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**LINGUAL REDUCTION IN BECKWITH-WIEDEMANN SYNDROME**

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**Abstract**

*Background and Purpose*:Management of the macroglossia of Beckwith-Wiedemann syndrome (BWS) is controversial. 432 individuals with BWS, 74% of whom had lingual reduction by one surgeon with one technique (n=319), were assessed to define dentoskeletal morphology, aerodigestive function and surgical morbidity.

*Methods*:The study population was evaluated either through a craniofacial team (n=371) or as part of a specific BWS research program (n=61) between 1985 and 2013. The standard protocol focused on perinatal history, airway stability, communication skills, dentoskeletal development and the performance of and outcome of lingual reduction surgery.

*Results*: Initial lingual reduction was performed before 13 months of age in 47%, between 13 and 24 months of age in 30% and over 24 months of age in 23%. 21 patients had multiple tongue reductions: 17 prior to being operated by the reporting surgeon. 319 initial reductions were performed by one senior surgeon using the "W" anterior technique. Perioperative morbidity was: upper airway compromise in 2%, lingual dysfunction in 0%, dehiscence requiring surgical revision in 2%. All "W" reduction patients returned to preoperative oral and general behavioral function, often with improvement in eating and saliva control, within 6 weeks postoperatively. Speech therapy was provided or recommended for 53% of all BWS individuals, of whom 1/2 had lingual reduction. Of the "W" reductions, preoperative anterior occlusion was: anterior cross-bite (ACB) = 52%, anterior open-bite (AOB) = 1%, ACB+AOB = 30%, edge-to-edge (ETE) = 8%, ETE+AOB = 7%, normal (NML) = 1%, NML+AOB = 1%. The anterior occlusion for 59 individuals who underwent lingual reduction before 2 years of age and were > 1 year postoperative was: reduced or eliminated ACB, AOB and/or ETE = 78% with 80% of these now NML or ETE, no change in occlusion = 10%, deteriorated occlusion = 12%. The anterior occlusion for 19 individuals who underwent lingual reduction after 2 years of age and were > 1 year post-lingual reduction was: reduced or eliminated ACB, AOB and/or ETE = 63% with 33% of these now NML or ETE, no change in occlusion = 16%, deteriorated occlusion = 21%.

*Conclusions*: For the macroglossia of BWS, 1. "W" anterior lingual reduction does not impair or facilitate speech development. 2. "W" anterior lingual reduction is more likely to reduce progressive dentoskeletal deformity for individuals operated before 2 years of age than older than 2 years. 3. "W" anterior lingual reduction has minimal morbidity.

*Key words*: Beckwith-Wiedemann syndrome, macroglossia, tongue reduction

**Introduction**

A number of seemly disparate physical findings were appreciated to be a syndromic constellation by two physicians, one pathologist reporting on three unrelated infant necropsies and one pediatrician reporting a kindred affecting three siblings, almost synchronously on either side of the Atlantic: Beckwith (Beckwith, 1963; Beckwith, 1969) and Wiedemann (Wiedemann, 1964). These findings included corporal and visceral overgrowth, omphalocele, and macroglossia. Honoring their deductive observations, this syndrome gained the dual eponym of Beckwith-Wiedemann syndrome (BWS). Subsequently recognized associative findings include excessively large birth weight, perinatal hypoglycemia, ear pits and/or creases, nevus flammeus of the forehead and asymmetry of the limbs. (Elliott et al., 1994) Initially thought to be a rare condition, it is now recognized that BWS is a rather common heritable birth defect occurring in about 1/13,700 live births. (Weksberg et al., 2010) Over the past 40 years, clinical attention has focused primarily on management of hypoglycemia (DeBaun et al., 2000), prenatal diagnosis (Grundy et al., 1985) and, more recently, the detection and management of the increased risk of embryonal cancers in early childhood for individuals with BWS (DeBaun and Tucker, 1998). More recently, the genomic error associated with these anomalies has been identified as an epigenetic alteration in imprinting and methylation of several genes on chromosome band 11p15.5. (Enklaaret al., 2006) Furthermore, a relationship between specific epigenotype and specific phenotype has also been documented. (Engel et al., 2000; DeBaun et al., 2002)

