Clinical Perspective

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MACROGLOSSIA: Clinical Considerations

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The clinical appearance characterized by tongue protrusion, lips apart, and mouth open is a familiar triad of findings in the practice of every orofacial myologist. In recent years, much attention has been focused appropriately on airway considerations to explain this triad.

The tongue is the most adaptable organ of the speech and respiratory apparatus and shows an impressive ability to adapt to environmental changes. Airway interference posteriorly often necessitates a forward adaptation of the tongue to maintain an open airway. A tongue thrust swallow may also indicate an adaptation to a reduced faucial isthmus size.

While not all prominent tongues can be linked to airway problems, by like token, not all large-appearing, forward-positioned tongues are pathologically large (macroglossia). The term *macroglossia* is an often used description for a variety of clinical observations that merit consideration by the orofacial myologist.

MACROGLOSSIA

Macroglossia, or pathologic enlargement of the tongue, may be congenital, primary or secondary, and is a more common condition than microglossia, or diminutive tongue. The congenital variety may be due to excessive muscular hypertrophy or hemihypertrophy, and although extremely rare, usually requires surgical correction. Primary macroglossia is typically caused by a systemic etiology (for example, acromegaly due to hyperpituitarism), while secondary involvement is produced by lymphangiomas, hemangiomas, cysts, or tumors.

Macroglossia can cause displacement of the teeth, leading to malocclusion. Posterior and anterior open bites occur with regularity, as well as mandibular prognathism and associated Class III malocclusion. Scalloping of the lateral margins of the tongue is a common finding due to the secondary pressure of the musculature against the teeth, although scalloping, per se, does not automatically signal a macroglossic condition.

Macroglossia often interferes with normal speech patterns and can produce respiratory difficulties, dysphagia, and salivary incontinence. Excessive drying of the mouth due to the lack of lip closure, may result in oral ulcerations and fissuring. Patients with a small oral cavity, due to a dentofacial deformity, may give the appearance of having macroglossia. In these cases the tongue is posturing forward due to the constricted oral environment created by the deformity. Poor muscle tone may also give the appearance of excessive tongue size. The flaccid muscle tone may be of neurological origin or simply a

result of lack of normal posture and use. It is a common clinical observation that a patient's tongue appears to reduce in size after some weeks of therapy exercises directed at normalizing tongue resting posture and improving muscle tone.

Since a "large"-appearing tongue and macroglossia are not always synonymous, differential diagnosis based on a sound data base is necessary to formulate an accurate treatment plan. Surgical correction should only be considered after a thorough correlation of the examination findings collected by the team members.

TONGUE SIZE

Although the tongue is a major focus to linguists, speech/language pathologists, orthodontists and orofacial myologists, it has been subjected to very little scientific study. The limited research data on tongue size may be attributed to the difficulty in evaluating a structure which can vary so much in size and shape. In addition, the tongue is a three-dimensional structure which suffers from the limitations of two-dimensional analysis. Nonetheless, some attempts have been made to evaluate tongue size.

Growth of the Tongue:

The tongue grows fairly steadily and approaches maximum size at or near eight years (Brodie, 1952). Hopkin (1967) measured the dimensions of neonatal and adult tongues post-mortem. 32 neonatal and 30 adult tongues, with approximately equal sex distribution, were studied. The neonatal tongues ranged from birth to 15 days and the adult tongues ranged from 29-85 years. Because the adult tongues were measured in a fresh state and the neonatal tongues were measured after preservation in formalin solution, all neonatal dimensions were increased by 7%. (One neonatal tongue was available in a fresh state and showed 7% shrinkage after 3 months in formalin).

The length of the tongue, measured from the tip of the epiglottis to the apex of the tongue, was 39.9 millimeters in the neonates and 79.8 millimeters in the adults.

The breadth of the tongue was measured at the widest part of the tongue. This measured 25.4 millimeters in the neonates and 51.9 millimeters in the adults.

Finally, the thickness of the tongue was measured at the free edge of the tongue at its widest part. This measured 8.7 millimeters in the neonates and 16.1 millimeters in the adults.

Thus, all the dimensions of the tongue appeared to

double in size by adulthood. While male tongues were slightly larger than female tongues, there was no statistically significant difference. There was a width variation in the size of the tongue within the neonatal group and within the adult group. Although there were no bony measures taken, a previous study by Hopkin (1963) demonstrated no relationship between the height of the tongue and the width of the upper arch.

Relative Size of the Tongue:

Although absolute measures of tongue size are of some importance, it is more relevant to study the tongue within the confines of the oral cavity. Vig and Cohen (1974) undertook a study to compare the size of the tongue with the maxillary space in both children and adults. They used 75 children who had been referred for orthodontic consultation and compared them to 26 adults who had been previously studied.

Lateral skull x-rays with the teeth in occlusion were used. Structures necessary to define the confines of the intermaxillary space and the tongue were defined and traced. A planimeter was used to estimate the area of the tongue shadow and intermaxillary space.

