al. (2006). Another study has investigated the pressure of the tongue on the teeth, before and after surgery, and found no clinically significant differences of tongue pressure after TRS (Fröhlich et al., 1992). In our group, interdental S, addental T and addental D speech errors were noticed.

Preoperatively, articulation errors commonly occur for bilabial sounds (e.g.,/p//b/) that are often produced as linguolabials. Other lingual sounds commonly affected include alveolar plosives (e.g.,/t//d/) and alveolar fricatives (e.g.,/s/z/) (Shipster et al., 2006). Commonly, alveolar sounds may be produced as linguadental sounds (e.g., the tongue tip protrudes through the teeth), or the tongue blade may contact with the alveolar ridge instead of the tongue tip. This result in incorrect articulatory placement of the tongue, and will affect speech intelligibility. The macroglossia disrupts the relationship between the anterior articulators, altering the place but not the manner of speech production (Shipster et al., 2006).

We agree with Chau et al., who concluded that anterior tongue reduction is a reliable and helpful procedure in cases in which macroglossia adversely affected speech (Chau et al., 2011). In a paper by Heggie et al., seven BWS patients were investigated after a stellate/anterior wedge reduction, and all patients were satisfied with the results except for two minor contour deformities (Heggie et al., 2013). Only from Tomlinson et al. was a detailed description available of the phonetic realization of articulation sounds in four adults with BWS. They reported a high prevalence of self-reported lisps, anterior production of sounds, and dental or interdental placement errors, with no significant subjective or objective impact on daily life, which was also seen in our study (Tomlinson et al., 2007). Van Lierde et al. provided a review of the literature regarding speech intelligibility and articulation before and after glossectomy in children with BWS with the keyhole technique (Van Lierde et al., 2010). Our results are comparable with data from this review.

van Lierde et al. studied four children with BWS who underwent glossectomy; in all four children, speech intelligibility, articulation, resonance, and oromyofunctional behavior remarkably improved (Van Lierde et al., 2012). Mixter et al. described a child with BWS who had difficulty elevating the tongue tip to produce the/t/d/and/l/sounds as well as positioning the tongue blade for the/s//z//sh/and/zh/sounds in the presurgical condition. After surgery, speech was unchanged, but the patient was more easily able to evaluate the tongue within the oral cavity (Mixter et al., 1993).

4.3. Study limitations

A major limitation of the present study is the small sample size and lack of phonological data in children with BWS before and after partial tongue surgery. The retrospective nature may cause a bias in some of the studied items.

4.4. Future research

The present study offers additional insight into the long-term effects after tongue reduction in patients with BWS. Functionality of the tongue is essential for taste and communication—key elements of daily life—and also may cause major impairments in physical appearance and psychological well-being, indicating the importance of studying these items. We advocate more extensive studies on consequences for taste and speech over a more prolonged period of time, preferably performed in a prospective manner. Likely the desired size of such studies will dictate the studies to be international studies.

5. Conclusion

We conclude that anterior wedge resection seems to be an effective technique to treat macroglossia in children with BWS without long-term complications for speech or taste perception.

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