Although macroglossia was recognized as one of the initial cardinal signs of the syndrome, its significance with respect to breathing, feeding, saliva control, speaking, dentoskeletal development and psychosocial integration was not appreciated until more recently. (Salman 1988; McCann et al., 1993) Surgery to reduce the tongue in BWS was advocated as early as 1970. (Aarons et al., 1970) The effect of lingual reduction for the macroglossia of BWS is minimally documented with respect to technique, timing, benefit and morbidity. (Aarons et al., 1970; Kveim et al., 1985; McManamny and Barnett, 1985; Siddiqui and Pensler, 1990; Mixter et al., 1993; Menard et al., 1995; Dios et al., 2000; Kacker et al., 2000; Tomlinson et al., 2007) Small numbers of patients and limited length of follow-up further compromise this limited literature. Hence the indications for, technique of, timing of and expected outcomes from lingual reduction for the macroglossia of BWS in infants and young children remain unclear.

This report communicates our experience with one specific technique of lingual reduction performed by the same senior surgeon over the past 28 years in 315 BWS individuals.

The objective of this report is to clarify the role of lingual reduction in the management of individuals with macroglossia secondary to BWS.

**Patients and Methods**

The records of 432 individuals with BWS evaluated and treated for macroglossia by one surgeon between 1985 and 2013 were reviewed retrospectively. The diagnosis of BWS was made by either clinical criteria or, more recently, positive DNA testing. For a clinical diagnosis, the presence of at least two of the five most common features of BWS was required. These features are: (1) macroglossia, (2) birth weight greater than the 90th percentile, (3) hypoglycemia in the first month of life, (4) ear creases or ear pits, and (5) abdominal wall defect (omphalocele, diastasis recti, umbilical hernia). (Pettenati et al., 1986)

The data were collated into the following categories: demographics, BWS non-craniofacial (hypoglycemia, omphalocele, malignancy, overgrowth), BWS craniofacial (airway, tongue, dentoskeletal, speech/language, hearing, feeding/swallowing, developmental) and non-BWS. Demographic data included birth date, gender, family history, gestational and delivery history. BWS non-craniofacial data included presence and magnitude of hypoglycemia; presence and management of omphalocele or other abdominal wall defects; presence and management of visceral malignancy; and presence and extent of overgrowth. BWS craniofacial data included perinatal and subsequent airway history; presence of, longitudinal changes in and intervention for macroglossia; dentoskeletal status with respect to anterior alveolar or incisor relationship, molar relationship and skeletal relationships; speech/language function noting delay or impairment and response to therapeutic intervention; hearing acuity; feeding/swallowing function including management of oral secretions; and cognitive/behavioral development. Non-BWS data consisted of any other significant medical history or findings not usually associated with BWS. This report focuses on those items relevant to macroglossia, its associated morbidities, its surgical treatment, and dentoskeletal development.

Data were descriptively analyzed for distribution according to several major binary alternatives: lingual reduction – yes or no; speech therapy – yes or no; anterior occlusal relationship – normal or abnormal.

**Results**

***Demographics***

The records of 319 individuals with BWS were reviewed and comprise the database for this report. The gender ratio was almost equal (M:F::0.9:1). A positive family history of BWS was present in 12% and there were a few families that might have had an affected member other than the propositus. The median gestational age at birth was 37 weeks, ranging from 25 to 42 weeks. Hypoglycemia was documented perinatally in 11% of individuals and persisted beyond the second week of life in 2%. The median age at initial evaluation was 13 months, with a range of 1-277 months. The majority of BWS individuals were American 263 (82%) with the following racial distribution: 90% caucasian, 4% African-American, 2% Hispanic, and 4% oriental. Fifty-six individuals from 25 countries comprise the non-American population.