The children in this study averaged 9.9 years of age, with a range of 5 years. The adults averaged 28.2 years of age, with a range of 10 years.

The ratio of the tongue area to the intermaxillary space in the child sample was 0.74. The ratio in the adult sample was 0.67. These data suggest that the tongue becomes relatively smaller when compared with the intermaxillary space, at least in this age group. The authors (1974) speculate that this difference is due to the differential rates of maturation of the skeleto-dental and muscular element, and partly due to the descent of the tongue and associated structures which occurs with growth of the cervical spine.

Childhood is also the time when the lymphatics demonstrate themselves most fully. The tonsils, in particular, compete for space in the posterior oral cavity. Considering the oral-facial complex in total, it is easy to see why some children with a relatively large tongue in proportion to the dimensions of the intermaxillary space, and large tonsils, will present with a chronic tongue-forward, mouth-open posture. This situation is not macroglossia, nor is it necessarily mouth breathing. The clinician should evaluate and should initiate therapy with reference to this time frame of the growth and development of the oral-facial complex.

The Tongue in Down Syndrome:

The child with Down syndrome (Figure 1) typically presents with a "relative" macroglossia. The term "relative macroglossia" was well discussed by Subtelny (1970), who pointed out that the morphology in Down syndrome finds a normal-size tongue in a small oral cavity due to maxillary retrusion and a restricted nasopharynx; hence, the tongue is macroglossic only in relation to reduced size or dimensions of adjacent structures or spaces. Nonetheless, a true macroglossia is not present.

The facial posture in Down syndrome often includes an habitual open-mouth posture with protruding tongue.

One other report (Oster, 1973) suggests that 60% of Down syndrome children have an "overly large or overly long tongue."

Figure 1

Patient with Down Syndrome and prominent, forward resting posture of the tongue. This is not a true macroglossia, but can be considered a "relative macroglossia." See text for discussion.

In addition to the aesthetics of the chronic open-mouth, tongue-protruded posture, a relative macroglossia may also result in difficulties in chewing, drinking and in speech articulation. Early medical, educational and therapy intervention is becoming more widely available for children with this syndrome. Parents, therapists and educational personnel can use behavior management techniques to improve the child's nasal hygiene strategies, lip closure and tongue posture. Oral muscle exercises and facilitating techniques can be used to improve tongue and lip muscle tone, posture and movement patterns during speech, chewing and swallowing. Children are referred for medical diagnosis and management when nasal congestion is a persistent problem.

Another reported treatment for the relative macroglossia is a partial tongue resection. Olbrisch (1982), a surgeon, reported on an oblique wedge resection carried out on 202 Down syndrome children. The resection preserved the nerves and blood vessels in an effort to protect the mobility of the tongue. Complications included breakdown of the tongue repair in 11 cases, which required resuturing.

Post-operative results on 102 of the children, at one year or more follow-up, are anecdotal. The reported results include increased mouth closed posture, fewer upper respiratory infections, increased nasal breathing, improved eating and speech and improved parental attitudes towards the children.

The improved aesthetics have also been demonstrated in an additional study. Klaiman, et al. (1987), studied speech as well as the speech aesthetics in 8 patients with Down syndrome, pre-operatively and at six months post-surgical resection of the tongue. While the aesthetic appearance of speech was judged to be improved in 7 of 8 patients, there was no difference in the acoustic production of speech. Currently, there is diminished inter-

national interest in tongue resections as a positive treatment mode for most patients with Down syndrome.

CLASSIFICATION OF MACROGLOSSIA

Macroglossia may essentially be classified as primary or secondary.

Primary

Primary macroglossia may result from a variety of diseases. These include hyperthyroidism, amyloidosis, cretinism, glycogen storage disease, and idiopathic hyperplasia. Beckwith's hypoglycemic syndrome, which includes macroglossia, also consists of neonatal hypoglycemia, mild microcephaly, umbilical hernia, fetal visceromegaly, and postnatal somatic giantism. In this syndrome, macroglossia is more of a symptom than a cause. Treatment is directed at the primary cause in order to correct the macroglossia.

Idiopathic lingual hypertrophy, although very uncommon, has been classified by Shafer (1968) into three categories: generalized hypertrophy without muscular hypertrophy; diffuse hypertrophy associated with generalized muscular hypertrophy; and unilateral hypertrophy of the tongue associated with ipsilateral or contralateral facial or body hypertrophy. Most patients are female and, in the first category or classification of Shafer, anomalies such as umbilical hernias and omphaloceles are found frequently. Biopsy specimens from these patients demonstrate an increase in the amount of cytoplasmic content in each muscle fiber and a 50% increase in the number of nuclei.

