MACROGLOSSIA
Warren E. Morgan, MD
May 28, 1992

Macroglossia is tongue enlargement that leads to functional and cosmetic problems. Although this is a relatively uncommon disorder, it may cause significant morbidity. There is no clear definition of macroglossia and it may be defined in relative, functional, or structural terms. Clinical studies are limited by this lack of a clear definition.

Normal speech and swallowing require normal tongue anatomy and function. Swallowing begins as the tongue mixes food with saliva to form a food bolus, which is then propelled into the pharynx by the tongue. Articulation also depends on the tongue's ability to alter the impedance of air, and change the resonant characteristics of the upper airway. In macroglossia, increased tongue bulk may impair these functions.

Differential growth of the tongue and other facial structures affects tongue position in infants and children. The tongue growth is similar to neural tissue, with the majority of growth occurring early. Maxillary growth is slower, but similar to the tongue. The mandible grows more slowly, with two peaks of growth. One occurs between the ages of 8-12 and the second growth phase occurs with puberty. This differential of growth leads to the tongue having a high and forward position in the early years which may accentuate macroglossia.
Macroglossia may be classified in several ways. A classification by Meyer et al divides the causes into generalized and localized based on the extent of tongue involvement. The localized causes can be broken down into four different categories. These include congenital, inflammatory, traumatic, and neoplastic lesions. The congenital causes include hemangioma, lymphangioma and lingual thyroid. Inflammatory causes include tuberculosis, actinomycosis, dental infection, syphilitic gumma, Riga disease, ranula, and sublingual calculus. Traumatic causes include dental irritation, hematoma, and postoperative edema. The neoplastic causes can be divided into malignant and benign lesions, with the malignant lesions including carcinoma and sarcoma. The benign lesions include granular cell tumor, neurofibroma, leiomyoma, and lipoma.

Generalized macroglossia can also be broken down into four groups. These are the congenital, inflammatory, traumatic, and metabolic causes. An inflammatory cause is chronic glossitis. Traumatic causes include postoperative edema. Metabolic causes are myxedema, amyloidosis, lipoid proteinosis, chronic steroid therapy, and acromegaly. The congenital causes are primary idiopathic macroglossia, cretinism, hemangioma, lymphangioma, Robinow Syndrome, Beckwith-Wiedmann Syndrome, Down Syndrome, Trisomy 4P Syndrome, Triploid Syndrome, generalized gangliosidosis syndrome, and any of the Mucopolysaccharidoses.

In several series the most common cause of macroglossia is lymphangioma. These are relatively rare congenital tumors of malformed lymphatic tissue that most commonly involve the tongue. The involvement may be partial or complete. These tumors typically present within the first year of life and are slow growing and progressive. Trauma or infection results in increased lymph production and an increase in tumor size. Subsequently, fibrosis may occur leading to more lymph obstruction or atrophy. About 7% of lymphangiomas have associated lymphatic malformation in the neck. Microscopic exam shows enlarged lymphatic channels interdigitating with normal muscle cells.

Beckwith-Wiedmann Syndrome is another common cause of macroglossia. This syndrome encompasses several congenital anomalies. The exact incidence is unknown, but has been estimated at .07 per 1000 births. The mode of inheritance is unknown with sporadic and familial cases being reported. The most common clinical findings are macroglossia, omphalocele, and postnatal gigantism. These patients also have characteristic craniofacial abnormalities of maxillary hypoplasia and a prominence of the occiput. For unknown reasons, these patients have an increased incidence of Wilm's Tumor, adrenal carcinoma, or hepatoblastoma. Another problem is hypoglycemia which causes seizures, mental retardation, and even death.

Macroglossia is a common but not obligatory manifestation of Beckwith-Wiedmann Syndrome. The degree of macroglossia varies with each patient. The enlargement is generalized and may cause a variety of difficulties with speech and feeding, as well as airway problems. The macroglossia is most prominent in infants but tongue size may remain above normal in childhood and adolescence. As the hyoid descends with age, the macroglossia may improve. Histologic examination shows hypertrophy and hyperplasia of muscle cells.

