The Modified Method of Tongue Reduction in Beckwith-Wiedemann Syndrome

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The indications for surgical tongue reduction for syndromic macroglossia are airway obstruction, swallowing problems with failure to thrive, dental deformities, articulation disorders, recurrent tongue bitten, oral incompetency, persistent drooling, and cosmetic concerns. However, various methods of tongue reductions have yielded mixed results. Our patient 1 year 11months old with Beckwith–Wiedemann syndrome received surgical tongue reduction at China Medical University Hospital. A modified complex mucous-muscular flap without interrupting the vascular base and preserving the tongue tip was formed. The longitudinal full thickness central wedge resection spared tongue tip apex with lateral extended resection. The transverse part anterior to the circumvallate papilla, was resected in partial thickness. Our approaches provide a shortened, non-ankylosed tongue. The benefits are better cosmesis, dynamic esthetic improvement when talking and relief of drooling, and one and half year, better comprehensive articulated pronunciation and her oral competency. It is important that tongue reduction addresses the global nature of the macroglossia and preserving a tapered tongue tip with length sufficient to permit normal tongue movements. Our method may be an alternative for tongue tailoring. (J Plast Surg Asso R.O.C. 2008;17:390∼398)

Key words: tongue reduction, Beckwith–Wiedemann syndrome, Macroglossia

Introduction

Beckwith–Wiedemann Syndrome (BWS) is an overgrowth disorder initially described by
Beckwith in 1963\(^1\) and Wiedemann in 1964.\(^2\) It was characterized by a collection of congenital anomalies and time-reliant abnormalities. Most common manifestations are exomphalos, macroglossia, and gigantism (EMG syndrome).\(^3\) The prevalence of BWS is around 0.07 per 1000 births.\(^4\) The syndrome results from chromosomal changes in the imprinted 11p15.5 region that cause amplified levels of the fetal growth factor IGF (insulin-like growth factor).\(^3\) Fifteen percent of cases are familial and 85 percent are sporadic.\(^5\),\(^6\) Other clinical features frequently correlated with BWS include postnatal somatic gigantism (88%), abdominal wall defects and hernias (80%), abnormal earlobe creases/pits (76%), hypoglycemia (63%), nevus flammeus of the of the forehead (62%), nephromegaly (59%), hemihypertrophy (24%), congenital heart defects (6.5%) and cleft palate (2.5%), and an increased incidence of childhood neoplasms.\(^6\),\(^9\) Macroglossia is seen in 80 to 99 percent of patients with BWS.\(^6\),\(^8\)-\(^10\) Surgical tongue reduction (partial glossectomy) endeavors to correct macroglossia to grant normal tongue function and appearance. Not all patients with BWS will require tongue reduction, as the condition is not phenotypically homogenous and, often, mildly symptomatic macroglossia will improve without intervention as a result of normal mandibular and skeletal growth.

For a child, teenager, or adult, an unusual tongue enlargement may imply some congenital syndromes such as BWS, Down syndrome, hypertelorism, hyperthyroidism (cretinism), and other diseases.\(^11\) The indications for surgical tongue reduction are airway obstruction, swallowing problems with failure to thrive, tongue protrusion, dental deformities, articulation disorders, recurrent lingual trauma, failure of oral competency, persistent drooling, cosmetic concerns and preservation of many functions of the jaws and oral cavity.

Vogel et al developed a detailed classification to define congenital and acquired forms of macroglossia.\(^4\) Congenital macroglossia comprises vascular and muscular forms as well as various syndromic symptoms and consequences of congenital tumors. Acquired macroglossia is a tongue enlargement that results from growth of some tumors, in addition to an effect of excessive development of the upper or lower jaw of the child, and other local insults. The enlarged tongue has both functional and cosmetic deformity, which may affect the oral airway, speech, and the development of the jaws.\(^12\) There are various methods of tongue reductions reported with mixed outcomes.\(^11\) A wedge-shaped resection of the tongue tip is suggested by Rheinwald with a width of 3 to 5 cm.\(^11\) Pichler and Trauner proposed tongue reduction of two sites: in the area of the dorsum and the tip.\(^11\) However, retaining tissue between the areas of resection often leads to insufficient correction of the problem.\(^11\) That is why Obwegeser and later Köle, modified the procedure of including a block excision of the tip and the middle part of the tongue.\(^11\) These methods are very much in common and recommend an efficient way to provide longitudinal as well as transverse tongue reduction. All the aforementioned techniques have the same shortcomings: postoperatively the patient loses the tip of the tongue, which is the most sensitive part for somatosensation and taste.\(^11\)

**Report of A Case**

The patient we reported was a one year 11 months old girl in September 2005. She suffered from respiratory distress at birth and received herniorrhaphey for omphalocele. The diagnosis was Beckwith-Wiedemann syndrome, also named as EMG syndrome. Physical examination showed only three features of the syndrome:
macroglossia, gigantism and hypertrophy of the clitoris (Figs. 1A, B). Her chromosomal study was normal. Upon examination it became clear that at first it would be necessary to reduce the child’s hypertrophied tongue, and only then would it be possible to solve sleep apnea, incomprehensive speech, persistent drooling problems, anterior open mouth and cosmesis.

