This book provides detailed description of disorders occurring during the development of tongue. The embryology, anatomy and physiology of tongue are also discussed in depth. Each disorder is explained very systematically under different subheadings such as introduction, etiology, clinical features, histopathologic features, differential diagnosis and management. The book provides extensive classifications, original clinical and histoplathology pictures which are easy to comprehend and relate to. The language and style of writing is very simple and easy to understand. This book is written with the undergraduate and post graduate medical/ dental students in mind, at the same time, it also provides comprehensive information on the developmental disturbances of tongue to any interested reader.

Dr. Sangeetha Ramu MDS Oral Pathology, Rajiv Gandhi University of Health Sciences. Currently working as Reader in the Department of Oral Pathology & Microbiology at Dayananda Sagar Institute of Dental Sciences, Bengaluru, India. She has been an academic achiever with more than 15 publications to her credit.

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Ramu, Bidadi Rajashekaraiah, Chandavarkar

Sangeetha Ramu
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A Comprehensive Guide

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Sangeetha Ramu

Premalatha B.R

Vidyadevi Chandavarkar

DEVELOPMENTAL DISTURBANCES OF TONGUE- A COMPREHENSIVE GUIDE

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AUTHORS

1. Dr. Sangeetha Ramu

Reader, Department of Oral Pathology and Microbiology

Dayananda Sagar College of Dental Sciences, Bangalore, India

Email: sangeethasrikanth04@gmail.com,

Phone no: +91 7760400400

2. Dr. Premalatha B.R

Reader, Department of Oral Pathology and Microbiology

JSS Dental College and Hospital,

JSS University, Mysuru, India

Email: prema.raaj@gmail.com,

Phone no: +91 9900281290

3. Dr. Vidya Chandavarkar

Reader, Department of Oral Pathology and Microbiology

School of Dental Sciences,

Sharda University

Greater Noida, India

Email: svidyac27@yahoo.co.in,

Phone no: +91 8800978814

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Dr. Sangeetha Ramu

INTRODUCTION

The tongue is the most agile, adept, and versatile appendage in the human body. It is essential for mastication, deglutition, taste, and speech.[1]The tongue is a highly specialized muscular organ occupying a large part of oral cavity that contributes to speech and complex sense of taste. The word tongue is derived from the Latin word 'Lingua' and Greek word 'Glossa'.

It is essential for mastication, deglutition, phonation, and gustatory function. In congenital absence of tongue survival is impossible without active support since suckling is impossible without the tongue. The tongue together with the cheek also plays an active role in mastication by keeping the bolus between the maxillary and mandibular teeth by preventing the food from falling into the sulcus. The subject of taste is of interest to most human beings either for its value in survival or in the appreciation of food and drink. But is particularly important to the dentist because taste is the main stimulant to saliva flow and the stimulation of saliva is important in oral health. [2]

The tongue occupies a large part of the oral cavity and can be affected by numerous lesions, either limited to the tongue or as a part of oral diseases or as a sign of systemic disorders. Thus the tongue is also called as a "mirror" of the human body.

Few local lesions have typical manifestation only on tongue, which help in definitive diagnosis, but there are many systemic diseases which have secondary manifestations on the tongue. Such manifestation on the tongue is useful as adjuncts for the diagnosis of the systemic diseases. They should be observed and recognized which can prompt the dentist to refer the case to one's medical colleague.

The main aim of this book is to enlighten the reader about the congenital anomalies of the tongue, since congenital tongue anomalies of tongue can occur as isolated events or as a part of clinical syndromes. Aglossia, syndromic microglossia, macroglossia, accessory tongue, long tongue, and cleft or bifid tongue are few of the congenital anomalies seen in relation to tongue. This book gives further insight to these and other congenital anomalies of the tongue.

CLASSIFICATION OF TONGUE DISEASES

1. **Congenital and developmental disorders**
    - Aglossia and microglossia
    - Ankyloglossia
    - Cleft tongue / bifid tongue
    - Ankyloglossum superius syndrome
    - Lingual varices
    - Lingual thyroid nodule
    - Variations in tongue movement
    - Patent thyroglossal duct cyst
    - Tongue thrusting
    - Lingual polyp
    - Midline fistula
    - Teratoma
    - Reactive lymphoid aggregate
    - Lingual cyst

2. **Local tongue disorders**
    - Fissured tongue
    - Median rhomboid glossitis
    - Benign migratory glossitis
    - Hairy tongue
    - Crenated tongue
    - Foliate papillitis
    - Leukokeratosis nicotine glossi

3. **Papillary changes in tongue**
    a) Atrophic / depapillation

    Local disease
    - Eosinophilic granuloma
    - Traumatic injuries
    - Lesions due to auto mutilation

- Allergic stomatitis
- Facial hemiatropy
- Cranial arteritis
- Chronic candidiasis

Systemic disease

- Iron deficiency anemia
- Plummer – Vinson syndrome
- Pernicious anemia
- Niacin deficiency
- Folic acid deficiency
- Peripheral vascular disease
- Dermatomyositis
- Diabetes
- Syphilis
- Zoster infection
- Tuberculosis

b) Hypertrophic

- White and black hairy tongue
- After antibiotic therapy
- After steroidal therapy
- Hydrogen peroxide therapy
- Immunosuppressive therapy
- Smoking
- High fever
- Constipation
- Hyperacidity

4. Neurological disease

- Glossodynia
- Dyskinesia
- Paralysis
- Trigeminal neuralgia
- Glossopharyngeal neuralgia

- Polyneuritis
- Neurofibromatosis
- Tongue trusting
- Dysgeusia
- Oropharyngeal duct cyst

5. Cyst

- Anterior median lingual cyst
- Bronchogenic cyst
- Epidermoid and dermoid cyst
- Lympho – epithelial cyst
- Mucus cyst
- Gastric mucosal cyst
- Paralytic cyst
- Thyroglossal duct cyst

6. Benign tumor

- Fibroma
- Glomus tumor
- Granular cell myoblastoma
- Leiomyoma
- Rhabdomyoma
- Plasmacytoma
- Neurofibroma
- Keratoacanthoma
- Traumatic neuroma
- Papilloma
- Adenoma
- Hemangioma
- Lymphangioma

7. Premalignant lesion and conditions

- Leukoplakia
- Lichen planus

- Oral sub mucous fibrosis

8. Malignant tumors

- Squamous cell carcinoma
- Adenocarcinoma
- Transitional cell carcinoma
- Verrucous carcinoma
- Mucoepidermoid carcinoma
- Reticular cell carcinoma
- Malignant lymphoma
- Malignant melanoma
- Metastatic tumor
- Sarcoma

9. Metastatic lesions from

- Kidney
- Liver
- Stomach
- Lung

10. Red and white lesions

- Leukoplakia
- Erytroplakia
- Lichen planus
- OSMF
- Candidiasis
- Psoriasis
- Focal epithelial hyperplasia
- White sponge nevus
- Pemphigus
- Syphilitic mucous patches
- Verruca vulgaris

11. Systemic diseases manifested in tongue

- Infections – bacterial, viral, & fungal
- Blood disorders
- Metabolic disorders
- Dermatological disorders
- Collagen & autoimmune disorders

12. Miscellaneous

- Pigmentation of tongue
- Phlebectasia

II. Laskin et al [3]

Localized lingual conditions	**Systemic lingual conditions**
1. Congenital or developmental - Fissured tongue - Lingual thyroid - Thyroglossal duct cyst - Dermoid cyst - Lymphangioma - Hemangioma - Median rhomboid glossitis	**1. Systemic Infectious** - syphilis - Tuberculosis - AIDS - scarlet fever (scarlatina)
2. Traumatic lesions - Traumatic ulcer - Pyogenic granuloma - Neuroma(traumatic neuroma) - Mucous extravasation cyst	**2. Blood dyscrasias** - Anemia - Leukemia

3. Infectious - Herpes simplex infection - Aphthous ulcers (recurrent aphthous stomatitis, canker sores) - Foliate papillitis - Candidiasis (Moniliasis, thrush)	**3. Metabolic diseases** - Diabetes mellitus - Hypothyroidism - Acromegaly - Vitamin B deficiency - Amyloidosis
4. Benign tumors 1. Papilloma 2. Lipoma 3. Rhabdomyoma 4. Neurilemmoma (Schwannoma) 5. Neurofibroma 6. Granular cell tumor	**4. Immunologic disorders** - Pemphigus - Benign mucous membrane Pemphigoid - Erythema multiforme (Stevens-Johnson syndrome) - Lichen planus

5. Neoplastic / Malignant tumors - Squamous cell carcinoma - Malignant salivary gland tumors - Sarcomas 1. Fibrosarcoma 2. Hemangiopericytoma 3. Alveolar soft-part sarcoma 4. Liposarcoma 5. Rhabdomyosarcomas 6. Leiomyosarcoma 7. Synovial sarcoma 8. Chondrosarcoma 9. Neurogenic sarcoma - Metastatic tumors	
6. Idiopathic lesions - Hairy tongue - Benign migratory glossitis (geographic tongue , erythema migrans)	

III. CLASSIFICATION OF LINGUAL LESIONS ACORDING TO THEIR CLINICAL CHARACTERESTIC [3]

1. Vesiculo-bullous

Primary herpes simplex

Recurrent herpes simplex

Erythema multiforme

Mucous membrane pemphigoid

Pemphigus vulgaris

2. Ulcerative

Squamous cell carcinoma
Pyogenic granuloma
Traumatic ulcer
Aphthous ulcer
Erythema multiforme
Lichen planus
Primary herpes simplex
Recurrent herpes simplex
Mucous membrane pemphigoid
Pemphigus vulgaris
Tuberculosis

Leukemia

Primary syphilis (chancre)

Tertiary syphilis (gumma)

3. Atrophic

Chronic candidiasis (median rhomboid glossitis)

Benign migratory glossitis (geographic tongue)