Secondary

Secondary macroglossia may be divided into five groups with the most common type being the lymphangiomatous variety. These lesions have been classified into four groups: (1) hygroma (cystic lymphangioma); (2) cavernous lymphangiomata; (3) capillary lymphangiomata; and (4) hemangiolymphangiomata (Figure II). The disease may be localized (only involving the anterior two-thirds of the tongue) or more diffuse, such as the cystic hygroma. Respiratory difficulties (airway obstruction) may arise with the diffuse variety, but are usually not encountered in localized disease. Surgical intervention is usually indicated since spontaneous regression is not common. Partial glossectomy is the treatment of choice in patients with localized disease, while in more diffuse cases extensive dissection and *en bloc* resection may be required.

Cystic lesions commonly result from aberrant tissue growth either embryologically or developmentally. Thyroglossal duct cysts are due to the persistence of all or part of the thyroglossal duct resulting in a cyst. The most common locations are the infrahyoid region and in the midline, although these lesions may also occur at the base of the tongue. Surgical excision of the cystic lesion and duct remnant is the treatment of choice.

The ranula is a mucous retention phenomenon which occurs in association with the ducts of the submaxillary or sublingual gland. This lesion develops as a slowly enlarging mass in the floor of the mouth. It is important

that this lesion be differentiated from a cystic hygroma which frequently involves the neck. Surgical treatment of the ranula should be directed at excision if the lesion is small, or at marsupialization (opening the cyst and emptying its contents) if it is extensive.

FIGURE II

Macroglossia due to hemangiolymphangioma of the tongue. Note the discolored area of the lesion. This patient cannot chew without biting the lateral margins of the tongue. This is a typical macroglossia associated with a space-occupying mass of the tongue. See text for discussion.

Dermoid cysts can produce a secondary type of macroglossia which is developmental in origin and occurs in the midline. The cyst is usually encountered in the floor of the mouth. However, it can protrude inferiorly, producing an extraoral swelling. Treatment of the lesion is surgical incision.

Solid lesions such as the lingual thyroid nodule, neurofibromatosis, granular cell tumors, and malignant tumors may also produce secondary macroglossia. The lingual thyroid gland tissue contained in the lingual thyroid nodule may be the patient's only functioning thyroid tissue. Diagnosis is confirmed by a radioactive I-131 scan. The treatment of choice is usually surgical excision. However, the presence of thyroid gland tissue should be determined or the patient must be placed on thyroid hormone therapy.

Neural tumors may occur anywhere in the oral cavity, but the tongue is the most frequent site. They are benign neoplasms which surround nerves and appear as smooth nodular lesions. Such tumors are usually unilateral and, if the lingual nerve is involved, can be extensive. Differential diagnosis from other cysts and tumors is essential and the treatment is surgical resection.

Granular cell lesions are tumors which are composed of dense granular cytoplasm. The most common site is the dorsum and anterior border of the tongue, but rarely do these tumors cause macroglossia. Treatment is local excision and recurrence is unusual.

Malignant tumors, when present on the tongue or the floor of the mouth, can produce macroglossia. The differential diagnosis is confirmed by biopsy and the patient is referred to an oncologist for treatment (Mason and Serafin, 1984).

DISCUSSION

The condition of macroglossia most often implies some space-occupying mass of the tongue that necessitates surgical excision or resection. In other instances, the macroglossia is a symptom or consequence of systemic disease.

For some patients, whose orofacial morphology creates the physiologic need for the tongue to protrude as a means of maintaining the airway, the tongue can appear pathologically enlarged either in comparison with adjacent structures (as in the relative macroglossia of Down syndrome), or due to systemic conditions which may contribute to the clinical judgment of tongue enlargement.

From a clinical examination standpoint, the term *macroglossia* should be considered a medical diagnosis to be avoided in orofacial examination report findings, unless established previously in a medical team setting. It would be rare to find a true idiopathic, or non-pathologic, macroglossia in clinical work in orofacial myology or speech and language treatment.

A simple clinical test for suspected macroglossia is to request that the patient bite on the back teeth. If the teeth are able to contact without biting the tongue, it would be difficult to make a case for idiopathic macroglossia.

Any suspicions of macroglossia should be accom-

panied by a thorough examination of the tongue. Coloration, masses, and changes in consistency of tissue should be recorded. Scalloping of the lateral margins of the tongue, conforming to the adjacent shape of the teeth, is quite prevalent in the normal population and does not signal a macroglossic condition.

The variety of airway interference factors well known to the orofacial myologist may necessitate or encourage a tongue-forward, lips-apart, mouth-open posture. This triad of findings may erroneously imply either mouth-breathing, or macroglossia, or both. Most often, it is clinically prudent to avoid labelling until objective assessment or verification can be made.

SUMMARY:

Macroglossia is a multifactorial condition that is almost always associated with a space-occupying mass of the tongue. The usual treatment is surgical resection.

The typical medical conditions that lead to macroglossia have been described. The orofacial myologist should be alert to the presence of possible pathology of the tongue during orofacial examination, and refer suspected instances of macroglossia to an appropriate medical resource for definitive diagnosis and treatment.

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