A stay suture was placed to draw the tongue forward so that the posterior tongue can be accessed, and this may be left for 48 hours postoperatively to assist in the wound and airway management. Incision margins are marked with ink, and local infiltration with a vasoconstrictive agent to reduce intraoperative blood loss. A modified complex mucous-muscular flap without interrupting the vascular base is formed. Further, a solid line marks the area of longitudinal full thickness wedge resection (1.8x1.2x1 cm³) sparing apex on the tip of the tongue (Fig. 2). The lower lateral extended resection of the tongue is outlined (0.5 to 0.7 cm in width) (Fig. 2). The transverse part as marked in a dotted line was resected lower to the circumvallate papilla by partial thickness in depth, 0.5 cm in length and 3 cm in width, as piece by piece to avoid injuring the neurovascular bundles that run inferior-laterally.

There were no contraindications and surgery was performed under general anesthesia. The tongue reduction was performed according to the aforementioned technique with maintaining taste buds. The parents agreed on the postoperative care to keep her intubated and sedated in pediatric intensive care unit for one week. Pain control is essential. Postoperative antibiotics for a clean contaminated wound are recommended for 3 doses. The postoperative course included tongue swelling and lymphostasis, which gradually normalized. The tongue swelling diminished gradually so that the tongue was confined within the oral cavity and regained its mobility. However, because the tongue is a muscle, a quick return to function greatly aids in shortening the time to normal recovery. Cold clear liquid diets are admirable early troches for the tongue.

During the second postoperative week, continuous pulse oximeter readings and observation in at least a step-down unit are advocated to maximize patient safety. When tongue swelling no longer presents a threat to the airway and the patient is able to tolerate an appropriate oral diet, she was discharged on the 15th hospitalization day. The girl felt quite well after the operation.

Perioperatively, we administer systemic steroids (Dexamethasone 2 mg, q6h) for one week to reduce the risk of edema. The nasogastric tube feeding started one day after surgery. Careful monitoring of blood glucose levels is required, as patients with BWS are at risk of hypoglycemia when fasting. The surgery resulted in sufficient tongue reduction so that she may close her mouth one month later. The tip of the tongue was somewhat wider as compared with normal, but moved easily. The girl was able to close her mouth with ease, applying no tension to her lips. Her tongue reduction surgery has provided several benefits as follows:

1) Reduction in tongue protrusion did good to cosmetic improvement.
2) Improvement in the visual appearance of speech made the child no longer talk with the tongue protruded.
3) Drooling was resolved as long as the problems were only caused by the large tongue.

Afterward, the patient consulted with our orthodontists. We started an active orthodontic treatment, and the detailed recommendations on further treatment were given to orthodontists. The girl was examined every 6 months after...
surgery and the tongue had almost regained its normal size, thus the oral competency was achieved. Her teeth have gradually accepted normal position; all appearance was normalized except there was negative over jet (1.5mm) in a mild class III occlusion (Figs. 3A, B, C).

Fig. 1. One year 11 months old girl with Beckwith-Wiedemann Syndrome presented characteristic macroglossia. She was not able to close her mouth.  
1A: Front view.  1B: Lateral view.

Fig. 2. The illustration of the operative method of this modified complex mucous-muscular flap. The solid line marks the area of longitudinal full thickness wedge resection (1.8x1.2x1 cm³) sparing apex on the tip of the tongue. The lower lateral extended resection of the tongue is outlined (0.5 to 0.7 cm in width). The transverse part as marked in a dotted line was resected lower to the circumvallate papilla by partial thickness in depth, 0.5 cm in length and 3 cm in width, to avoid injuring the neurovascular bundles that run inferior-laterally.
Discussion

Indications for surgical tongue reduction in patients with macroglossia include airway obstruction, sleep apnea, feeding difficulties, specific articulation or dental disorders, recurrent local trauma, and cosmetic concerns. Where airway obstruction or sleep apnea is the major indication, it is important to examine for tonsillar and adenoidal hypertrophy, which may contribute and should be treated accordingly. Although not an absolute indication for surgery, dental deformities in BWS including mandibular prognathism, anterior open bite, and malocclusion are secondary to the effects of macroglossia and are not attributable to somatic gigantism or growth hormone effects. In severe cases, desiccation of the protruding tongue can occur and inability to achieve oral competence can result in drooling. Tongue protrusion may give the appearance of mental retardation, which can cause tremendous distress to patients and families and result in poor bonding with parents, social ostracism, and low self-esteem.