Vitamin B deficiency

Anemia

Diabetes mellitus

Lichen planus

4. Cystic

Thyroglossal duct cyst

Mucous extravasation cyst

Dermoid cyst

5. Benign and malignant neoplasms of the tongue

Benign

Fibroma

Papilloma

Lipoma

Rhabdomyoma

Leiomyoma

Neuroma

Neurofibroma

Neurilemmoma

Granular cell tumor

Malignant

Squamous cell carcinoma

Salivary gland tumor

Sarcoma

Metastatic tumor

6. Congenital and developmental tongue lesions

Fissured tongue

Lingual thyroid

Lymphangioma

Hemangioma

Median rhomboid glossitis

V. CLASSIFICATION OF DISEASES OF TONGUE (Vanderwaal & Pindborg) [4]

A) Congenital & Developmental Disorders

Aglossia

Ankyloglossia

Ankyloglossum superius syndrome

Fordyce's spots

Hemangioma

Hypoglossia-hypodactylia syndrome

Lingual polyp

Lingual thyroid

Lymphangioma

Macroglossia, congenital

Microglossia

Midline fistula

Osteochondroma

Teratoma

B) Local Tongue Diseases

Coated and hairy tongue

Crenated tongue

Foliate papillitis

Geographic tongue

Leukokeratosis nicotina glossi

Macroglossia, acquired

Median rhomboid glossitis

Plicated tongue

C) Tongue Lesions as Part of an Oral Disease

Allergic stomatitis

Angioneurotic edema

Candidiasis

Eosinophilic granuloma

Focal epithelial hyperplasia

Hemiatrophy

Injuries and ulcerations

Melanoplakia

Myositis ossificans

Necrotizing sialometaplasia

Phlebectasia

Recurrent aphthous ulceration

Verruciform xanthoma

D) Lingual Changes in Systemic Disease

Infectious diseases

Blood dyscrasias

Metabolic disorders

Dermatologic disorders

Collagen diseases (immunopathies)

E) Neurologic Disturbances

Atrophy

Dyskinesia

Glossodynia

Neuralgias

Paralysis

Paresthesia

Tongue thrusting

F) Cysts

Anterior median lingual cyst

Bronchogenic cyst

Epidermoid and dermoid cyst

Gastric mucosal cyst

Lymphoepithelial cyst

Mucous cyst

Parasitic cysts

Thyroglossal duct cyst

G) Benign Tumors and Tumor like lesions

Fibroma

Glomus tumor

Granular cell tumor

Keratoacanthoma

Leiomyoma

Lipoma and lipoblastomatosis

Mesenchymoma

Neurogenic tumors

Papilloma

Plasmacytoma, solitary

Pyogenic granuloma

Rhabdomyoma

Salivary gland tumors, benign

H) Premalignant Lesions and Conditions

Premalignant lesions

Premalignant conditions

I) Malignant Tumors

Carcinoma

Malignant lymphoma

Malignant melanoma

Metastatic tumors

Salivary gland tumors, malignant

Sarcoma

EMBRYOLOGY [5]

Development of the tongue starts in the 4th month of the intrauterine life. The tongue develops from the pharyngeal arches in the floor of the developing mouth. Each pharyngeal arch arises as a mesodermal thickening in the lateral wall of the foregut and then it grows ventrally to become continuous with the corresponding arch of the opposite side.

The medial - most parts of the mandibular arches proliferate to form two lingual swellings. The lingual swellings are partially separated from each other by another swelling that appears in the midline. This median swelling is called the tuberculum impar. Immediately behind the tuberculum impar, the epithelium proliferates to form a downgrowth (thyroglossal duct) from which the thyroid gland develops. The site of this downgrowth is subsequently marked by a depression called the foramen caecum.

Another midline swelling is seen in the region of the medial ends of the second, third, and fourth arches. This swelling is called the hypobranchial eminence. The eminence soon shows a subdivision into a cranial part related to the second & third arches (called the copula) and a caudal part related to the 4th arch. The caudal part forms the epiglottis.

	Anterior two-thirds of the tongue	**Posterior one-third of the tongue**
Formation	Formed by the fusion of the tubercular impar and the two lingual swellings	Formed from the cranial part of the hypobranchial eminence
Arch	Mandibular arch	In this situation, the 3rd arch mesoderm grows over the second arch mesoderm to fuse with the mesoderm of the first arch. The posterior one-third of the tongue is thus

		formed by third arch mesoderm.
Nerves	Lingual branch of the mandibular nerve, which is the post-trematic nerve of this arch, is the sensory nerve; and the chorda tympani, which is the pre-trematic nerve of the first arch, is the nerve of taste.	Glossopharyngeal nerve.

[The posterior – most part of the tongue is derived from the fourth arch. It is supplied by the superior laryngeal nerve.]

The musculature of the tongue is derived from the occipital myotomes.

Nerve supply: hypoglossal nerve

The epithelium of the tongue is at first made up of a single layer of cells. Later it becomes stratified and papillae become evident.

Taste buds are formed at the terminal branches of the innervating nerve fibres.

ANATOMY OF THE TONGUE

The tongue is a muscular organ situated in the floor of the mouth. It is conical in shape and presents with the following features.

1. A root
2. A tip
3. A body, which has
    a) Curved upper surface or dorsum, and
    b) An inferior surface.

The dorsum is divided into oral and pharyngeal parts. The inferior surface is confined to the oral part only.

The tip of the tongue forms the anterior free end which, at rest, lies behind the upper incisor teeth.[6]

The tongue consists of two parts, namely

1. Oral part – the anterior 2/3rd that lies in the mouth
2. Pharyngeal part – the posterior 1/3rd that lies in the pharynx.

The two are divided by a faint V – shaped groove, the sulcus terminalis. The two limbs of the 'V' meet at a median pit, named the foramen caecum. The oral and pharyngeal parts of the tongue differ in their development, topography, structure and function.[6]

ORAL (PRESULCAL) PART:

The presulcal part of the tongue is located in the floor of the oral cavity. It shows the following areas

Apex: touches the incisor teeth

Margin: contact with gums and the teeth

Superior surface (dorsum): related to the hard and soft palates.

On each side in front of the palatoglossal arch, there are four or five vertical folds, the foliate papillae, which represent vestiges of larger papillae found in many other mammals.

The dorsal mucosa has a longitudinal median sulcus and is covered by filiform, fungiform, and circumvallate papillae.

The mucosa of the inferior (ventral) surface is smooth, purplish and reflected onto the oral floor and gums: it is connected to the floor anteriorly by the lingual frenulum. The deep lingual vein which is visible lies lateral to the frenulum on either side. The plica fimbriata, a fringed mucosal ridge directed anteromedially towards the apex of the tongue, lies lateral to the vein.[6]

The tongue has no internal bony skeleton. It is mass of the muscle that must provide its own unique skeletal base. And it does; its movements spring from a "hydrostatic skeleton". The biomechanical principle for such a construct is the maintenance of constant volume despite instantaneous shifts in shape. The device depends on two physical properties:

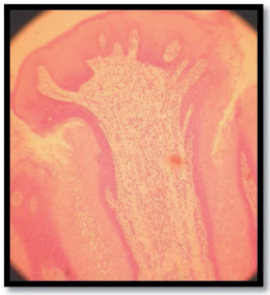
- The incompressibility of water
- The capacity to contract along three planes of the space[1]

Types of papillae

The majority of taste buds on the tongue sit on raised protrusions of the tongue surface called *papillae*. There are four types of papillae present in the human tongue:

Fungiform papillae – These papillae are found as isolated, elevated mushroom – shaped papillae scattered between the filiform papillae and are approximately 150 – 400 µm in diameter. They are covered by a relatively thin epithelium that may or may not be keratinized and have a vascular core of lamina propria. Taste buds may be found on the surface.[7]

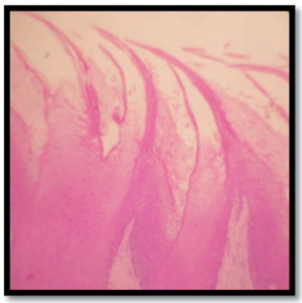
These are present mostly at the apex of the tongue, as well as at the sides. They are distinguished by bright red colour and are innervated by the facial nerve.



Fungiform papilla

Filiform papillae – [L.filum, thread or rather file-like] are tapered, thread like structures that are composed of epithelium and the underlying lamina propria. They lie in oblique, transverse rows that become parallel to the sulcus terminalis toward the base of the tongue.[8]These are thin, long papillae "V"-shaped cones that don't contain taste buds but are the most numerous and give the tongue a characteristic velvety appearance. These papillae are mechanical and not involved in gustation. They are characterized by increased keratinization.

Each filiform papilla consists of a central core of lamina propria with smaller, secondary papillae branching from it. The filiform papillae are highly abrasive during mastication when the bolus is compressed against the palate.[7]



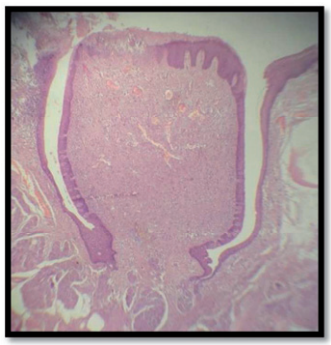
Filiform papilla

Foliate papillae - These are ridges and grooves towards the posterior part of the tongue found on the lateral margins. They are innervated by the facial nerve (anterior papillae) and glossopharyngeal nerve (posterior papillae).[8]

Circumvallate papillae – (vallate = looking like a rampart, so called because each vallate papilla is flanked by a trench-like groove) they are large in size - 1-2 mm in diameter. Only about 3-14 of these papillae are seen in most people,

and they are present at the back of the oral part of the tongue. They are arranged in a circular-shaped row just in front of the sulcus terminalis of the tongue. They are associated with the ducts of von Ebner's glands and are innervated by the glossopharyngeal nerve. These papillae have a connective tissue core that is covered on the superior surface by a keratinized epithelium. The epithelium covering the lateral walls is non keratinized and contains taste-buds.

Groups of mucous glands are also seen within the muscle of the tongue, particularly in the posterior part, and these are unencapsulated.[7]



Circumvallate papilla

Taste buds are small structures on the upper surface of the tongue, soft palate, and epiglottis that provide information about the taste of food being eaten. The human tongue has about 10,000 taste buds.

