Review Article

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HYPODEVELOPMENT OF THE TONGUE: EMBRYONIC BASIS AND CONSEQUENCES FOR THE DEVELOPMENT OF CRANIOFACIAL STRUCTURES

Tereza Guida Paola Genovez[1], Juliana Silvério Campanati Mendonça[2], Janete MB Galante[3], Ana Lúcia Pompéia Fraga de Almeida[4], Marcia R Gomide[5], Gisele S Dalben[6]

1. Department of Pediatric Dentistry, Hospital for Rehabilitation of Craniofacial Anomalies, University of São Paulo
2. Department of Pediatric Dentistry, Hospital for Rehabilitation of Craniofacial Anomalies, University of São Paulo
3. Department of Pediatric Dentistry, Hospital for Rehabilitation of Craniofacial Anomalies, University of São Paulo
4. Department of Prosthodontics, Bauru School of Dentistry, University of São Paulo
5. Department of Pediatric Dentistry, Hospital for Rehabilitation of Craniofacial Anomalies, University of São Paulo
6. Department of Pediatric Dentistry, Hospital for Rehabilitation of Craniofacial Anomalies, University of São Paulo

ABSTRACT:

The present paper describes the embryogenesis of the tongue, as well as possible factors that interfere with its development. Changes caused by hypodevelopment of the tongue (hypoglossia and aglossia) and resulting alterations on the craniofacial complex are also discussed. Even though the occurrence and case reports of these alterations are scarce, knowledge on this issue is of special interest to dental professionals, since the tongue plays a fundamental role during the development of craniofacial structures.

Keywords: Aglossia,Hypoglossia,Tongue Hypoplasia,Tongue Hypogenesis,Tongue Alterations.

INTRODUCTION:

Examination of the tongue is very important in oral diagnosis and should comprise assessment of several aspects, including the size, shape, mobility and symmetry, since this muscle plays a very important role in the development of craniofacial structures.

Hypodevelopment of the tongue is a rare occurrence and may present as hypoglossia, also named as microglossia, which is manifested as a small or rudimentary tongue, typically with absence of the two anterior thirds of the tongue, with persistence of only the posterior third. [1, 2] Some individuals may also present aglossia, which is the complete absence of the tongue at birth. [3- 8]

EMBRYONIC DEVELOPMENT OF THE TONGUE

From an embryonic standpoint, formation of the tongue is initiated at the fourth week at the pharyngeal floor, from the ventral portion of pharyngeal arches (I, II, III, and IV) at the midline below the primitive mouth. Mesenchymal proliferation gives rise to several prominences on the mouth floor. Initially, one prominence (the median lingual swelling) appears at the midline of the mandibular process; later, it is surrounded by other prominences, the lateral lingual swellings. These prominences quickly increase in volume and fuse with the median lingual swelling forming a large bulk, which gives rise to the mucous membrane of

*Corresponding Author Address: Dr Marcia Ribeiro Gomide Email: marcinha@usp.br

the two anterior thirds of the tongue; thus, in summary, this part of the tongue appears by mesenchymal proliferation of the first pharyngeal arch (mandibular).

The posterior third of the tongue, or lining mucosa of the tongue root, is formed from the hypopharyngeal eminence, a large prominence at the midline originated from the mesenchyme of the third arch, which quickly covers the second arch, thus precluding its subsequent participation in the development of the tongue. Some specialists divide this eminence in an anterior copula (which gives rise to the lining mucosa of the tongue root) and a hypopharyngeal eminence (which gives rise to the epiglottis).

The tongue is separated from the mouth floor by downward ectodermal growth at its periphery, which subsequently degenerates forming the lingual groove, which allows tongue mobility. The tongue muscles have a different origin: they are originated from the occipital somites, which migrate forward at the tongue area, accompanied by their nerve supply, the 12th cranial nerve (hypoglossal). [9]

This complex tongue development explains its innervation. Since the mucosa on the two anterior thirds of the tongue is derived from the first arch, its innervation is supplied by the 5th cranial nerve (trigeminal), whereas innervation of the posterior third is supplied by the 9th cranial nerve (glossopharyngeal), since it is derived from the third pharyngeal arch; as previously mentioned, the motor innervation to tongue muscles is supplied by the 12th cranial nerve. [9]

In the fetus and neonate, the lingual artery is a major vessel, larger than the anterior or middle vertebral arteries. The external carotid, which eventually supplies the tongue and mandible through the lingual artery, develops from the third arch. Before appearance of the external carotid, at about five weeks, the first arch is supplied by the stapedial artery, a temporary vessel that arises from the dorsal aorta. Injury to this artery or its premature involution can interfere with the vascular supply to the area and is believed to be responsible for several craniofacial malformations, especially the so-called first arch syndromes, which comprise anomalies related to abnormal development of the first arch [10]. Thus, the presence of hypoglossia can be explained by a failure of normal development of the first arch at or before the fourth embryonic week [2]. Weckx *et al* [11], in their most reasonable assumption, indicate a teratogenic factor that occurs in the fourth week of embryonic life, which is the period of initial development of oral structures. Other authors [12, 13] suggest that the etiology of congenital hypoglossia include chromosomal aberrations (trisomy I) and other facial dysmorphology syndromes.