There is no consensus as the best timing to carry out the surgery but generally under the age of two years is thought to be favorable to prevent the lower jaw from being pushed forward. Nevertheless, this has not yet been proven. Although surgery may be delayed well past the age of 6 months in the majority of cases, gross macroglossia may require earlier tongue reduction, as a successful reduction may negate the need for a surgical airway or prolonged nasogastric feeding. However, earlier intervention carries increased anesthetic and airway risks, and there is also a risk of significant regrowth of the tongue postoperatively, as insulin-like growth factor 2 is still circulating in relatively
high levels in the neonatal period. Staged reductions may also improve long-term outcomes in these patients. Regardless, gross macroglossia in the neonate remains a difficult management problem, even for experienced practitioners with a full range of options of reduction techniques.

The management of macroglossia has been reported as early as 1658. Historically, boiling water was injected into the tongue successfully to reduce tongue size. There were several proposed procedures for tongue reduction. The key to most of these techniques engrosses reduction of the central tongue bulk and tongue length. Thus one third to one-half of the tongue should be resected anterior to the circumvallate papillae. Care must be taken to avoid damaging the neurovascular bundles that run inferior-laterally. Standard tongue reductions may result in ankylosed, globular tongues with an insensate tip whereas central tongue reduction does not address the anterior tongue. An anterior keyhole method used the KTP or the CO₂ laser for tongue reduction, which removes both anterior and the wedges of the tongue. The keyhole technique helps in providing a reduced, functional and sensate tongue. The complications of the keyhole tongue reduction technique are similar to those of other techniques, which include tongue edema, wound dehiscence and wound infection. The efforts to reserve tongue tip should not be underestimated. One special concern in this technique is post-operative edema which requires either prolonged intubation with neuromuscular blockade or a pre-operative tracheotomy. The described surgical procedure by Kruchinsky results in significant longitudinal and transversal tongue reduction, while at the same time maintaining integrity of the tongue tip. Unless the technology as radiofrequency ablation is able to remarkably decrease size without compromising function, the treatment of macroglossia will likely remain unchanged. Despite the various techniques described, correction should be adapted to the individual patient. Staged procedures or re-operations may also be necessary. A successful reduction allows for the tongue to rest behind the lower dental alveolus and enough tissue should be left for the tongue to be able to protrude and allow moistening of the lips. Our approach has achieved the goal.

Operation reports did not suggest that any of the subjects in this study had tongue hemihypertrophy preoperatively; however, hemihypertrophy has a prevalence of 24 percent in the BWS population, and it is to be anticipated that in some occasions modification of surgical techniques will be necessary to reduce asymmetry. Histologic examination of tongue tissue in BWS invariably demonstrates microscopically normal muscle and nerve fibers and thus surgical reduction is theoretically curative with adequate resection. On the other hand, extensive resection risks damage to the neurovascular bundles, and the global nature of the macroglossia poses difficulties in reconstructing a normal tongue. Correction of the macroglossia prevents the development of these deformities and has been documented to cause regression in some cases. Speech may also be improved following correction of macroglossia. There are no deglutition abnormalities as they are able to initiate swallow using tongue tip and to hold bolus in oral cavity.

The treatment of BWS patients requires a multidisciplinary approach that includes orthodontics, orthopedics and surgical intervention. It is also necessary to address the significance of diagnosis at an early age and timely treatment to reduce the development of dento-skeletal alterations with a tendency to Class III malocclusion and an open-bite.
Conclusions

Pediatric patients with symptomatic macroglossia and BWS who undertake surgical tongue reduction are unlikely to have fully normal tongue function and appearance as adults. The most common abnormality is that of continued disproportionate tongue bulk, and many patients report having a short tongue tip. Specific speech sound errors are very common but do not result in significant levels of self-awareness about speech or influence on interactions. It is important that the surgical technique used addresses the global nature of the macroglossia and aims to retain a tapered tongue tip with length sufficient to permit normal tongue movements. We achieved an acceptable tongue bulk and adequate length of tongue tip by transverse and longitudinal resections of tongue at one stage.

Reference

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改良的舌頭縮小術於 Beckwith-Wiedemann 症候群病人

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本文敘述一例為 Beckwith-Wiedemann 症候群病人，接受改良的橫向和縱向舌頭縮小術。巨舌會造成呼吸道堵塞、吞嚥問題、牙齒畸形、發音不清、反覆性舌頭被咬、閉口不能以及流口水，導致無法長期存活；且縮小手術後可能達不到完全正常的舌功能及外型。患者為 1 歲 11 個月大女孩，罹患 Beckwith-Wiedemann 綜合症候群，她接受了舌頭縮小術。藉由設計好的黏膜肌肉皮瓣，避免舌下血管神經傷害。術後，舌頭腫脹於兩週後消退，舌頭較短且不僵硬，此術式提供了幾項好處 1) 靜態舌頭外凸提供外觀的改善。2) 談話時，舌頭也不再突出。3) 解決流口水問題。經過一年半的追蹤，她的舌頭可自由運用於講話時；而在關閉口腔時，也不會咬到舌頭；同時講話也較先前清晰，讓旁人易於了解。本文提供的方式可以使整體的舌頭縮小，保存正常的舌尖外型，並維持足夠長度允許正常舌頭的運動。我們的經驗是一個臨床上舌頭縮小術可行的方式。