

Reduction glossectomy for macroglossia in children

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Background/aim Although several surgical techniques have been described for reduction glossectomy in children, many general pediatric surgeons are still skeptical about the treatment of patients with macroglossia because of potential surgical complications. The aim of this study was to describe our initial surgical experience with reduction glossectomy in a series of eight patients with macroglossia.

Materials and methods A retrospective file review was carried out for all patients who underwent reduction glossectomy during the period from October 2009 to December 2014. Charts were designed to collect the following data from the patients' files: age; etiology of macroglossia; and full clinical examination including the functional respiration, deglutition, phonation deficit, and dental occlusion alterations.

Results The records of eight patients were retrieved. Five patients, in whom macroglossia affected the tongue width and length, underwent peripheral glossectomy, whereas two patients, in whom macroglossia affected the tongue in all directions, underwent key-hole glossectomy. In one patient in whom only half the tongue was affected, a central longitudinal strip was excised. Motor and sensory

functions, especially taste sensation, were not affected by resection. Speech articulation errors were corrected in cases that started speech. One complication occurred in one patient who had undergone a key-hole glossectomy: wound infection, followed by partial wound disruption.

Conclusion Partial glossectomy for macroglossia in children is both feasible and safe. It results in minimal complications. Many clinical problems caused by the pathology improve after surgery repair. The recommended surgical technique should be based on the extent of involvement of the tongue. *Ann Pediatr Surg* 11:115–119
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Introduction

Macroglossia is a generalized term used to describe the tongue that protrudes beyond the teeth during a natural resting posture [1]. The Myer classification, on the basis of the extent of involvement, subdivides macroglossia into generalized or localized [2]. Vogel classification, on the basis of etiology, divides macroglossia into true or relative [3]. When a primary disorder of tongue tissue leads to macroglossia, it is termed true and when affected secondarily, such as by amyloidosis, it is referred to as relative macroglossia. True macroglossia originates from several causes: idiopathic muscular hypertrophy as occurs with Beckwith–Wiedemann syndrome [4,5], vascular malformations such as angiomas and lymphangiomas [6], tumors such as dermoid cysts, teratomas, myoblastomas, rhabdomyomas, sarcomas, fibromas, and plasmocytomas [7], inflammatory disease such as glossitis and Ludwig's angina [8], allergy-related edemas [9], pharmacologic alterations [10], endocrine disorders such as gigantism, acromegaly, myxedema [11,12], infective pathoses such as tuberculoma of the tongue [13], and dermatosis (pemphigus) [14]. Relative macroglossia may occur with Down syndrome [12].

In terms of frequency, the most frequent causes are idiopathic muscular hypertrophy, Down's syndrome, lymphangiomas, angiomas, and fibromas [15].

Macroglossia can affect all oral functions such as breathing, chewing, swallowing, and speech [3] and can cause esthetic problems such as perception of mental retardation, widened interdental spaces, and mandibular prognathism [3,16].

The multiple techniques advocated for tongue reduction show that an ideal procedure is yet to emerge. This is understandable as the condition is relatively rare, with a variation in the degree of macroglossia. Also, there remains no consensus on the timing of tongue reduction. The collection of follow-up objective data is also difficult [17].

Several surgical procedures were described for glossectomy. The surgical techniques may be subdivided into two groups: glossectomy along the median line and peripheral glossectomy. Both techniques include the resection of a tissue portion and the subsequent suture of the margins [15].

With the glossectomy procedures, mobility of the tongue will not be significantly decreased [18]. The lateral, downward, and protrusive movements will usually remain unchanged, although movement of the tongue cephalad may be somewhat restricted. The more muscle removed from the anterior tongue, the less upward mobility the tongue will retain. Taste sensation appears to be unaltered after glossectomy [18]. Even though the primary taste buds for sweetness are located in the anterior tongue, the other taste buds (sourness, bitterness, saltiness) seem to be stimulated sufficiently, by sweets, to provide the appropriate sensation [19].

The aim of this study was to describe our initial surgical experience with reduction glossectomy in a series of eight patients with macroglossia.

Fig. 1



Peripheral reduction glossectomy. (a) Preoperative photo of a 6-month-old female patient with Beckwith–Wiedemann syndrome, (b) marking the peripheral incisions and vertical mattress suture, (c) excision of peripheral tissue, (d and e) peripheral reduction glossectomy is completed, (f) outcome 5 weeks after surgery.