***BWS Craniofacial Findings Prior to Tongue Reduction***

*Breathing* – Perinatal breathing issues, requiring interventions ranging from supplemental oxygen to intubation with ventilatory support, occurred in 50% of patients. Seven individuals had a tracheotomy prior to our evaluation. Often associated with prematurity, it was difficult to causally associate the respiratory issues with the macroglossia. Respiratory insufficiency rarely persisted beyond three months of age. Diagnoses for the patients with prolonged respiratory difficulties included pulmonary pathology with impaired oxygenation, tracheomalacia and sleep apnea (both obstructive and central).

*Feeding/swallowing* – Almost half (45%) of BWS neonates were described by their mothers as having difficulty initiating oral feeding. An additional 5% had problems transitioning to baby food and/or solids. Feeding/swallowing difficulties persisted beyond three months of age for only about 10% of individuals who were managed with either trans-nasal tube gavage or a surgical feeding gastrostomy. Some mothers reported successful breast nursing of their BWS infant while others were unable to have the BWS infant latch-on. Aspiration was reported in only 4 BWS infants.

*Tongue* – Macroglossia was diagnosed for all of the patients that comprise this report. Of these, 80% were symmetric and the remainder had lingual hemihypertrophy. 319 individuals underwent the same three-dimensional peripheral lingual “W” pattern tongue reduction (see Surgical Technique below) by the same senior surgeon between 1985 and 2013. The age range at tongue reduction was 2-200 months with a median age of 13 months.

*Palate* – Cleft palate occurred infrequently (3%) with 9 complete cleft palates and 2 submucous clefts.

*Speech/language* – 53% of the individuals in this report were receiving or recommended to receive speech therapy prior to tongue reduction. Assessment for factors known to predispose individuals for needing speech therapy (e.g. prematurity, intracranial bleed, delayed development, hearing loss, family history of speech therapy) documented a positive correlation between those BWS individuals with two or more risk factors and receiving speech therapy. In contrast, those with no or only one risk factor were unlikely to receive speech therapy. Specific assessment for those phonemes thought to be at risk with impaired tongue mobility showed no acoustic difference among BWS individuals without tongue reduction, BWS individuals with tongue reduction and normals. However, the anatomic production of such phonemes usually was different in the tongue reduction patients.

*Dentoskeletal* – The anterior occlusal (alveolar, in edentulous infants, or incisal) relationship at initial presentation represents the natural history of the effect, if any, of macroglossia with or without primary mandibular hypertrophy and/or maxillary retrusion on dentoskeletal development. A normal anterior occlusal relationship was present in only 1% of BWS individuals without tongue reduction. Of the 99% with abnormal anterior occlusion, the relationships were: negative anterior cross-bite alone (52%); edge-to-edge alone (8%), anterior openbite alone (1%); mixed negative anterior cross-bite or edge-to-edge with anterior openbite and/or lateral openbite (39%). An additional 3 BWS individuals had anterior lateral openbite alone that was associated with lingual hemihypertrophy.

***BWS Craniofacial Findings Following Surgical Tongue Reduction***

*Breathing* – There were 10 individuals who had pre- and post-lingual reduction sleep studies that documented resolution or significant improvement in obstructive sleep apnea post-operatively. In addition, 4 individuals had marked reduction or elimination of supplemental oxygen following tongue reduction surgery. Two individuals had respiratory compromise following tongue reduction that required re-intubation with subsequent successful extubation. One individual required peri-operative tracheotomy: the local ENTs recommended tracheotomy for this infant but parents preferred to try tongue reduction first; the child was subsequently decannulated successfully. Of those individuals with pre-existing tracheotomy prior to tongue reduction surgery, all 7 were successfully decannulated post-operatively.

*Feeding/swallowing* – Following lingual reduction, presurgical normal oral feeding resumed within 4-6 weeks. Improved feeding was reported postoperatively for 57% of lingual reduction individuals. No individual who was an oral feeder prior to tongue reduction surgery became non-oral post-operatively.

