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National trends in tongue reduction surgery for macroglossia in children☆,☆☆

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ABSTRACT

Objectives: To examine the frequency of partial glossectomy performed for the indication of macroglossia in children within the United States, assessing for differences in rates of intervention across various demographics.

To identify potential morbidities associated with partial glossectomy in this population and determine how such factors may influence length of stay and cost of admission following tongue reduction surgery.
Study Design: Retrospective cross-sectional study.
Setting: The Kids' Inpatient Database 2003, 2006, 2009, and 2012.
Subjects: Patients under age 5 diagnosed with macroglossia who underwent partial glossectomy.
Methods: Demographics were analyzed and cross tabulations, linear regression modeling, and multivariate analysis were performed.
Results: During the four-years studied, partial glossectomy was performed in 196 children under age 5 with macroglossia. A disproportionately higher rate of intervention was seen in white children ($p = 0.001$), patients undergoing surgery in the mid-west ($p < 0.001$) and patients in the highest socioeconomic quartile ($p = 0.015$). Most patients underwent glossectomy in their second year of life. The average length of stay in patients who underwent partial glossectomy for macroglossia was 9.59 days (Range 1–211 days, median 3.45 days) and the average cost was $56,602 (median $16,330).
Conclusion: Partial glossectomy for macroglossia is typically performed prior to age 2 in the United States. A higher rate of intervention is seen in white children, those who have surgery in the mid-west and affluent children even when controlling for confounding variables.
Level of evidence: III.

1. Introduction

Macroglossia is a condition in which the tongue protrudes beyond the level of the dentition or alveolar ridge in resting position (Prada et al., 2012; Choi et al., 2013). This condition can lead to mandibular growth disturbances, speech and swallowing problems, and open bite deformity. Described by Vogel, true macroglossia is secondary to histologic enlargement of the tongue musculature and may occur in isolation, as manifestations of systemic conditions such as hypothyroidism or amyloidosis, or in the context of a genetic condition such as Beckwith–Wiedemann Syndrome (BWS) (Prada et al., 2012). Relative macroglossia displays no histological change and is often seen in Down syndrome or micrognathia (Choi et al., 2013) (Balaji, 2013). The clinical presentation of BWS is variable, though classically includes macroglossia in 80–99% of patients along with hypoglycemia, omphalocele, hemi-hypertrophy and auricular pitting (Kadouch et al., 2012). The birth prevalence of BWS alone is estimated at 1 in 12,000 or 0.0083% (Cohen, 2005). Due to the variable expressivity and presentation of these patients, the diagnosis of BWS is frequently made well after the neonatal period,

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and the true birth prevalence of this condition may be significantly higher (Kadouch et al., 2012).

The secondary consequences of untreated macroglossia merit consideration. Over time, an enlarged tongue may lead to malocclusion related to open bite deformity or maxillofacial deformities such as prognathism. Over 20% of patients with macroglossia have respiratory difficulties and at least 34% have feeding compromise necessitating nutritional interventions such as feeding tube placement (Prada et al., 2012). A 20-year retrospective review by Kadouch et al. suggested a need for surgical intervention in 43% of patients with macroglossia related to BWS (Kadouch et al., 2012). Partial glossectomy or anterior tongue reduction remains the primary surgical intervention for macroglossia (Chau et al., 2011). However, optimal timing of surgery remains unclear due to variabilities in severity of macroglossia and patient comorbidities (Choi et al., 2013) (Kadouch et al., 2012). Some authors believe timing of intervention should be dictated by severity of symptoms such as airway obstruction, dysphagia, or impaired speech. Others suggest that surgical intervention should be performed prior to age 1 as a means of preventing later functional impairments related to malocclusion and mandibular prognathism (Kadouch et al., 2012).

In 2012, Kadouch et al. reported that 70% of tongue reduction procedures were performed for a number of indications including significant tongue protrusion and associated clinical issues such as feeding problems, drooling, and swallowing difficulties. However, 30% of their procedures were performed for the indication of tongue protrusion alone (Kadouch et al., 2012). Up until this point, the evidence supporting the indications and efficacy of partial glossectomy for macroglossia has been based on a few small case series; the lack of higher level evidence studies has hindered formation of practice guidelines to optimize outcomes in patients with macroglossia.