CLINICAL MANIFESTATIONS

As with all malformations, internal and external factors during fetal development play an important role in the occurrence of tongue malformations.[5] According to Hernández *et al*. [1], there is no gender predilection or genetic implications in the occurrence of aglossia and hypoglossia. Their etiology should be investigated focusing on some types of fetal aggression at the first weeks of intrauterine life, such as teratogenic drugs (corticoids, tranquilizers, etc.), intoxications, radiations, viral infections, undernourishment, emotional stress, etc., even though the pathogenesis of these alterations remains undefined.

Hypodevelopment of the tongue may occur in isolation [1,5,11,14] or associated with other anomalies, especially limb defects, or associated with syndromes [10,15,16,17,18 ,19]

Most cases are part of the aglossia-adactyly spectrum [10,15,16] and the oromandibular-limb hypogenesis syndrome (Hanhart's syndrome). [17-19] Tongue changes have also been related to the agnathia-holoprosencephaly spectrum [20], situs inversus [2,21] and cleft palate (Fig. 1) [22] and also associated with disorders as the Moebius syndrome and Robin sequence[7,23] and hypothyroidism. [23]

The earliest report on congenital aglossia is ascribed to Jussieu in 1718 [24], and the first literature review describing an aglossia-adactyly case was published by Rosenthal in 1932. [25] The publication of Weckx *et al*. in 1990 [11] discussed the 47 cases of aglossia-adactyly syndrome reported in the available literature. Fewer cases of isolated hypoglossia or aglossia have been reported [5, 6, 25, 26]. Hypodevelopment of the tongue may lead to imbalance in facial structures.

Since the function is directly related to the shape, all functions of the stomatognathic system may me impaired since, the greater the deformity, the greater will be the functional damage. Total or partial absence of the tongue is usually followed by palate atresia (Fig. 2), lower lip defects (Fig. 3), mandibular hypoplasia (Fig. 4) [1, 5, 11, 19, 22, 23, 27] and poor transverse development of the alveolar arches due to the lack of muscle stimulation; the alveolar arches remain small and narrow, leading to severe tooth crowding (Fig. 5). There may also be hypodevelopment of the lower and medium thirds and of the chin (Fig. 6), besides tooth agenesis, especially of mandibular incisors (Fig. 7). [1]

CLINICAL MANAGEMENT

With regard to the treatment of occlusal changes, some authors suggest mandibular expansion or distraction osteogenesis with retention. [1, 7, 28] Yamada et al [29] performed bone lengthening for mandibular hypoplasia and orthodontic treatment with satisfactory outcomes in three cases of micrognathia and hypoglossia. Even

though Ribeiro *et al*. [22] suggested therapeutic planning with evaluation of the mouth floor tissue available for reconstruction of a tongue-like organ, after 18 months of age, a later report by Weckx *et al*. [11] addresses the impracticality of a direct surgical approach to correct malformations of the tongue in patients with aglossia, suggesting a multidisciplinary therapeutic team approach, i.e. supportive therapy, including phoniatry and orthodontic treatment. The phoniatric therapy is also intended to stimulate or facilitate the functions of mastication, swallowing, and speech. Absence of the tongue has significant influence on the transverse growth of the mandible and maxilla. This contributes to the lack of space available for the eruption of permanent teeth. Orthodontic treatment should be initiated as soon as the patient can safely accept, so that any dental malocclusion can be corrected and the space decreased between the palate and the floor of the mouth.

The accumulation of food secondary to hypoglossia represents a very serious problem for proper oral hygiene. [11] A high caries rate with a rudimentary tongue has been previously reported. [5,23] The causes of high caries index could be lack of self-cleansing by the tongue or composition of the saliva, which might predispose to dental caries [8, 23] (Figs. 5 and 8). These patients should also receive preventive dental care.