Structure of taste buds

Taste buds are small ovoid or barrel – shaped intraepithelial organs about 80µm high and 40µm thick. They extend from the basal lamina to the surface of the epithelium.[10] Each taste bud is flask-like in shape, its broad base resting on the corium, and its neck opening by an orifice, the gustatory pore, between the cells of the epithelium. The bud is formed by two kinds of cells: supporting cells and gustatory cells.

The *supporting (sustentacular) cells* are mostly arranged like the staves of a cask, and form an outer envelope for the bud. Some, however, are found in the interior of the bud between the gustatory cells.

The *gustatory (taste) cells*, which are chemoreceptors, occupy the central portion of the bud; they are spindle-shaped, and each possesses a large spherical nucleus near the middle of the cell. The peripheral end of the cell terminates at the gustatory pore in a fine hair-like filament, the gustatory hair.

The central process passes toward the deep extremity of the bud, and there ends in single or bifurcated varicosities.

The nerve fibrils after losing their medullary sheaths enter the taste bud, and end in fine extremities between the gustatory cells; other nerve fibrils ramify between the supporting cells and terminate in fine extremities; these, however, are believed to be nerves of ordinary sensation and not gustatory.

It is known that there are five taste sensations:

Sweet, Bitter, and Umami (now sometimes called Savory), which work with a signal through a G-protein coupled receptor; and Salty and Sour, which work with ion channels.

Localization of taste and the human "tongue map"

Contrary to popular understanding that different tastes map to different areas of the tongue, taste qualities are found in all areas of the tongue. [12] The original "tongue map" was based on a mistranslation by Harvard psychologist Edwin G. Boring of a German paper that was written in 1901. Sensitivity to all tastes occurs across the whole tongue and indeed to other regions of the mouth where there are taste buds (epiglottis, soft palate).[13]

PHARYNGEAL (POSTSULCAL) PART

The postsulcul part of the tongue constitutes its base and lies posterior to the palatoglossal arches. Although it forms the anterior wall of the oropharynx, it is described here for convenience. Its mucosa is reflected laterally onto the palatine tonsils and pharyngeal wall, and posteriorly onto the epiglottis by a median and two lateral glossoepiglottic folds which surround two depressions or valleculae.

The pharyngeal part of the tongue is devoid of papillae, and exhibits low elevations. The ducts of small seromucous glands open on the apices of these elevations. Lymphoid nodules embedded in the submucosa are collectively termed as lingual tonsil. [14]

Arterial supply of the tongue:

It is chiefly derived from the lingual artery, a branch of the external carotid artery. The root is also supplied by the tonsillar and ascending pharyngeal arteries. [6]

Venous drainage:

The arrangement of the veins of the tongue is variable. Two venae comitantes accompany the lingual artery and one vena comitante accompanies the hypoglossal nerve. The deep lingual vein is the largest and the principal vein of the tongue. [6]

Lymphatic drainage:

The tip of the tongue drains bilaterally into the submental nodes. The right and left halves of the remaining part of the anterior two-thirds of the tongue drain unilaterally into the submandibular nodes. A few central lymphatics drain bilaterally into the same nodes.

The posterior one-third of the tongue drains bilaterally into the jugulo-omohyoid nodes. Since most of the lymph from the tongue ultimately drains into the jugulo-omohyoid nodes, these are known as lymph nodes of the tongue. [6]

Nerve supply:

Motor nerves: All intrinsic and extrinsic muscles, except the palatoglossus, are supplied by hypoglossal nerve. The palatoglossus is supplied by the cranial part of the accessory nerve through the pharyngeal plexus.

Sensory nerves: The lingual nerve is the nerve of general sensation and the chorda tympani is the nerve of taste for the anterior two-thirds of the tongue. The glossopharyngeal nerve is the nerve for both general sensation and taste for the posterior one-third of the tongue. The posterior – most part of the tongue is supplied by the vagus nerve through the internal laryngeal branch.[6]

Muscles of the tongue:

The tongue is divided by a median fibrous septum, attached to the body of the hyoid bone.

There are two types of muscles in the tongue:

Extrinsic muscles (extending outside the tongue and moving it bodily)	**Intrinsic muscles (present wholly within the tongue and altering the shape)**
genioglossus	superior longitudinal
hyoglossus	inferior longitudinal
styloglossus	transverse
palatoglossus	vertical

EXTRINSIC MUSCLES

Genioglossus:

It is fan shaped muscle which forms the main bulk of the tongue.[6] It is triangular in sagittal section, lying near and parallel to the midline. It arises from a short tendon attached to the superior genial tubercle behind the mandibular symphysis, above the origin of geniohyoid. From this point it fans out backwards and upwards.

The **inferior fibres** of genioglossus are attached by a thin aponeurosis to the upper anterior surface of the hyoid body near the midline.

The **intermediate fibres** pass backwards into the posterior part of the tongue.

The **superior fibres** ascend forwards to enter the whole length of the ventral surface of the tongue from the root to apex, intermingling with the intrinsic fibers.[14]

Vascular supply: It is supplied by sublingual branch of lingual artery and the submental branch of facial artery.

Innervation: The genioglossus is innervated by the hypoglossal nerve.

Actions: The main action of the genioglossus is forward traction of the tongue to protrude its apex from the mouth. Acting bilaterally, the muscles of the two sides depress the central part of the tongue, making it concave from side to side. When acting unilaterally, the tongue diverges to the opposite side.

Applied aspects: The attachment of genioglossus to the genial tubercles prevents the tongue from sinking back and obstructing respiration; therefore anesthetists pull the mandible forward.

Hyoglossus:

Hyoglossus is thin and quadrilateral, and arises from the whole length of the greater cornu and the front of the body of the hyoid bone.

Relations: Superficial surface is related to the digastric tendon, stylohyoid, styloglossus and mylohyoid, the lingual nerve and submandibular ganglion, the sublingual gland, the deep part of the submandibular gland and duct, the hypoglossal nerve and deep lingual vein.

Deep surface is related to the stylohyoid ligament, genioglossus, the middle constrictor and the inferior longitudinal muscle of the tongue and the glossopharyngeal nerve.

Posterioinferiorly it is separated from the middle constrictor by the lingual artery.

Vascular supply: It is supplied by sublingual branch of lingual artery and the submental branch of facial artery.

Innervation: The hyoglossus is innervated by the hypoglossal nerve.

Actions: It depresses the tongue.

Styloglossus:

The styloglossus is the shortest and the smallest of the three stylohyoid muscles. It arises from the anteriolateral aspect of the styloid process near its apex, and from the styloid end of the stylomandibular ligament. Passing downwards and forwards, it divides at the side of the tongue into a longitudinal part, which enters the tongue dorsolaterallly to blend with the inferior longitudinal muscle in front of hyoglossus, and an oblique part, overlapping hyoglossus and decussating with it.

Vascular supply: It is supplied by the sublingual branch of the lingual artery.

Innervation: It is innervated by the hypoglossal nerve.

Action: It draws the tongue up and backwards.

INTRINSIC MUSCLES:

Superior longitudinal	**Inferior longitudinal**	**Transverse**	**Vertical**
It lies beneath the mucous membrane of dorsum of tongue.	It is a narrow band lying close to the inferior surface of the tongue between the genioglossus and the hyoglossus.	It passes laterally from the median fibrous septum to the submucous fibrous tissue at the lingual margin, blending with the palatopharyngeus.	It extends from dorsal to the ventral aspect of the tongue in the anterior region.

Vascular supply: Intrinsic muscles are supplied by lingual artery.

Innervation: All intrinsic lingual muscles are innervated by the hypoglossal nerve.

Actions: The intrinsic muscles alter the shape of the tongue.

- Contraction of the superior and inferior longitudinal muscles tends to shorten the tongue.
- Superior longitudinal muscles turn the apex and sides upwards to make the dorsum concave.
- Inferior longitudinal muscles pull the apex down to make the dorsum convex.
- Transverse muscle narrows and elongates the tongue
- Vertical muscle flattens and widens the tongue.

Acting alone or in pairs and in endless combination, the intrinsic muscles give the tongue precise and highly varied mobility, important not only in alimentary function but also in speech.[6]

Taste Buds

Taste buds are the organs which act as taste receptors, found in small groups in the papillae. These develop early in foetal life, those around the circumvallate papillae being fully formed by about the 14th week of intrauterine life.[2]

A young adult has a total of around 9000 taste buds, most of which are on the tongue and situated on the papillae on its dorsum. The thin keratinized projections of the filiform papillae rarely have taste buds on them. The smooth, rounded projections of the fungiform papillae each have a few taste buds on their surface. The foliate papillae have five or more taste buds on each of the lateral walls of the ridges. The foliate papillae possess about 1500 taste buds in total.[2]

In the circumvallate papillae the central dome is keratinized, but the sides of the groove are non-keratinized and contain some 250-300 taste buds in each papilla. The number of taste buds in the circumvallate papillae decreases with age. [14, 15]

STRUCTURE OF TASTE BUDS:

A typical taste bud contains three main type of cells designated as Types I to III, together with the basal progenitor cells (or Type IV cells). Their tips extend up into a pit or taste pore, which contains a gel-like material made up of glycosaminoglycans, ascorbic acid (Vitamin C) and a number of enzymes.

Type I cells: Around the periphery of the taste bud and between the other cells are the Type I or dark cells. About 2/3 of the cells in a taste bud are type I cells. They have narrow, dense cell necks, with fibrillar or tubular elements and end in a flattened surface with about 40 short stubby microvilli.

These are separated from the pit substances by a basement membrane which lines the inner surface of the pit. The presence of apical granules suggests that their function is to secrete the pit substances. In light microscopic descriptions these cells were termed as sustentacular cells.

Type II cells **(light cells)**: These cells have relatively thick microvilli projecting into the taste pit and have a number of vesicles immediately below these. They have a smooth walled apically situated nucleus. They do not extend down to the base and they do not show any synapse-like features. About 20% of the cells in the taste buds are of type II.