*Tongue* – Perioperative morbidity was infrequent and included respiratory distress (see above), delayed hemorrhage requiring transfusion (3 cases) and wound dehiscence (2 cases). Several children had premature suture disruption with secondary healing: only one of these required surgical revision. Two children developed post-operative tongue surface irregularities for which parents requested surgical removal. One child had recurrent sublingual mucoceles that were managed surgically. None of these patients had any permanent disability due to the tongue reduction. One child had complete dehiscence as well as dehiscence of a synchronous (different surgeon) umbilical hernia repair: this infant was found to be malnourished on pre-albumin levels, responded to gavage feeding and healed both the tongue and abdominal wall without complication. While a decrease in tongue motility, especially protrusion, was noted for 80% of patients during the first postoperative year, this did not impair feeding or speech. Of the 319 individuals who initially underwent tongue reduction by this surgeon, 3 received, and a fourth is recommended, a second reduction due to persistent macroglossia and improved but not resolved severe dentoskeletal deformity: each of these had the initial reduction within the first 6 months of life and had 10/10 macroglossia.

*Speech/language* - Of those BWS individuals who were already speaking at the time of tongue reduction surgery, none had persistent impairment of speech post-operatively. Of the large majority who were not yet speaking or only had minimal speech at the time of tongue reduction surgery and were cognitively normal, none had impairment of articulation related to the tongue.

*Dentoskeletal* – The anterior occlusion for 59 individuals who underwent “W” pattern anterior lingual reduction before 2 years of age and were at least one year postoperative was: reduced or eliminated anterior cross-bite, anterior openbite and/or edge-to-edge = 78% with 80% of these now normal or edge-to-edge; no change in occlusion = 10%; and deteriorated occlusion = 12%. The anterior occlusion for 19 individuals who underwent lingual reduction after 2 years of age and were at least one year post-lingual reduction was: reduced or eliminated anterior cross-bite, anterior openbite and/or edge-to-edge = 63% with 33% of these now normal or edge-to-edge; no change in occlusion = 16%; and deteriorated occlusion = 21%.

***Tongue Reduction Surgical Technique***

The patient is nasotracheally intubated following induction of mask general anesthesia. A single dose of parenteral steroid is administered to minimize postoperative swelling. No prophylactic antibiotics are administered unless there is another indication for doing so such as congenital heart disease.

Three traction sutures are placed into the tongue: one at the tip and two on either side. Using these sutures, the tongue is protruded from the mouth and spread with moderate tension. The mandibular incisors, or alveolus if the incisors are unerrupted, are palpated through the dorsum of the tongue and that point marked as the most dorsal aspect of the central “V” excision. The rays of the “V” are 1.5 – 2cm long depending on the age of the child. Two lateral lines are extended posterolaterally from the tips of the central “V” toward the posterior aspect of the mandibular alveolar arch on each side thereby creating the “W” excision pattern. The dorsum is reflected using the traction sutures, and a reciprocal “W” excision pattern is drawn on the ventral lingual mucosa. Care must be taken to preserve sufficient ventral mucosa to prevent tethering of the tongue to the floor of the mouth. The dorsum is reexposed and the mucosal incisions are cut with a scalpel and then the muscle is divided for approximately 2/3rd of its bulk with the cutting cautery. The muscle is resected as beveled wedges to diminish anterior tongue bulk. Individual bleeders are identified as cut, clamped and cauterized. The ventrum is reexposed and after cutting the mucosa with a scalpel, the remainder of the muscle is divided with the cutting cautery and final hemostasis secured. The tongue is reconstructed with 2-0 polyglycolyic acid sutures at the tips of the dorsal and ventral mucosal flaps and as vertical mattress sutures in the midline and along the lateral wedges. Dorsal to ventral mucosal approximation is secured with running, locking 3-0 polyglycolyic acid sutures. This completes the surgical aspect of the lingual reduction.