Materials and methods

The study was designed as a retrospective file review of all patients who underwent reduction glossectomy in Tanta University Hospitals and affiliated hospitals during the period from October 2009 to December 2014. The study was approved by IRB. Charts were designed to collect the following data from the patients' files: age; etiology of macroglossia; and full clinical examination including the functional respiration, deglutition, phonation deficit (evaluated by a speech therapist for all children older than 2 years), and dental occlusion alterations (evaluated by an oral and maxillofacial surgeon).

In the presence of a musculoskeletal deformity with a malocclusion, the sequence of surgical intervention was either reduction glossectomy, followed by orthognathic surgery, orthognathic surgery, followed by reduction glossectomy, or both in one surgical stage. The first option was used when extensive orthodontics were necessary before orthognathic surgery, and the size of the tongue impeded the required orthodontic movements. The second sequencing option was indicated if occlusion instability developed after orthodontics and orthognathic surgery.

Surgical technique

After the parents had signed an informed consent, the surgery was planned. The choice of the surgical

technique was made on the basis of the type of macroglossia. In patients with macroglossia affecting mainly the width of the tongue, peripheral glossectomy was performed (Fig. 1). However, in patients with macroglossia affecting all dimensions of the tongue, a key-hole glossectomy was performed (Fig. 2). In one patient with only half the tongue affected, a central longitudinal strip was excised (Fig. 3).

Peripheral glossectomy

A marker pen was used to highlight the amount of tissue on the tongue periphery on either side to be removed. We have modified the Dingman and Grabb [20] technique by starting with the insertion of a vertical mattress sutures along the incision line to minimize blood loss. The incision was made just peripheral to the vertical mattress sutures by a scalpel and then electrocautery was used to complete the excision, taking more tissue from the ventral aspect than the dorsal aspect. Wounds were closed using interrupted polyglactin sutures (Vicryl; Ethicon, New Jersey, USA) (Fig. 1).

Key-hole glossectomy

We used the technique described by Morgan *et al.* [21]. The tongue resection consists of an anterior wedge combined with a posterior circular incision, which gives

Fig. 2



Central key-hole glossectomy. (a and b) Preoperative photos of a 12-month-old female patient with Beckwith–Wiedemann syndrome, (c) marking the key-hole incision, (d) excision of central tissue, (e) reduction glossectomy is completed, (f) outcome 12 weeks after surgery showing healing after partial dehiscence at the tip of the tongue.

the appearance of a key-hole. The incision line was marked either by a marker pen or by diathermy, ensuring that the resected area was symmetrical around the midline of the tongue to ensure a good lingual contour and cosmetic result (Fig. 2).

The initial resection should be conservative as it is very difficult to replace tissue bulk in this area. Tongue bulk can be further reduced by beveling the posterior incision outward. The incision was carried out using a scalpel and was then completed with electrocautery for better hemostasis. The resected area was then closed in three layers with vicryl suture, which reduces the tongue in all dimensions. Manipulation was limited to the dorsal aspect of the tongue than the ventral surface to avoid injury to the lingual nerves, arteries, and hypoglossal nerves.

Central strip glossectomy

In case of hemihypertrophy of the tongue, the incision line was marked by a diathermy, ensuring that the remaining part of the tongue was symmetrical around the midline of the tongue (Fig. 3).

Follow-up

The patients were scheduled for follow-up visits every month in the first year and every year thereafter, during which the following parameters were recorded: disappearance of teeth marks on the edges of the tongue; proper positioning of the tongue within the oral cavity;

improvement or resolution of respiratory disorders; improvement in swallowing and phonation; preservation of taste, heat and pain sensitivity; and improvement in tongue mobility.