Type III cells: have slender peg like tips with no microvilli. They contain vesicles which resemble the synaptic vesicles found in nerve fibres and they appear to synapse with adjacent nerve fibres. They are termed serotonergic cells because they react with anti-serotonin antibody reagents.

Type IV cells: At the base of the taste buds there are morphologically less well defined, type IV cells which are thought to be the progenitor cells for the other

three types. All taste buds have similar structures and there does not appear to be a variation based on taste sensitivity. The life of a taste bud cell is between 2 and 30 days. All taste buds contain nerve fibres - after entering the taste bud these lose their myelin sheaths and end closely adjacent to all the cell types. [2,14,15,18]

Sensory mechanisms of taste:

Taste may be defined as the detection and recognition of liquid phase stimuli. Taste (or gustation) is a sensation that is developed well before birth. Initially, therefore, it may comprise part of the monitoring system of the amniotic environment. Substances which stimulate taste receptors are referred to as sapid; the process involved in taste perception is termed gustation. To stimulate taste receptors, a substance must be in solution and if they are not ingested in solution they are solubilised in saliva. The sapid solution then reaches the taste pits where it reacts with pit content and finally with the cells of the taste bud which are the taste receptors.[2]

The nerve pathways which carry taste information from the tongue are all autonomic pathways and signals are carried in small myelinated, slowly conducting axons to reach the tractus solitarius and its nucleus where they synapse. The secondary neurons cross the midline and travel in the medial lemniscus to the thalamus which then communicates with the lower part of the postcentral gyrus and the hippocampal gyrus.[2]

There are connections with the salivary nuclei to give a reflex pathway and there is a link via reticular neurons to the hypothalamus and limbic cortex.[2]

The perception of tastes depends upon the mixture of signals reaching the sensory cortex. The different tastes which human beings can perceive have been classified as sweet, bitter, salt and sour.[2]

There is localization of taste sensitivity in the mouth. The areas of the tongue innervated by the facial nerve fibres respond to sweet, salt and sour stimuli and are arranged progressively more posteriorly along the edge.

The dorsum is more sensitive to sour and bitter stimuli. The areas of the mouth innervated by fibres of glossopharyngeal nerve respond to bitter gustants. [2]

Taste producing substances dissolved in the oral fluid, act by forming a weak attachment to receptors on the microvilli of the gustatory cells. The nature of

this combination of the way in which electrical impulses are generated in the gustatory nerve cells is unknown. The binding of substance to the receptors must be weak because washing the tongue with water can abolish the taste produced by any of these substances.[2, 5]

Modalities of Taste:

In marked contrast to the sense of smell there are only four basic modalities of taste: sweet, sour, bitter and salt. In the tongue sweet sensitivity in greatest at the tip, sour at the sides, bitter at the back while salt sensitivity is more homogenous but greater at the tip. Mid-dorsum is insensitive to all tastes.[2]

Substances producing basic taste sensations:

Except in the case of hydrogen ions and sour taste there is a clear rationalism between chemical constitution and basic or primary taste sensation.
Sour -The sour taste is due almost entirely to hydrogen ions and the degree of sourness is roughly related to the degree of dissociation. Some acids have other tastes as well; for e.g. citric acid in sweet as well as sour and picric acid in bitter and sour.[2]

Amino acids may taste sour, bitter, sweet or nasty depending on the concentration and varying among different subjects. Weak organic acids such as acetic acid that occurs in vinegar, are sourer than would be expected from their degree of dissociation. The threshold pH for HCl is 3.5 and for acetic acid 3.8.[2]

Salty: Sodium chloride in the reference substance for pure salty taste, through oddly enough at threshold concentration (0.02M) it tastes sweet. The anion contributes most to the taste of salts but the cation can modify the anionic effect. Potassium salts tend to be bitter as well as salty and potassium iodides are only bitter. Salts of heavy metals such as mercury have a metallic taste and lead acetate and beryllium salts a sweet taste.

Bitter: Many different types of chemical substances produce bitter tastes. Quinine sulphate is the classical bitter substance with a threshold of 0.006008m. Strychnine hydrochloride is even more with a threshold at 0.0000016m. Other alkaloids such as morphine and nicotine are bitter, as also are caffeine urea, phenylurea, and certain salts such as magnesium sulphate. Many sweet substances have a concomitant bitter taste or after taste e.g. saccharin. This double taste is most apparent as the substance travels from the

front of the tongue, where sweet tastes are appreciated to the back where bitter sensitivity is particularly developed.

Sweet: The sweet taste is associated primarily with organic compounds except for certain inorganic salts of lead and beryllium. Sucrose is the standard reference for sweetness, with the threshold concentration of 0.01M. Fructose in sweeter than sucrose and maltose, galactose and lactose are less sweet than glucose. However, the taste sensation is not the same for all these sugars.

Synthetic sweeteners, such as saccharine, dulcin and cyclomate are used as substitutes for sucrose in diabetics and obese persons in whom sugar intake must be reduced. A substance called gymnemic acid, extracted from the leaves of an Indian plant called gymnema sylventre, selectively abolished the sensation of sweet, leaving bitter, salt and sour unaffected.[20]

Development and Aging:

Taste buds appear very early in the tongue of the human foetus, at 7 to 8 weeks of gestation, but mature appearing taste buds are not observed until later in gestation.

Development is not complete at birth but numbers continue to increase. Behavioral testing of human newborns reveals that the ability to discriminate between the taste stimuli is present at birth, indicating that some attribute of taste preference behavior are innate and do not require any experience for expression.[5]

Role of saliva in taste function:

Saliva is essential for normal taste function. It is usually difficult to taste food with a dry mouth. Saliva not only acts as a solvent for chemical stimuli in food, but also transports their stimuli to the taste receptors. At rest, gustatory receptors are covered with a layer of fluid that extends into the taste pores and bathes the receptor surface of the microvilli. For taste buds in the fungiform papillae it presumably consists of pooled saliva from all the salivary glands.

Taste buds in the circumvallate and foliate papillae are bathed in saliva derived from Von Ebner's glands. Taste buds on the palate, larynx and pharynx are covered in fluid secreted by a large number of small salivary glands draining onto the surface of the epithelium. Gustatory stimulation alters salivary flow and composition; the fluid environment may alter during

transduction. Although this microenvironment, acting as a transport system may merely plays a passive role in taste mechanism, it may have a more active role. Components within the taste pore may control access and removal of stimuli and interact with tastant during the initial event in receptor – stimuli interaction.[5]

FUNCTIONS OF TONGUE

The tongue serves numerous functions. An awareness of these functions is important in diagnosing and managing local and systemic disorders affecting the tongue. The main functions of the tongue are:

1. Speech: The tongue interrupts the air passage through mouth or pharynx thereby producing consonants. Certain consonants like c, d, j, n, t, z, l, g etc. require movements of the tongue.[15]

2. Mastication: The tongue is very important for mastication. Jenkins divides the masticatory role of tongue into three phases:

- The tongue may have a direct crushing effect on food by pressing it against the hard palate
- The tongue pushes the food in between the occluding surfaces of the teeth and helps to mix in the saliva.
- The sensory nerve endings of the tongue enable it to select those parts of the food mass that are sufficiently well masticated to be ready for deglutition.[15]

3. Deglutition: Deglutition is a reflex response triggered by afferent impulses in the trigeminal, glossopharyngeal, and vagus nerves. The process is initiated by the voluntary action of collecting food on the tongue and propelling it backward into the pharynx.[16] The muscles involved in the process are the mylohyoid and pharyngeal constrictors. The bolus is pushed backwards by raising the back of tongue. It is sucked from the mouth into the pharynx by creating a negative pressure, while airways are still closed, by rapid relaxation of muscles of tongue and pharynx. This process has been described by Ardran and Kemp as being like toothpaste pressed from the tube. [17]

4. Taste: Taste is one of the chemical senses. Basically, four primary qualities are distinguished: [18]

1	Bitter	base of the tongue
2.	Salty	widespread, but greatest at the tip
3.	Sweet	tip of the tongue
4.	Sour	lateral margins

The chemical stimulus is converted into nerve impulses through nerve endings located within the taste buds.

5. Digestion: Tongue has a slight digestive function by virtue of salivary lipase, present in the serous lingual salivary gland.

6. Barrier function: Mucosa covering the tongue acts as a barrier protecting the deeper tissues from mechanical damage. It also prevents the entry of microorganisms and toxic substances. Chemo-receptors and mechanoreceptors in the tongue surface sense the nature and mechanical properties of ingested food, and prevent the digestion of noxious substances.[19]

7. Jaw development: Muscular pressure from the tongue is an important factor in determining the shape of the mandibular arch and the position of anterior and posterior tooth segments. The congenital and developmental anomalies of the tongue significantly affect jaw development and increase in the tongue size in adults will cause other deformities and spacing of teeth.[15]

8. Secretion: The major secretion of the tongue is provided by minor salivary gland activity which maintains the moist surface of the oral mucosa.[15]

9. Defence mechanism: Secretory immunoglobulin system plays an important role in body defense.[15]

10. Maintenance of oral hygiene: By virtue of its movement the tongue can reach all parts of the oral cavity removing food debris from the gums, vestibule and floor of the mouth. Thus, it helps in maintenance of oral hygiene.[15]

11. Respiration: Electromyographic studies of the tongue muscles (genioglossus, in particular) indicate that the position of the jaw and the tongue, by contributing to lingual muscular tonus, influence respiratory control. These studies have led a better understanding of various sleep apnea syndromes and the role of severe malocclusion and hypotonicity of tongue in such problems. [15]

12. Suckling: It is the highly specialized form of ingestion characteristic of the infant mammal; in humans, the mother's nipple and surrounding areola are drawn deep into the open mouth, the everted lips form a seal, and the dorsum of the tongue is applied to the nipple and areola, which are rhythmically compressed by the jaw and tongue, discharging milk onto the dorsum of the tongue at about the level of the row of vallate papillae. The lingual muscular activity generates a peristaltic wave moving posteriorly that compresses or

milks the nipple and propels milk toward the pharynx. Ultrasound imaging of the infant tongue during suckling demonstrates that the medial portion of the tongue into which the genioglossus is inserted moves in relation to the lateral portion, the styloglossus and hyoglossus insertions.[15]