Individuals with hypoglossia may also present with glossoptosis, due to posterior sliding from the region of insertion of tongue muscles, which are usually hypoplastic (Figs. 7A and 9). These defects often lead to severe breathing difficulties. [1]

Since tongue morphogenesis is integrally linked to the normal development of the thyroid gland, an abnormal tongue morphogenesis could potentially be associated with functional thyroid disorders. [23]

Due to these relevant orofacial changes, Perko in 1972 [30] mentioned that, since feeding is not possible without the suckling reflex (due to absence of the tongue), the survival of affected individuals would be threatened. However, the cases reported by Goto *et al*. [14] and Khalil *et al*. [5] managed to cope with the condition and were able to distinguish various tastes since a young age, despite the presence of only a rudimentary tongue. Their speech was surprisingly understandable, although they could not normally pronounce the consonants that are formed with the tongue tip.

While being fed, a normal infant thrusts the tip of his or her tongue onto the upper gum to obstruct the mother’s nipple or bottle against the hard palate, thus restraining the ingestion of fluid. At the swallowing phase, the tongue drives both the solid bolus and fluid into the oropharynx. At the same time, contraction of the soft palate occludes

the nasopharynx from the fluid bolus. [31] Conversely, the infant affected by hypoglossia cannot satisfactorily control the ingestion of fluid. Such children tend to have hypertrophy of structures in the mouth floor. The affected child then attempts to compensate for the deficient tongue by upward and downward thrusts of the vestigial tongue against the hard palate. [11]

Despite the significant anatomical changes, in some cases the speech [1, 5, 18, 32] and swallowing are not much affected by this condition [10, 19, 24, 33], due to elevation of the mouth floor. Individuals with hypodevelopment of the tongue may have clear speech and their swallowing may be adapted and regarded as normal [11] ; they may also have taste perception of foods. [5,34]

CONCLUSION:

Knowledge on the possible etiology and orofacial changes caused by aglossia and hypoglossia are fundamental in dental clinics, so that dental professionals may properly act in the treatment and follow-up of oral health of individuals affected by these malformations.

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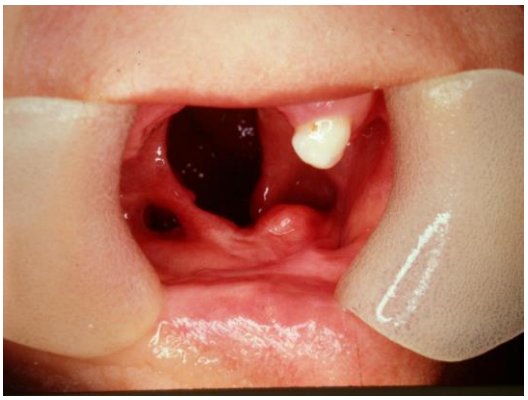
FIGURES:



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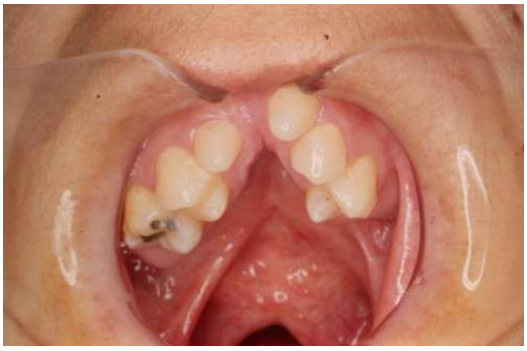
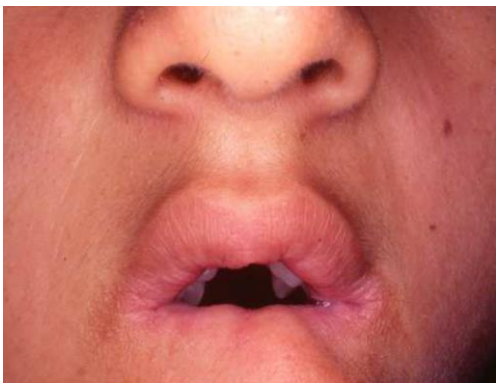


Figure 2



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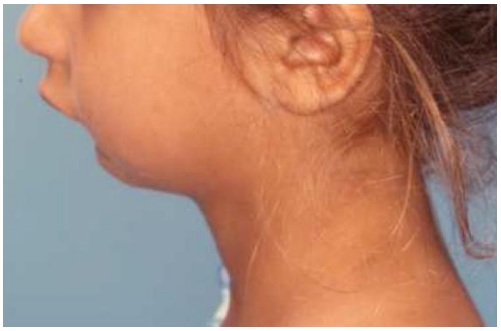
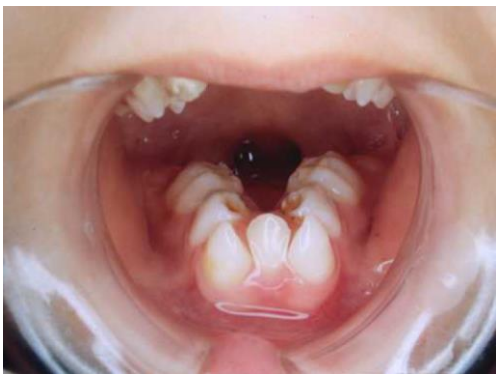