Understanding diseases on a national scale is imperative for improving outcomes and developing effective treatment algorithms and interventions. The Agency for Healthcare Research and Quality is a division within the United States Department of Health and Human Services that has been collecting patient data for years through the Healthcare Cost and Utilization Project (HCUP). The HCUP has several patient databases, including the Kids' Inpatient Database (KID), which is the only all-payer, pediatric inpatient database in the United States. This database contains information on patient demographics, procedures, charges, and utilization of hospital services by pediatric patients making it suitable for outcomes study of infrequent diseases such as macroglossia (HCUP, 2014b).

The first goal of this study was to query the KID to determine the frequency of partial glossectomy for macroglossia on a national scale and assess for possible variation in rates of intervention based on demographic factors. Additional goals of this study were to assess whether concurrent syndromic features are associated with differences in postoperative care for isolated macroglossia (iMG) and syndromic macroglossia (sMG). We also sought to identify any differences in length of stay and cost of admission following partial glossectomy within these two groups.

2. Materials and methods

We reviewed admissions records from the 2003, 2006, 2009, and 2012 KID, which is part of the HCUP sponsored by the Agency for Healthcare Research and Quality. These datasets contain data drawn from 36, 38, 44 and 44 states, respectively, amounting to over 29 million discharges. By utilizing four KID databases, we sought to account for sample size variation and control for potential variation in overall rates of intervention over time. *International Classification of Diseases, Ninth Revision, Clinical Modification* (*ICD-9-CM*) codes were used for assessment.

To estimate the national frequency of partial glossectomy performed for the indication of macroglossia, the birth prevalence of macroglossia was first established. All newborns with a diagnosis code for macroglossia (ICD-9 750.15) were identified and this number was divided by the total number of newborns in the database. Children with macroglossia were further subdivided into those with and without syndromic features by querying for additional codes associated with known syndromic conditions (Table 1). Patients with relative macroglossia (trisomy 21) and those who underwent tongue reduction for the indication of lymphatic malformation were excluded from the analysis.

A cohort of patients under age five who underwent partial glossectomy for the indication of macroglossia were then identified (*ICD-9-CM* 25.2 and 750.15). The age at which these patients underwent partial glossectomy was determined (Fig. 1), and the total number of patients with macroglossia who underwent partial glossectomy was calculated while accounting for a series of demographics including gender, race, socioeconomic status and geographic region (separated by the regional divisions used by the United States Census Bureau). Estimated differences in rates of intervention across these demographic factors were also determined by dividing the intervention group by the birth prevalence group for each demographic (Table 2). A summary of all *ICD-9-CM* diagnosis and procedure codes used is presented in Table 1.

Length of stay was determined for each surgical admission to identify national trends in the macroglossia population (Fig. 2). Length of stay in addition to total charges was further analyzed to identify factors that may influence these variables such as the presence of syndromic features. The cost of admission was determined by applying cost-to-charge ratios calculated from data reported to the Centers for Medicare and Medicaid Services, thereby providing an estimate of the all-payer inpatient cost-to-charge ratio by hospitals (HCUP, 2014a) (Newhouse et al., 2003).

2.1. Statistical Analysis

Statistical analyses were performed using SAS 9.4 (SAS Institute Inc, Cary, NC). Discharge counts under 11 observations were masked per the HCUP data-use agreement. Data were presented directly and weighted. All mean values were presented with the standard error. Student *t*-tests and z-tests were performed to make pairwise comparisons, and Pearson χ^2 tests were used to test the hypothesis of random distribution of statistical events across multinomial coding. Multivariate analysis was performed using the Fisher Scoring Algorithm. This protocol was reviewed and approved by an institutional review board.

Table 1
The fourteen diagnoses codes that were considered to be associated with syndromic macroglossia (sMG). Patients without any of these diagnoses were classified as isolated macroglossia (iMG).

Syndromic diagnoses	ICD-9 code
Atrioseptal defect	7455
Ventriculoseptal defect	7454
Other congenital anomalies of circulatory system	7470
Persistent fetal circulation (pulmonary hypertension)	74783
Anomalies of pulmonary artery	7473
Other specified anomalies of heart	74689
Tracheoesophageal fistula	7503
Unspecified cleft palate	74900
Hydrocephalus	7423
Reduction deformities of brain	7422
Microcephaly	7421
Down syndrome	7580
Autosomal anomalies	7585
Other specified anomalies	75989