13. Symbolic functions: Disease, deformity and pain in the tongue can imply personality traits. The tongue may figure prominently in sexual encounters, playing both a physical and symbolic phallic function.[15]

CLASSIFICATION OF CONGENITAL AND DEVELOPMENTAL DISORDERS

A) According to Burkit tongue lesions can be classified as

Inherited, Congenital and Developmental Anomalies

- Variations in tongue morphology and function
- Variations in tongue movements
- Fissured, Plicated, or scrotal tongue
- Patent thyroglossal ducts, thyroglossal duct cyst and lingual thyroid
- Cleft, lobed, bifurcated and tetrafurcated tongues
- Aglossia (hypoglossia)
- Macroglossia
- Bald or depapillated tongues
- Localized enlargement and papillomatosis

B) Congenital or developmental anomalies of tongue (Working classification)

- Aglossia and Microglossia
- Hypoglossia
- Macroglossia
- Ankyloglossia
- Cleft tongue / Bifid tongue
- Fissured tongue / Scrotal tongue
- Benign migratory glossitis
- Lingual thyroid
- Lingual Varices
- Lingual Polyp
- Thyroglossal duct cyst
- Dermoid cyst
- Cartilaginous and osseous choriostoma
- Midline fistula
- Lymphangioma
- Hemangioma
- Fordyces granules
- Teratoma

AGLOSSIA AND MICROGLOSSIA

Aglossia and its modification, microglossia, are rare congenital anomalies. It was apparently first described early in the eighteenth century by de Jussieu. In many cases, aglossia has been associated with other congenital anomalies, especially those of the extremities. [21]

Aglossia literally means the absence of the entire tongue. To the best of our knowledge aglossia without other oral or generalized malformations has rarely been reported and only in conditions that were incompatible with life. Besides, in those cases just the anterior two thirds of the tongue, which is the part of the tongue that develops in front of the buccopharyngeal membrane, were missing. The term partial aglossia would, therefore, be more appropriate. [4]

Microglossia is an uncommon developmental condition of unknown cause that is characterized by an abnormally small tongue. Isolated microglossia is known to occur, and mild degrees of microglossia may be difficult to detect and may go unnoticed. However, most reported cases have been associated with one of a group of overlapping conditions known as oromandibular-limb hypogenesis syndromes. These syndromes feature associated limb anomalies, such as hypodactylia (i.e., absence of digits) and hypomelia (i.e. hypoplasia of part or all of a limb). Other patients have had coexisting anomalies, such as cleft palate, intraoral bands, and situs inversus. Microglossia frequently is associated with hypoplasia of the mandible, and the lower incisors may be missing. [22]

The different combinations and groups of the anomalies have been listed under different syndromes like Goldenhar syndrome, Moebius syndrome, aglossia adactylia syndrome, Hanhart syndrome, glossopalatine ankylosis syndrome, limb deficiency- spleno gonadal fusion syndrome, and Charlie M syndrome. There have been variations in anomalies and frequency of overlapping features. [23]

Embryology and etiological hypothesis:

Aglossia and microglossia is a rare anomaly caused by failed embryogenesis of the lateral lingual swellings and tuberculum impar from the fourth to eighth gestational weeks.[24]

Clinical features:

An isolated case of aglossia has been reported in the literature. The finding of a constricted opening surrounded by thin membranous structure between oral cavity and pharynx resembled the persistent buccopharyngeal membrane and is comparable with a similar finding reported by Higashi and Edo.
The tongue with its highly co-ordinated muscular activity helps in sucking, swallowing and manipulating food during chewing. The history from the child's mother and the trial with food in the clinic revealed that she had no marked difficulty in such activities. The child was able to manipulate the food and this reflected the adaptive capacity of human beings. [23]

Proper pronunciation, particularly that of lingual consonants is dependent upon the movement of tongue. The speech of the girl was affected. It was slurred but understandable. She could manage to pronounce with some difficulty. This reflected obviously the role of tongue in proper pronunciation of words. [23]
The perception of taste was surprising. Taste buds are located in different lingual papillae. So, in case of aglossia, the perception of taste was unexplainable. There could be the possibility of some lingual mass in the form of rudimentary tongue or buried in adjacent structures which could not be detected clinically. No attempt was made to detect the taste buds histologically because of practical unfeasibility. [23]

A life-threatening event in the Pierre Robin syndrome is glossoptotic airway obstruction. Several explanations for such an obstruction range from an essentially passive phenomenon to a more active event as a result of negative pressure generated by deglutition and inspiration.[4]

Khalil et al have reported a case of 30-year-old man with aglossia. The entire tongue was absent. The development of the maxilla and mandible were affected. However the patient was able to cope with the oral functions with some difficulty.[23]

Kantapura et al correlated thyroid dysfunction in a patient with aglossia, reporting a Thai girl who had aglossia, micrognathia, microsomia, collapse of the mandibular arch with congenital absence of mandibular incisors, persistence of buccopharyngeal membrane, microcephaly and mild developmental delay. Thyroid function tests indicated hypothyroidism.[23]

Aglossia-adactylia syndrome:

The facies is usually sharp and narrow with a receding chin, producing a birdlike appearance. Involvement of the extremities varies from bilateral peromelia to agenesis of a single digit. Syndactyly and absence of fingernails have also been reported. There is no evidence that heredity plays a role in the genesis of this anomaly. Neither is there any sex predilection.

The tongue, though apparently completely absent in some patients, may be present as a small nubbin located posteriorly in the mouth, consisting essentially of that part normally developed from the copula. Speech is not severely impaired. The sublingual muscular ridges and salivary glands are hypertrophic. The mandible is usually small, and the anterior alveolar ridge may be underdeveloped. Cleft palate, persistence of the buccopharyngeal membrane and bony fusion of the jaws have also been.[21]

Treatment and prognosis:

Treatment of the patient with microglossia depends on the nature and severity of the condition. Surgery and orthodontics may improve oral function. Surprisingly, speech development often is quite good but depends on tongue size.[22]

In most cases of congenital microglossia non-surgical techniques such as positioning, nasogastric intubation, nasopharyngeal airway, and temporary endotracheal intubation are successful. Only in severe cases is a tongue-lip adhesion required with posterior placement of the tongue incision just above the floor of the mouth and stripping of the origin of the genioglossus muscles to provide secure control of glossoptosis in combination with a flap created on the mucosa of the lower lip. The flap is based on the labial sulcus and turned back over the alveolar ridge to be sutured to the margin of the tongue incision. A suture is passed through the base of the tongue for 6 to 10 days as described by Routledge. In an extreme case a tracheostomy may be required.[4]

HYPOGLOSSIA-HYPODACTYLIA SYNDROME:

The hypoglossia – hypodactylia syndrome was first reported by Rosenthal in 1932 as 'aglossia congenita' and later the present term was proposed by Hall. Total or partial absence of the tongue associated with abnormalities of the limbs is a rare but well-known phenomenon. The cause of the syndrome is unknown. No inheritance pattern has been noted, and until 1971, just nine cases had been reported. Because in almost all of the reported cases there is actually hypoglossia or microglossia, together with variable anomalies of the limbs, the term hypogiossia-hypodactylia syndrome seems to be more appropriate than the previously used term aglossia-adactylia syndrome.[4]

Etiology:

During development of the embryo, the anterior tongue is formed from three structures of the first branchial arch, the two lateral lingual swellings and a median swelling, the tuberculum impar. The second and third branchial arches form the posterior part of the tongue. Therefore, hypoglossia is attributed to the failed growth of the two lateral lingual swellings and tuberculum impar, and the second and third branchial arches**.** [25]s

Although a number of theories have been proposed, the etiology of hypoglossia is largely unknown. Internal and external factors during fetal development play an important role in all malformations, including the development of tongue anomalies. Also, hyperfunction and hypofunction can influence development after birth. The important influence of tongue malformation on postnatal tooth alignment and jaw development is well known.[25]

Classification:

In 1971 Hall introduced a classification that he called the oromandibular-limb hypo-genesis syndrome:

Type 1

1. Hypoglossia
2. Aglossia

Type 2

1. Hypoglossia-hypodactylia
2. Hypoglossia-hypomelia (peromelia)
3. Hypoglossia-hypodactylomelia

Type 3

1. Glossopalatine ankylosis (ankyloglossum superius syndrome)
2. With hypoglossia
3. With hypoglossia-hypodactylia
4. With hypoglossia-hypomelia
5. With hypoglossia-hypodactylomelia

Type 4

1. Intraoral bands and fusion
2. With hypoglossia
3. With hypoglossia-hypodactylia
4. With hypoglossia-hypomelia
5. With hypoglossia-hypodactylomelia

Type 5

1. Primary biliary cirrhosis of the liver (Hanhart syndrome)
2. Charlie M syndrome
3. Pierre Robin syndrome
4. Mobius syndrome
5. Amniotic band syndrome [4]

Clinical features:

The following features are essential for the diagnosis of this syndrome.

1. Reduction in the size of the tongue in several cases hypoglossia and ankyloglossia were observed.
2. Micrognathia due to underdevelopment or absence of bone and teeth in the midline segment of the mandible or maxilla.
3. Limb anomalies of varying severity may be syndactyly, adactyly, hypomelia; one or more limbs may be affected. [25]

Other reported oral manifestations of the syndrome consist of micrognathia, gingival abnormalities, high-arched or cleft palate, glossopalatine ankylosis, defects in the lower lip, intraoral bands, absence of lateral mandibular incisors, and hyper-

trophy of the sublingual glands. Intelligence is normal, and eventually speech may be surprisingly good.[4]

Management:

Surgical treatment was designed to improve the tongue, lip, and mandibular function. Treatment consisted of maxillary and mandibular osteotomies. It should be planned on a long-term basis with a multidisciplinary approach used for improvement of oro-facial abnormalities.[4]

MACROGLOSSIA, congenital

Macroglossia means long or large tongue. The synonym *glossocele* is sometimes used. It is difficult to establish clinical criteria for assessment of the size of the tongue, as either normal, too small (microglossia), or too large. Usually, the size of the tongue is related to the size of the mandible. Macroglossia can be divided into two forms: congenital and acquired as suggested by Vander Waal & Pindborg.[4]

Causes of macroglossia:

Pseudomacroglossia includes any of the following conditions, which force the tongue to sit in an abnormal position:

- Habitual posturing of the tongue
- Enlarged tonsils and/or adenoids displacing tongue
- Low palate and decreased oral cavity volume displacing tongue
- Transverse, vertical, or anterior/posterior deficiency in the maxillary or mandibular arches displacing the tongue
- Severe mandibular deficiency (retrognathism)
- Neoplasms displacing the tongue
- Hypotonia of the tongue.