Figure 4



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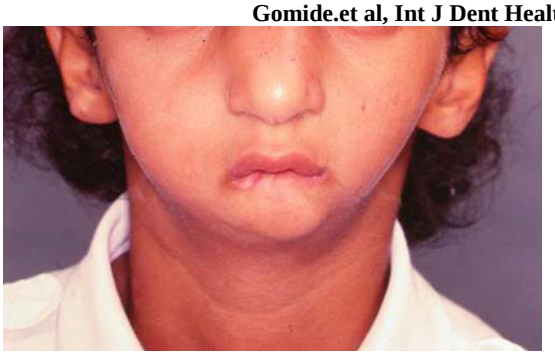


Figure 6A

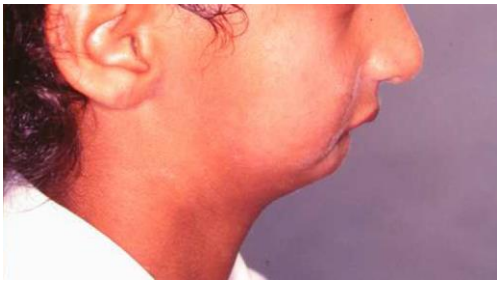
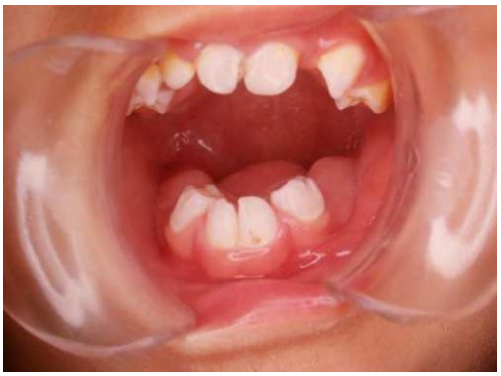


Figure 6B



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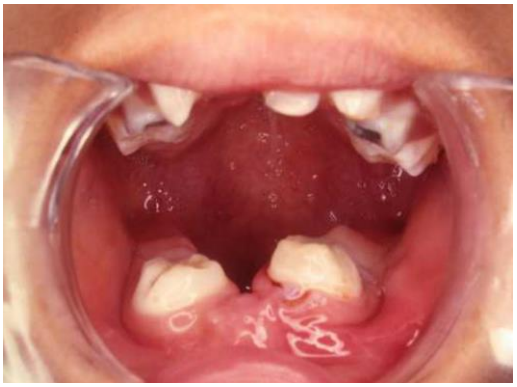

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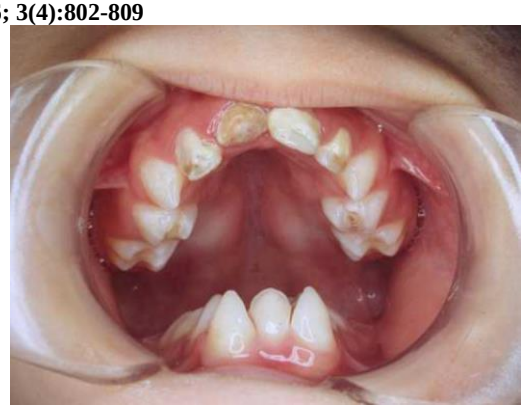

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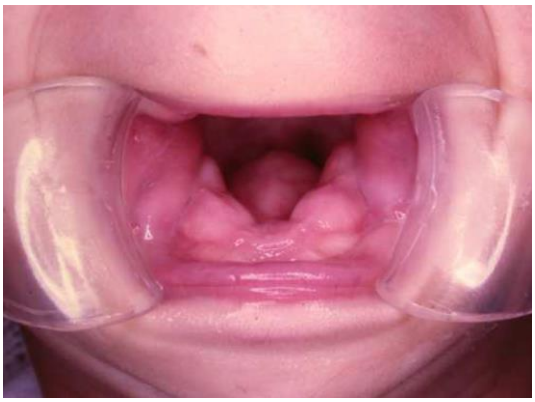

Figure 9