Results

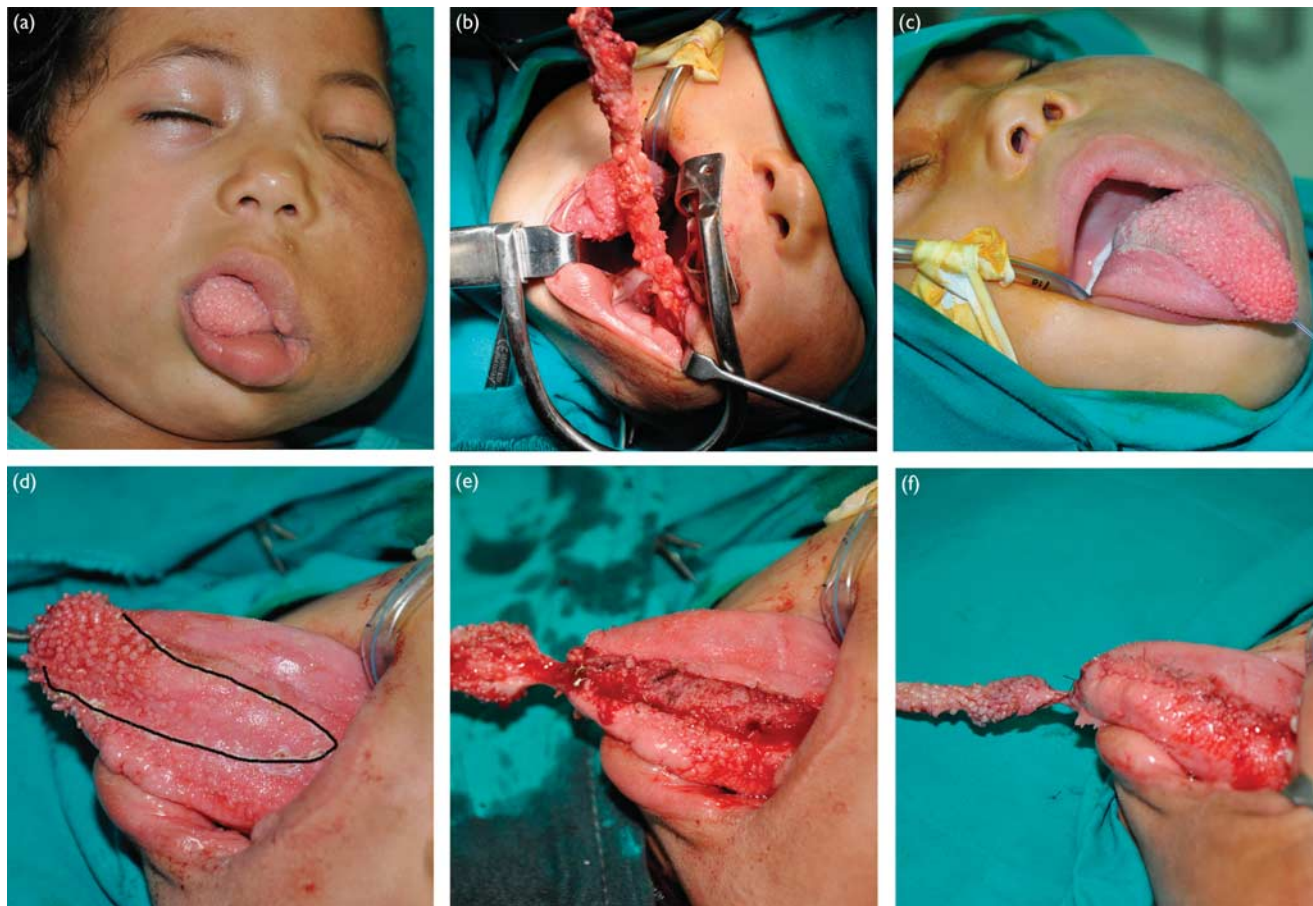
Eight patients' records were retrieved. Three were males and five were females. Their ages ranged from 6 months to 6 years, median 3 years. Three patients had lymphatic malformations, two had hypothyroidism, and three had Beckwith–Wiedemann syndrome.

Clinical examination showed that the tongue was enlarged in width and length, or in all dimensions. There were also disturbances in phonation, malocclusion, recurrent chest infection, drooling of saliva with angular cheilitis, and sleep apnea (Table 1).

Articulation errors were found in bilabial sounds (e.g. Arabic for B, M, that are often produced as lingo-labials), alveolar plosives (e.g. Arabic for t, d), and alveolar fricatives (e.g. Arabic for s, z). Five patients, in whom macroglossia affected the tongue width and length, underwent peripheral glossectomy, whereas two patients, in whom macroglossia affected the tongue in all directions, underwent key-hole glossectomy. In one patient with only half the tongue affected, a central longitudinal strip was excised.