True macroglossia can be subdivided into two main subcategories, congenital causes and acquired causes.

Congenital causes

- Idiopathic muscle hypertrophy
- Gland hyperplasia
- Hemangioma
- Lymphangioma
- Down syndrome
- Beckwith-Wiedemann syndrome
- Behmel syndrome
- Lingual thyroid
- Gargoylism
- Transient neonatal diabetes mellitus

- Trisomy 22
- Laband syndrome
- Lethal dwarfism of Blomstrand
- Mucopolysaccharidoses
- Skeletal dysplasia of Urbach
- Toliner syndrome
- Autosomal dominant inheritance
- Microcephaly and hamartoma of Wiedemann
- Ganglioside storage disease type I

Acquired causes (Categories have been assigned to simplify the list, but there can be overlap of a particular etiology into more than one of these categories.)

- **Metabolic/endocrine**
  - Hypothyroidism
  - Cretinism
  - Diabetes
- **Inflammatory/infectious**
  - Syphilis
  - Amebic dysentery
  - Ludwig's angina
  - Pneumonia
  - Pemphigus vulgaris
  - Rheumatic fever
  - Smallpox
  - Typhoid
  - Tuberculosis
  - Actinomycosis
  - Giant cell arteritis
  - Candidiasis
  - Scurvy
  - Pellagra
- **Systemic/medical conditions**
  - Uremia
  - Myxedema
  - Hypertrophy
  - Acromegaly

- Neurofibromatosis
- Iatrogenic macroglossia

- **Traumatic**
  - Surgery
  - Hemorrhage
  - Direct trauma (e.g. biting)
  - Incubation injury
  - Radiation therapy
- **Neoplastic**
  - Lingual thyroid
  - Lymphangioma
  - Hemangioma
  - Carcinoma
  - Plasmacytoma
- **Infiltrative**
  - Amyloidosis
  - Sarcoidosis [26]

Clinical features: The first report of macroglossia was a description of oral lymphatic malformation, in 1854, by Virchow and Uber. [27]

Congenital macroglossia is an uncommon condition with significant morbidity. The characteristic picture is tongue protrusion that may lead to dental and facial abnormalities, mucosal exposure and dryness, exposure to trauma, dysphagia and difficulty in phonation, airway obstruction, salivation and growth delay.[27]

Macroglossia most commonly occurs in children and can range from mild to severe. In infants, macroglossia may be manifested first by noisy breathing, drooling, and difficulty in eating. The tongue enlargement may result in lisping speech. The pressure of the tongue against the mandible and teeth can produce a crenated lateral border to the tongue, open bite, and mandibular prognathism. If the tongue constantly protrudes from the mouth, it may ulcerate and become secondarily infected or may even undergo necrosis. Severe macroglossia can produce airway obstruction.[22]

Most cases are based on the presence of an extensive hemangioma, lymphangioma, neurofibroma**,** or cyst. The tongue may be involved locally or in total. In a series of five patients with congenital macroglossia in India, there was

one case of idiopathic or muscular macroglossia. The patient was not a cretin and did not suffer from Down's syndrome. Muscular macroglossia may occur unilaterally. It occurs usually in those individuals who are mentally defective and short-lived. [4]

Some congenital syndromes often express macroglossia in their phenotypes, most commonly Down syndrome (1 per 700 live births) and Beckwith-Wiedemann syndrome (0.07 per 1000 live births). In Beckwith-Wiedemann syndrome, 97.5 per cent of patients have macroglossia.[26]

Beckwith-Wiedemann syndrome was first described by Beckwith in 1963, and later by Wiedemann, in 1964, as exophthalmos/omphalocele, macroglossia and gigantism syndrome (EMG/OMG syndromes).[27]

Macroglossia is a characteristic feature of Beckwith-Wiedemann syndrome, a rare hereditary condition that includes many other possible defects, such as the following:

- Omphalocele (i.e., protrusion of part of the intestine through a defect in the abdominal wall at the umbilicus)
- Visceromegaly
- Gigantism
- Neonatal hypoglycemia

In Beckwith-Wiedemann syndrome, the tongue usually shows a diffuse, smooth, generalized enlargement.[22]Individuals with Beckwith-Wiedemann syndrome have an increased risk for several childhood visceral tumors, including Wilms' tumor, adrenal carcinoma, hepatoblastoma, rhabdomyosarcoma, and neuroblastoma. Facial features may include nevus flammeus of the forehead and eyelids, linear indentations of the earlobes, and maxillary hypoplasia (resulting in relative mandibular prognathism). Most examples of Beckwith-Wiedemann syndrome are sporadic, but 10% to 15% of cases show autosomal dominant inheritance with preferential maternal transmission. The genetic basis is complex, involving a variety of alterations within two domains of imprinted growth-regulator-v genes on chromosome 11p15.[22]

Today it is related to other malformations. It is a genetic syndrome of overgrowth that is relatively common, and characterized by congenital abnormalities, such as visceromegaly, macroglossia, abdominal wall defects,

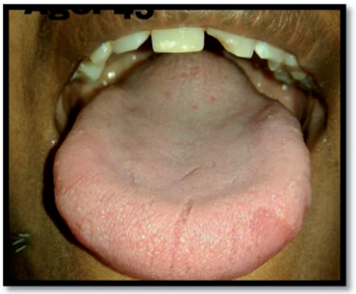
pre and postnatal overgrowth and neonatal hypoglycemia. It is a polymorphous syndrome subject to a variable combination of signs and symptoms. Among the several anomalies mentioned, macroglossia is the most common manifestation of the syndrome, found in 82% to 99% of the individuals affected. It may be associated with a spectrum of craniofacial alterations. The syndrome may cause difficulty in swallowing, phonation and even respiratory problems due to inability to keep the tongue inside the mouth. This may lead to dryness, ulcerations and even infections in the tip of the tongue.[27]

Moreover, untreated macroglossia may result in impaired craniofacial development, with open bite and inclined incisors, leading to prognathism. The histological examination of macroglossia shows muscle hyperplasia or even normal histology. Many authors approach this anomaly by waiting for the child to grow, since the tongue tends to accommodate inside the mouth as the jaw grows. [27]

In patients with lymphangiomas, the tongue surface is characteristically pebbly and exhibits multiple vesicle-like blebs that represent superficial dilated lymphatic channels. The enlarged tongue in those with Down syndrome typically demonstrates a papillary, fissured surface. [22]

In patients with hemifacial hyperplasia, the enlargement will be unilateral. Some patients with neurofibromatosis also can have unilateral lingual enlargement. [22]

The secondary effects of a grossly enlarged tongue merit serious consideration for active treatment. Prolonged exposure can cause ulceration and necrosis of the mouth and tip of the tongue. Maxillofacial abnormalities including anterior open bite, prognathism, and an increased angle between the ramus and body have been described. Noisy breathing, drooling, and the unsightly appearance of a protruberant tongue, particularly in children, can cause distress. Difficulties in swallowing due to limited movement of the enlarged tongue can lead to poor weight gain and failure to thrive. Problems with articulation occur, particularly the expression of consonants requiring the tip of the tongue to be in contact with the alveolar ridge or roof of the mouth. The most serious and life threatening complication is airway obstruction, which is more common in generalized or posterior lingual enlargement than in anterior enlargement of the tongue. [28]



Macroglossia

Histopathologic features:

The microscopic appearance of macroglossia depends on the specific cause. In some cases, such as the tongue enlargement seen with Down syndrome or in edentu1ous patients, no histologic abnormality can be detected. When macroglossia is due to a tumor, a neoplastic proliferation of a particular tissue can be found (e.g., lymphatic vessels, blood vessels, neural tissue). Muscular en1argement occurs in those with hemifacial hyperplasia and Beckwith-Wiedemann syndrome. In the patient with amyloidosis, an abnormal protein material is deposited in the tongue. [22]

Diagnosis:

The evaluation of a patient with macroglossia should begin with a thorough history and physical examination, which may allow the recognition of a syndrome of which the enlarged tongue is one component. Assessment of the tongue should include examination for masses and changes in colour and consistency. Thyroid function tests, isotopic imaging of the thyroid gland, chromosomal studies, and urinary mucopolysaccharide assay may be indicated. Patients with chronic airway obstruction should be assessed for pulmonary hypertension and cardiac decompensation with electrocardiography, chest radiography, arterial blood gas analysis, and Doppler echocardiography. Computed tomography and magnetic resonance imaging may be useful to delineate soft tissues and to show the extent of tumours and other masses. Microscopic examination of tongue tissue in primary macroglossia may be unhelpful, but biopsy is useful for localised lesions of the tongue that occur in chronic granulomatous and neoplastic disorders. Biopsy of other potentially

affected tissue (rectum, skin, gums) is indicated to diagnose definitively amyloidosis. [29]

Management:

Management of congenital macroglossia presents the following problems:

a. Preservation of taste
b. Restoration of normal size and shape of the tongue
c. Correction of the dental arch deformity and malocclusion of teeth by orthodontic treatment
d. Correction of defective articulation by speech therapy

An inverted V-shaped incision, as described by Hendrick and Antonio in 1956, seems preferable to meet the requirements in the surgical treatment of congenital macroglossia. In case of an angiomatous type of macroglossia, radiation therapy should not be used because of the increased risk of an induced malignancy with such treatment. [4]