Another common cause of macroglossia is Down Syndrome. Down Syndrome is the most common genetic disorder in man, occurring in about 1 out of 700 live births. The
incidence increases with maternal age. The syndrome results from nondisjunction and 
the affected individual has an extra chromosome 21. These patients have numerous 
characteristic physical findings and mental deficiency. The six characteristic facial traits 
include: 1) epicanthus, 2) oblique lid axis, 3) saddle nose, 4) macroglossia, 5) 
hypotonia, and 6)microgenia. The macroglossia is accentuated by their general 
hypotonia, causing some authors to refer to this as a relative or pseudomacroglossia. 
The histology shows hyperplasia and hypertrophy similar to Beckwith-Wiedmann 
Syndrome.

Macroglossia causes a variety of signs and symptoms. These include tongue protrusion, 
which exposes the tongue to trauma. This exposure also leads to mucosal drying and 
recurrent upper respiratory tract infections. Other symptoms include speech 
impediment, swallowing difficulties, airway obstruction, drooling, and failure to thrive.

Evaluation of macroglossia begins with a careful history and physical exam to identify 
any 
undiagnosed syndrome. Laboratory tests are guided by clinical suspicions. Thyroid 
function tests are used to rule out hypothyroidism. Thyroid scan may identify a lingual 
thyroid. Patients with chronic airway obstruction may need CXR and EKG to evaluate 
right heart failure. Treatment options include observation, orofacial therapy, and 
surgery. Patients that have minimal symptoms may be observed since changes in tongue 
position may improve the disorder.

Orofacial therapy uses a palatal device to stimulate muscular tone and proper tongue 
position. This is used in patients with Down Syndrome to counteract their hypotonia 
and improve their tongue position. Limbrock et al reported their results using orofacial 
therapy in a series of 89 patients with Down Syndrome. 25% of patients were unable to 
complete the therapy due to lack of compliance, improper fitting and loss of follow-up. 
Tongue protrusion improved in 63% and drooling improved in 68%.

The majority of cases of macroglossia are treated surgically. Indications for surgery 
include airway obstruction, speech difficulties, dysphagia, and cosmetics. The 
procedure of choice is partial glossectomy. Surgical goals are to reduce the tongue size 
and produce improved function. Good airway management is important in the 
postoperative period. For small children and infants this may be done with tracheotomy, 
and in older children prolonged intubation may be adequate.

Different authors have advocated a number of different approaches for tongue 
reduction. Rather than using one approach, each case should be evaluated and the 
appropriate surgical approach chosen. Different resections provide reduction in different 
directions. Regardless of the approach taken the initial resection should be conservative 
to prevent permanent problems from overly aggressive resection. The technique used is 
an anterior wedge resection with posterior key hole. The lateral incisions are beveled 
out to decrease tongue bulk. The tongue is then closed in a T.

Surgical results have been good. In a series of nine patients with macroglossia, Siddiqui 
and Pensler reported good results. The patients in their series were equally divided into 
those with Down Syndrome, Beckwith-Wiedmann Syndrome, and Lymphangioma. 
62% had improvement in speech. Drooling and deglutition difficulties improved in
80%. No major complications occurred, but patients with lymphatic malformations did significantly worse than the other groups.

Olbrisch reported his results with tongue reduction in 300 patients with Down Syndrome. After surgical treatment, complete mouth closure was present in 60% and another 25% could close their mouths during the day. Airway diseases were reduced by 83% and speech was improved in 45%. Revision operations were required in 6% of patients. The complications were minor, with two patients experiencing minor bleeding postoperatively.

Macroglossia results from a variety of disorders, and may affect cosmetics and function. The main treatment is partial glossectomy, but the initial treatment should be conservative. Successful treatment require appropriate rehabilitation and long term follow up.

Case Presentation

The patient was noted, shortly after birth, to have an extensive lymphatic malformation of the left neck, tongue and epiglottis. She was otherwise healthy, with no airway or feeding problems. At 10 months she underwent excision of the lymphatic malformation of the neck. She tolerated the procedure well and recovered uneventfully. Although there were no functional problems, her tongue was thickened and protruded from her mouth. At 21 months she underwent tracheotomy and partial glossectomy. She tolerated these procedures well. Several weeks later the supraglottic component of the lymphatic malformation was treated with the laser vaporization. There was improvement following the initial tongue resection, however there was still tongue protrusion noted at various times during the day. Due to cosmetic concerns she then underwent repeat tongue reduction at 24 months. She tolerated this procedure well and has no tongue protrusion with good closure of the mouth at one-month follow-up.

Bibliography


[Return Grand Rounds Archive Index](#)

[Return to BCOM Otolaryngology Home Page](#)