An orogastric tube is passed to decompress the stomach and the patient is emerged from anesthesia with removal of the nasotracheal tube in the operating room. A small caliber silastic nasogastric feeding tube is passed to facilitate nutrition and weaning from parenteral narcotics until the child is willing to take po. Elbow restraints are used until the feeding tube is discontinued and then the restraints are discontinued also. The child is transferred from the operating room to the post-anesthetic recovery room and then to the intensive care unit for overnight observation. Intravenous morphine and gavage or po acetaminophen+hydrocodone elixir are administered as needed for pain control.

The child is allowed to take a soft diet as soon as he or she is willing to do so. There are no restrictions on feeding tools, excepting sharp objects (e.g. fork): mother’s breast, nipple bottle, spouted cup, regular cup, tube-syringe feeder, spoon, fingers, etc. The child is discharged home once satisfactory po intake and comfort with oral analgesics are achieved. This occurs on an average three days after the operation, in our experience, with a range of two to nine days. Other than avoiding hard, sharp foods (if already part of the child’s diet), there are no specific post-discharge instructions. Speech/language development and dentoskeletal status are evaluated annual in small children and biennially in older individuals.

**Discussion**

Macroglossia was recognized as a major phenotypic indicator of the syndromic constellation of findings now referred to as Beckwith-Wiedemann syndrome (BWS) as early as Beckwith and Wiedemann’s seminal reports of 1963-4. (Beckwith, 1963; Beckwith, 1969; Wiedemann, 1964) It is one of the five most common anomalies associated with BWS and is used as a major criterion for the clinical diagnosis. Macroglossia occurs in the large majority of individuals affected with BWS, ranging from 82-100% of reported large series. (McManamny and Barnett, 1985; DeBaun and Tucker, 1998; Elliott et al., 1994) Nonetheless the literature is inconclusive regarding a number of issues related to macroglossia: (1) the definition of macroglossia; (2) the natural history of unoperated macroglossia in BWS including its effect upon breathing, eating, saliva control, speech, dentoskeletal development, and psychosocial integration; and (3) the effect of lingual reduction upon the afore listed functional and growth issues.

A quantitative definition of macroglossia has remained elusive because the tongue changes shape and size with muscular activity. We define macrogossia as a tongue that protrudes beyond the lips more than 50% of the time when awake and asleep in the infant and young child or can only be contained within the oral cavity with lip seal by conscious effort.

The natural history of unoperated macroglossia in BWS has not been documented in a rigorous fashion. Based upon case or small series reports, it has been speculated that macroglossia can interfere with breathing by causing either acute or chronic upper airway obstruction with development of secondary pulmonary hypertension and cor pulmonary in the latter. Rimell et al (1995) objectively reported airway issues in 13 BWS patients from two medical centers evaluated and treated between 1986 and 1993. Of these, 2 underwent tracheostomy within the first 3 months of life and 7 tonsillectomy/adenoidectomy after 18 months of age for upper airway obstruction. They felt that macroglossia, specifically enlargement of the base of the tongue, accounted for the neonatal airway issues and recommended therapeutic tracheotomy rather than anterior lingual reduction, which they felt was an inadequate procedure for the problem. However there was only one case report to support this assertion. Our experience with successful decannulation following the anterior “W” lingual reduction in all 7 of our BWS patient who presented with a tracheotomy contradicts their conclusion. While 50% of BWS infants in the current study had perinatal airway issues these seemed more related to prematurity than to macroglossia. That only 7 out of 338 BWS individuals in this report received tracheotomy and for indications that were impossible to assess objectively, suggests that the need for tracheotomy in the BWS neonate is very infrequent. While a small number of our BWS infants with respiratory issues had improved oxygenation and/or diminished sleep apnea following anterior lingual reduction, the numbers are too small to clearly assert a causal relationship. In older children with BWS and upper airway obstruction, these authors stated that the obstruction “most likely results from tonsil and adenoid hypertrophy, rather than from macroglossia.” While our data suggests that Rimell et al. have overestimated the upper airway morbidity in BWS, probably due to patient selection bias due to their discipline of Otolaryngology, the importance of evaluating all forms of airway obstruction in a BWS patient with breathing issues rather than assuming the pathology is the enlarged tongue is supported by their data.