ANKYLOGLOSSIA

The term 'ankyloglossia' comes from the Greek words 'ankyloglossiakilos' for crooked or loop and 'glossa' for tongue. [33]

Ankyloglossia, or tongue-tie, is a congenital developmental condition in which the tongue is abnormally fixed to the floor of the mouth or the lingual aspect of the gingival mucosa (or both). It is characterized by the presence of a short, thick lingual frenum resulting in limitation of tongue movement. No epidemiologic figures are available from the literature. On the basis of a rough estimation, the prevalence is probably less than one case in 1,000 births. [4]

It has been reported to occur in 1.7% to 4.4% of neonates and is four times more common in boys than in girls. In adults, mild forms are not unusual, but severe ankyloglossia is a relatively uncommon condition that has been estimated to occur in about 2 to 3 of every 10,000 people. [22]

Kupietzky A et al stated that its incidence varies from 0.02% to 5%, depending on the study, its definition of ankyloglossia, and the population examined. The incidence among outpatients of a children's hospital with breast-feeding problems was almost 3 times higher (13%). Two independent studies of oral anomalies in neonates found a significant predilection (3x) for ankyloglossia in males. [31]

Etiology, physiology and possible pathology:
During early development, the tongue is fused to the floor of the mouth. Cell death and resorption free the tongue, with the frenulum left as the only remnant of the initial attachment. Tongue-tie is the result of a short fibrous lingual frenulum or a highly attached genioglossus muscle, affecting from 0.02% to 4.4% of newborn infants. The lingual frenulum usually becomes less prominent as a natural process of the child's growth and development, when the alveolar ridge grows in height and the teeth begin to erupt. This process occurs during the first six months to five years of life. Ankyloglossia is defined as complete if there is a total fusion between the tongue and the floor of the mouth or partial if it arises from a short lingual frenulum, the latter being by far the most common type. [33]

The role of a short lingual frenulum as a cause of breastfeeding difficulties has been described in multiple anecdotal reports linking partial ankyloglossia to decreased tongue mobility and a potential inability to latch on properly. [33]

It is important to remember that the swallowing mechanism of the newborn and infant is different from the adult or older child. It has been noted that for successful nursing to occur, the infant must latch on to the mother's areola with his/her upper gum ridge, buccal fatty pads and tongue. Suckling begins with the forward movement of the jaw and tongue. The tongue helps to make a better seal, but with minimal active action. The anterior edge of the tongue thins, cupping upwards to begin a peristaltic ripple back toward the throat. At the same time, the lower jaw squeezes milk from the ductules. Finally, the posterior part of the tongue depresses to allow milk to collect in the oropharynx before swallowing. It is clear that restriction of the tongue movements must be quite extreme to interfere with sucking and swallowing. [33]

Acevedo AC et al stated that ankyloglossia could be observed as part of a syndrome or as an isolated trait. Little is known about the pathogenesis of ankyloglossia. Cleft palate with ankyloglossia with a dominant X-linked mode of inheritance has been reported in the literature and more recently, mutations in the transcription factor TBX22 gene have been identified in some of these families. [32]

Classification of ankyloglossia:

The term 'free tongue' is defined as the length of tongue from the insertion of the lingual frenum into the base of the tongue to the tip of the tongue. Because the tongue is a muscle, which in young children is flexible and often difficult to stabilize, placing a dental instrument at the insertion point and approximating the tip of the tongue helps determine this measurement. A Boley gauge is then used to measure this distance. [30]

A group of 322 children, ranging in age from 18 months to 14 years, were examined for the length of free tongue and then evaluated for clinical evidence of speech and oral problems. Assessment of these measurements resulted in the development of the following descriptions and categories of ankyloglossia. [30]

<u>Classification of Ankyloglossia Based on "Free Tongue" Length (Kotlow)</u>

Clinically acceptable, normal range of free tongue: - 16 mm

Class I: Mild ankyloglossia: 12 to 16 mm

Class II: Moderate ankyloglossia: 8 to 11 mm

Class III: Severe ankyloglossia: 3 to 7 mm

Class IV: Complete ankyloglossia: less than 3 mm [31]

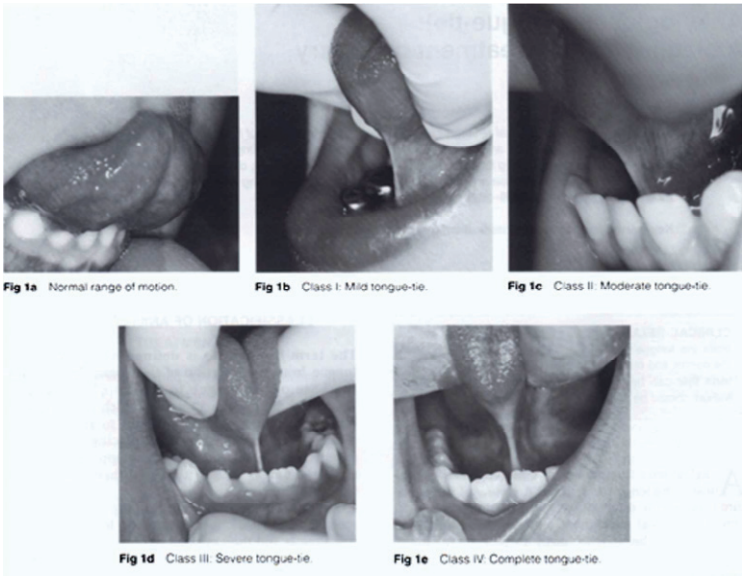


Fig 1a Normal range of motion. **Fig 1b** Class I: Mild tongue-tie. **Fig 1c** Class II: Moderate tongue-tie. **Fig 1d** Class III: Severe tongue-tie. **Fig 1e** Class IV: Complete tongue-tie.

Wallace distinguished four clinical groups of ankyloglossia patients:

1. Infants referred for surgery when less than 2 years old because the parents believe that the tongue-tie should be "snipped" lest it subsequently interfere with speech.
2. Children between 2 and 4 years, who are slow to speak properly, and anxious parents blame the tongue-tie.
3. Children from age 4 to young adulthood, who, in addition to a tongue tie, usually have a serious cause for defective speech, such as an underdeveloped mandible, mental retardation, or stammering.

4. Children whose tongue-tie has recurred after snipping. [4]

Structural guidelines were developed to assist in determining if the lingual frenum required revision. A normal range of motion of the tongue is indicated by the following criteria:

1. The tip of the tongue should be able to protrude outside the mouth without clefting
2. The tip of the tongue should be able to sweep the upper and lower lips easily, without straining.
3. When the tongue is retruded, it should not blanch the tissue lingual to the anterior teeth.
4. The tongue should not place excessive forces on the mandibular anterior teeth.
5. The lingual frenum should allow a normal swallowing pattern.
6. The lingual frenum should not create a diastema between the mandibular central incisors.
7. In infants, the underside of the tongue should not exhibit abrasion.
8. The frenum should not prevent an infant from attaching to the mother's nipple during nursing.
9. Children should not exhibit speech difficulties associated with limitations of the movement of the tongue. [30]

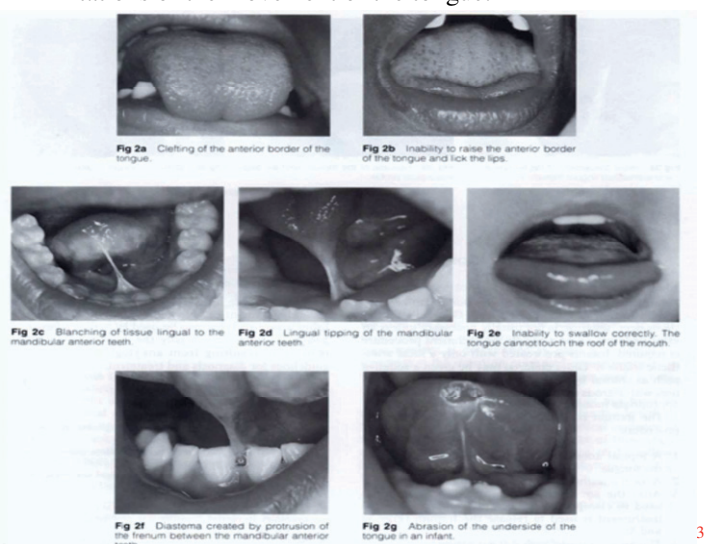


Fig 2a Clefting of the anterior border of the tongue.

Fig 2b Inability to raise the anterior border of the tongue and lick the lips.

Fig 2c Blanching of tissue lingual to the mandibular anterior teeth.

Fig 2d Lingual tipping of the mandibular anterior teeth.

Fig 2e Inability to swallow correctly. The tongue cannot touch the roof of the mouth.

Fig 2f Diastema created by protrusion of the frenum between the mandibular anterior teeth.

Fig 2g Abrasion of the underside of the tongue in an infant.