Postoperative complication occurred in one patient who underwent a key-hole glossectomy. Wound infection for 2

Fig. 3



Central strip glossectomy. (a) Preoperative photos of a 4-year-old female patient with lymphangioma of the left side of the cheek and tongue, (b) transoral excision of the lymphangioma of the cheek, (c) markedly enlargement of the left half of the tongue, (d) marking the incision, (e) excision of the central strip, (f) reconstruction of the tongue with symmetry of both halves.

days, followed by partial wound disruption involving the tip of the tongue occurred. The patient was reoperated 3 months later and the disrupted part was repaired after releasing the partial ankylosis that developed during healing.

Motor and sensory functions, especially taste sensation, were not affected by resection. Speech articulation errors were corrected in cases who started speech.

Discussion

The tongue is involved in many functions, including swallowing, phonation, breathing, and normal development of the alveolar process and facial bone structure [22]. Macroglossia compromises airway by obstruction and often leads to obstructive sleep apnea. It hinders growth of the adjacent tissues, leading to an uncoordinated anatomical relationship, causing malocclusion, making speech and mastication problematic. Recurrent upper respiratory tract infection compromises the general health, whereas uncontrolled drooling of saliva predisposes to angular cheilitis and supra-added bacterial/fungal infections. In addition, normal psychological and social well-being and interaction are often affected [1].

The mainstay of surgical treatment of macroglossia is to provide a tongue that can function in the most efficient

manner in terms of form and function [15]. There has been no general agreement on the optimal timing of tongue reduction in infants. Kopriva and Classen [23] recommended that the optimal time for tongue reduction procedures is after 6 months of age, coinciding with a reduction in the rate of tongue growth. Davalbhakta and Lambery [24] recommend deferring the surgical treatment up to the time in which there is a lingual reduced or ceased growth, a condition that occurs between the ages of 15 and 18 years. In this study, we operated patients as young as 6 months old. We did not delay repair because of the important functional limitation caused by macroglossia.

Several procedures have been described to correct macroglossia [1]. As per our experience, we perform peripheral glossectomy [20] in cases where macroglossia affects the length and width of the tongue. However, in cases where the tongue is affected in all dimensions (length, width, and thickness), we perform key-hole glossectomy [21]. A tailored resection is planned according to the affected part of the tongue as in the case of the patient with hemihypertrophy of the tongue in this series.

We did not encounter any intraoperative complication. In the literature, many operative complications have been reported: excessive bleeding, airway obstruction second-

Table 1 Clinical findings

Clinical features	Number of patients
Tongue enlargement in length and width	5
Tongue enlargement in all directions	2
Enlargement of half of the tongue	1
Disturbance in phonation	4
Malocclusion	7
Recurrent chest infection	5
Drooling of saliva with angular cheilitis	3
Sleep apnea	1

ary to tongue edema, lingual nerve injury leading to anesthesia and loss of taste sensation, and hypoglossal nerve injury leading to motor dysfunction [22]. We recommend starting with a series of vertical mattress hemostatic sutures just central to the incision line; such sutures, in addition to traction sutures at the tip of the tongue, minimize blood loss significantly.

Apart from one case of partial wound disruption, we obtained a favorable outcome, with improvement in mouth occlusion and mastication. Speech disorders were corrected. Recurrent upper respiratory tract infections and uncontrolled drooling of saliva were controlled. This is in agreement with many authors in the literature who reported favorable results [25–27].

The impact of partial glossectomy has been studied and improvements in intelligibility [28–30], articulation [31], or phonetic placement of the tongue have been reported [32]. However, Egyedi and Obwegeser published a report on 18 cases. Post operatively, they had 14 patients with decreased movement of the tongue, seven patients who developed speech difficulties, and two patients with anesthesia of the tip of the tongue [33,34].

Conclusion

Partial glossectomy for macroglossia results in minimal complications when performed by an experienced surgeon. All clinical problems caused by the pathology improve after surgery repair. We recommend peripheral glossectomy for cases of macroglossia involving the length and width of the tongue and key-hole glossectomy in cases of macroglossia involving all tongue dimensions.

Acknowledgements

Conflicts of interest

There are no conflicts of interest.

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