It has been speculation that macroglossia interferes with swallowing. The current study confirms that almost half of BWS neonates will have some feeding difficulties but these are usually short term and not of nutritional consequence. However, this includes a subpopulation that requires prolonged gavage feeding via nasogastric or gastrostomy tube. Further study is required to determine if these infants have other risk factors for impaired feeding such as marked prematurity, upper airway issues, and/or CNS dysfunction. Nonetheless, that over half of parents reported improved feeding following recovery from lingual reduction, suggests that macroglossia does indeed have some, albeit mild, negative effect on feeding.

The effect of macroglossia in BWS on oral competency, with respect to both drooling and appearance, is unreported. However, some literature on macroglossia in Trisomy 21 individuals asserts that it causes “stretching of the lower lip” and drooling that is “difficult to control” (Champion, 92) as well as “frequent airway and throat infections” (Olbrisch, 85) Postoperative consistent oral continence of the tongue was reported for almost all patients in the current study. Multiple parents have commented upon the “normal” appearance of their BWS child after the tongue reduction and the elimination of inquiries as to whether the child had Down syndrome, a common lay misinterpretation of the protruding tongue in BWS children according to the parents of our BWS patients. Others have asserted that BWS individuals “grow into” their enlarged tongue without any secondary morbidity, although supportive data has not been presented. In our evaluation of older BWS individuals without early tongue reduction, most were tongue continent unless they concentrated on a task and then the large tongue protruded. One cost of volitional oral continence seemed to be the hypertrophic expansion of the oral cavity with attendant dental and jaw pathologies.

Macroglossia has been assumed to interfere with speech intelligibility. (Van Borsel et al., 1999) The current study documents that non-BWS factors seem much more likely to be the causal agents for the speech/language problems in half of the BWS population rather than the macroglossia. Furthermore, lingual reduction surgery seems to neither impair nor significantly enhance speech. However spontaneous correction of anterior crossbites and anterior and lateral openbites following early anterior lingual reduction would eliminate the sibilant distortions usually seen with these dentoskeletal anomalies.

 Several reports have documented, using established cephalometric criteria, a characteristic dentoskeletal deformity of BWS individuals without lingual reduction surgery. This phenotype includes anterior open bite, mandibular prognathism with an abnormally obtuse gonial angle and increased effective mandibular length, and splaying of the dentition with labioversion. (Friede and Figueroa, 1985; McCann J et al., 1993) Based upon a small number of patients, Menard et al. (1995) concluded that mandible was not truly prognathic in young children with BWS and could even be retrognathic but became truly prognathic, using standard cephalometric criteria, in older BWS individuals who had not undergone lingual reduction. They noted that mandibular alveolar protrusion was present in all of the 11 BWS individuals they studied. They concluded that it was the alveolar protrusion combined with an excessively obtuse gonial angle that produced the anterior open bite and the illusion of mandibular prognathism. The maxilla, in BWS, has been shown to be displaced ventrally, with an excessively obtuse craniomaxillary angle, and deficient vertically. (Menard et al.,1995) The current study did not use cephalometric analysis due to the young age of most of the patients at our center. That only 1% of our BWS patients had normal anterior dental/alveolar relationships on initial evaluation prior to any tongue reduction regardless of age, not only reflects the prevalence of pathologic jaw relationships in this population but also belies the oft quoted assertion that they “grow into their tongue” without co-morbidity. Concerns regarding the psychosocial consequences of the BWS facies have been raised (Menard et al., 1995) but not documented. While the current study did not investigate this issue, two of our adolescent tongue reduction patients specifically requested surgery because they were “tired of thinking to keep my tongue in my mouth”.

Lingual reduction has been advocated for persistent macroglossia due to a variety of etiologies, including syndromic, neoplastic and vascular malformation. There is no consensus in the literature regarding the indications for, timing of or technique used for lingual reduction regardless of etiology. That there is a relationship between dental position and lingual pressure on the teeth is an accepted principle of orthodontics. (Proffit, 1978) Attempts to quantify the effect of lingual reduction upon pressure exerted on the teeth by the tongue have not been successful. (Frohlich et al., 1993) Nonetheless, orthodontists at times refer patients with anterior open bites, excessive interdental spacing and/or bimaxillary protrusion to surgeons for lingual reduction in the belief that the excessively large tongue is responsible for the malocclusion. (Becker, 1966; Ingervall and Schmolker, 1990). It was, therefore, logical to hypothesize that early lingual reduction in individuals with macroglossia secondary to BWS would minimize or eliminate the unfavorable dentoskeletal aspects of that syndrome. While performance of lingual reduction based upon this hypothesis has been reported (McManamny, 1984; Giancotti et al., 2002), long-term outcome studies attempting to confirm the validity of this hypothesis have been few. Menard et al. (1995) reported on six BWS individuals who underwent “keyhole” pattern lingual reduction between the ages of 10 and 76 months of age, with a mean age at reduction of 41 months. Of these, the molar occlusion was Angle class I for all excepting the child operated upon at 76 months of age who was Class III 3 ½ years later. Follow-up ranged from 2 years to 14 ½ years with a median of about 7 ½ years. Comparatively, four BWS individuals without lingual reduction were studied cross-sectionally. Of these, two, ages 1 and 7 years at evaluation, had a Class I occlusion while the 11 and 18 year old were Class III. Detailed longitudinal pre and post lingual reduction cephalometric analysis was provided for only two patients. Unfortunately, the effect of lingual reduction upon the mandibular alveolar protrusion, which was documented by these authors as one of two critical pathologic findings, the other being an excessively obtuse gonial angle, was not reported. These authors hypothesized that the “craniofacial features seen with Beckwith-Wiedemann syndrome are secondary to the macroglossia associated with the syndrome …”. A single long-term BWS case report of occlusal and cephalometric findings following lingual reduction at 8 years of age with perioperative adjunctive orthodontic intervention (chin cap and tongue crib) and definitive edgewise treatment in the early to mid teens, by Miyawaki et al. (2000), documented normal occlusal, skeletal and facial profile relationships at 18-7 years of age. Thus while the published findings regarding the dentoskeletal consequences of lingual reduction performed in growing children with BWS are suggestive of benefit, a rigorous outcome study is yet to be published. The findings of the current study suggest that early lingual reduction can reverse the deleterious effect of macroglossia upon the developing teeth and jaws. We do have a small number of individuals who underwent early tongue reduction and now are in adult dentition: none of these has required jaw surgery to correct persistent negative anterior cross-bite and dentoskeletal disharmony. Longitudinal documentation of dental occlusion and maxillofacial growth in a larger population of BWS individuals, currently in progress, is necessary to define the specific effect(s) of early lingual reduction.

**Conclusions**

1. Untreated macroglossia of BWS is associated with unfavorable dental occlusion in most affected individuals that requires a combination of orthodontics and jaw surgery in the teens for correction.
2. Early surgical lingual reduction (<24 months of age) using the anterior “W” pattern minimizes secondary dentoskeletal deformity.
3. “W” pattern lingual reduction for BWS macroglossia has minimal morbidity for breathing and eating.
4. “W” pattern lingual reduction for BWS macroglossia reduces or eliminates obstructive sleep apnea and facilitates tracheotomy decannulation for some individuals with an unstable upper airway due to macroglossia.
5. “W” pattern surgical lingual reduction does not seem to impair or facilitate speech/language development.
6. Impaired speech/language development in BWS may be related to risk factors other than macroglossia or dentoskeletal deformity.
7. A prospective study of surgical lingual reduction is necessary to accurately define its indications, benefits, morbidity, ideal timing and ideal technique.

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