3

Lingual frena in the complete (Class IV) tongue-tie category should be revised because they severely restrict the tongue's movement. Many lingual frena in the severe (Class III) category also benefit from revision. Children often adapt to the short attachment (Class III) with fatiguing efforts. Release of this frenum is often recommended. Children with moderate (Class II) and mild (Class I) ankyloglossia are the most difficult to evaluate. Most of these children appear to have normal speech patterns and are able to effortlessly fulfill most of the criteria listed above. [30]

In addition to the guidelines cited, there are additional reasons for lingual frenum revisions. These include abnormalities in tongue function during swallowing, difficulty in eating or drinking, difficulty in playing wind instruments, difficulty in licking ice cream cones and, during the adult years, instability of dentures as well as impairment of certain social activities. Flexibility of the floor of the mouth is also an important factor in determining the effect of ankyloglossia. Some of the children examined displayed normal mobility of the tongue in conjunction with a flexible floor of the mouth; others displayed restricted tongue movement, when the tension of the floor of the mouth exhibited little or no flexibility. The tension was associated with a pulling of the tissue behind the mandibular incisors or the development of a diastema between the mandibular central incisors. It also appeared that, in Class I and Class II ankyloglossia, a natural lengthening of the free tongue might occur as a child grows. [30]

Clinical features:

Ankyloglossia can be complete or partial. Only in extreme cases of ankyloglossia may nursing and feeding become a problem shortly after birth, necessitating the surgical removal of the fibrous band. Mild cases of ankyloglossia may go un-noticed for a few years, until the time that speech becomes impaired. [4]

Other features of tongue-tie are:

1. Heart shaped tongue when raised or protruded out
2. “V” shaped notch at the tip of the tongue
3. Patients cannot usually extend the tongue tip beyond the upper gums

4. Patient cannot move the tongue from side to side
5. Usually deviate while swallowing the tablets
6. Patients have problems while swallowing
7. Digestive problems due to inability to chew properly
8. Difficulty in licking an ice cream cone [34]

It should be emphasized that, without further research, clinicians should not advise parents that the existence of ankyloglossia will result in a malocclusion in their child. Although there is lack of scientific evidence proving a relationship between speech disorders and ankyloglossia, a consensus seemingly exists that ankyloglossia is not a cause of a delay in speech onset. Ankyloglossia may interfere with articulation. The existence of ankyloglossia in the newborn may result in breastfeeding difficulties, including ineffective latch, inadequate milk transfer, and maternal nipple pain. An infant with ankyloglossia may experience difficulty latching on to the nipple and may compress the nipple against the gum pad instead of the tongue, resulting in nipple pain and an inefficient, inadequate seal. A mother experiencing such pain may often contemplate switching the baby to a bottle. Indiscriminate or immediate clipping of all or most lingual frenula, however, is not universally recommended. It is crucial to understand the mechanism of feeding and its relationship with oral structures to comprehend the suggested role of ankyloglossia in feeding problems. Many of the reports on this suggested mechanism are anecdotal, and evidence-based studies are lacking. A recent study of 3,000 infants concluded that:

1. Ankyloglossia represents a significant proportion of breast-feeding problems
2. Surgical interventions in the presence of significant ankyloglossia facilitated successful and improved breast-feeding. [31]

The possible sequlae of ankyloglossia remain controversial, and the range of suggested complications is great. Among the suggested complications found in the literature are:

1. Lower incisor deformity

2. Gingival recession; and
3. Malocclusions [31]

Dentists are encouraged to learn more about the mechanisms of breast-feeding in the lactation literature. Ankyloglossia may have sequelae beyond those of speech or feeding difficulties. Children may be teased by their peers for their anomaly. Social issues include the inability to lick ice cream, play a musical wind instrument, and even kiss. [31]

Ankyloglossia may occur with increased frequency in various congenital syndromes, including Opitz syndrome, orofaciodigital syndrome, Beckwith-Wiedemann syndrome, Simpson-Golabi-Behmel syndrome, and X-linked cleft palate. [31]

Management:

The need for surgical correction of ankyloglossia is exceptionally small and is perhaps justified only in severe congenital cases when nursing and feeding become a problem or in adults in whom the frenum interferes with the wearing of a denture. According to Wallace, no frenulum should be divided at all before the age of 4 years and thereafter only if the mechanism of tongue protrusion is too feeble to stretch or rupture it.

Other authors have confirmed that statement by mentioning that nursing techniques do not require that the short frenum be cut, and because of the dangers associated with this so-called simple operation, it should not be performed. [35]

When surgical intervention is justified, the attachment of the undersurface of the tongue should be divided at least 1.27 cm horizontally and sewn up vertically. This method has the advantage over other techniques, such as the Z-plasty, that the resulting scar is situated on the undersurface of the tongue, well away from the area that concerns the prosthodontist. Injudicious cutting of the frenum in a new-born can cause hemorrhage; the tongue may become too mobile and may be swallowed, causing asphyxia. There is also a possibility of a subsequent infection at the base of the tongue, with the formation of a large ulcer and a spreading stomatitis. [4]

ANKYLOGLOSSUM SUPERIUS SYNDROME

Ankyloglossum superius syndrome is characterized by the attachment of the tongue to the hard palate and by limb malformations.

Etiology:

A genetic disorder is the most important or perhaps the only factor in the etiology, although intrauterine environmental factors have been considered as well. A possible relationship with the hypoglossia-hypodactylia syndrome has been mentioned by several authors.

Clinical features:

Very few cases have been reported. In one of the described cases jejunal and ileal atresias were extensive. A case of attachment of the tip of the tongue to a cleft palate without anomalies of the extremities has also been reported.

Management:

Treatment consists of surgical separation of the tongue from the palate.[4]

BIFID TONGUE

Cleft tongue, Lobed, Bifurcated, Tetrafurcated Tongue

Cleft, or bifid, tongue describes the condition in which there is cleavage of the tongue due to lack of fusion of the lateral halves. It is an extremely rare phenomenon. The tongue may be completely or only partially cleft. [4]

A completely cleft or bifid tongue is a rare condition that is apparently due to lack of merging of the lateral lingual swellings of this organ. A partially cleft tongue is considerably more common and is manifested simply as a deep groove in the midline of the dorsal surface. The partial cleft results because of incomplete merging and failure of groove obliteration by underlying mesenchymal proliferation.[26]

Etiology:

The tongue anterior to the circumvallate papillae is formed from two lateral tubercles that fuse in the midline between fourth and the fifth embryonal week. Failure of fusion of the tubercles causes cleft (bifid; lobulated) tongue. Lobulated tongue (i.e. division of the tongue into two, three, or four lobes) is associated with oro-facial-digital syndrome. Bifid tongue is also seen in association with median cleft of the mandible. It also may occur as an isolated phenomenon or be combined with cleft palate. [21]

Clinical features:

The lack of fusion of the lateral halves of the tongue may result in the formation of two complete tongues. Also, the occurrence of trifid tongue and even further divisions has been mentioned. None has been reported without other grave errors of development. A deep groove in the midline of the dorsal surface of the tongue, as can be seen in many persons, could perhaps be regarded as an extremely mild form of partially cleft tongue. However, this has no clinical significance. The same is true of a small cleft in the tip of the tongue. [4]

The frequency of isolated cleft tongue is unknown, but Witkop and Barros found an incidence of 1 per 1,000 among Chileans. [21]

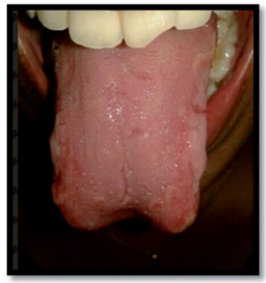
Double tongue, formed as a result of a developmental anomaly within a lingual tubercle, has previously been described in association with a cleft palate (Bartholdson et al 1991; though no salivary gland component was described),

and the tongue was reconstructed at 40 days. Lingual hamartoma and cleft palate have also been described. [36]

Cleft tongue can be of a feature of oral—facial—digital syndrome in association with thick, fibrous bands in the lower anterior mucobuccal fold eliminating the sulcus and with clefting of the hypoplastic mandibular alveolar process. [26]

Clinically, cleft tongue alone is of little significance, except for hygiene problems, i.e. food debris and microorganisms may collect in the base of the cleft which may lead to local irritation. [37]

It is seen associated with various syndromes: Orofacial digital syndrome I and II, Meckel's syndrome, Trisomy 21 (Down) syndrome, Cryptophthalmos (Fraser's) syndrome, Ectodactyly-ectodermal-dysplasia-clefting (EEC) syndrome, Coffin - Lowry syndrome, Robinow's syndrome, Goldenhar syndrome, Klippel-Feil syndrome. [15]



Bifid tongue

Treatment:

Despite the apparent severity of these various tongue malformations, surgical repair of congenital tongue clefts is frequently successful and case reports often refer to normal shape and function in patients followed for some years.[15]

PLICATED TONGUE

Plicated tongue, also called *lingua dissecta, fissured tongue,* or *scrotal tongue,* represents a change in the dorsal surface of the anterior two-thirds of the tongue. Several definitions of plicated tongue have been suggested in the literature, such as "a tongue with or without a central fissure which show parallel fissures lateral to the midline, or fissures at right angles to the long axis of the tongue." Another description is "a tongue being characterized by furrows, one extending anteroposteriorly and others laterally over the entire anterior surface". [4]

Fissured tongue is a congenital malformation affecting the dorsum of the tongue. This anomalous condition can be differentiated from pathologic conditions by observing the papillae. In the scrotal tongue the papillae will extend into, and completely cover, the mucosa of the furrow. In the pathologic condition causing deep fissures, the depth of the fissures will be denuded of papillae. Occasionally the hypertrophy of the papillae causes a "hairy" tongue. [38]

Glossal double fissures were found in a study of prenatal and postnatal human specimens. Histologic analysis suggested that the double fissure appearance was a reflection of the topography of the subjacent muscle bundles and that true fissuring, when present, was probably a secondary phenomenon akin to acquired plication. [4]

Etiology:

The cause of plicated tongue is unknown, although a developmental nature seems most likely. A polygenic or autosomal dominant mode of inheritance is suspected as this condition is seen with increased frequency in families with an affected proband. Extrinsic factors such as chronic trauma or vitamin deficiencies are considered possible causes of fissured tongue as well.[4,26]

Clinical features:

The fissures may be relatively shallow or may be deep, in which case food debris will accumulate and result in inflammation. Depending on the pattern of the grooves, and several sub classifications have been proposed in the literature, such as *foliaceus, cerebriform,* and *transverse.*[4]

In a study among children in the Republic of South Africa, six different patterns of tongue fissuring were observed *(1)* Plication, *(2)* Central longitudinal fissuring, *(3)*

Double fissures, *(4)* Transverse fissuring arising from a central fissure, *(5)* Transverse fissuring without a central fissure, and *(6)* Lateral longitudinal fissures.[4]

In the same study, some tongues exhibited more than one pattern. The author suggested that most tongue fissuring seen in children should be considered variations in normal anatomy rather than an abnormality. The condition is found more frequently in the elderly. There is no predilection for any particular race. Some reporters have showed a slightly male predilection. The prominence of the condition appears to increase with increasing age.[26]

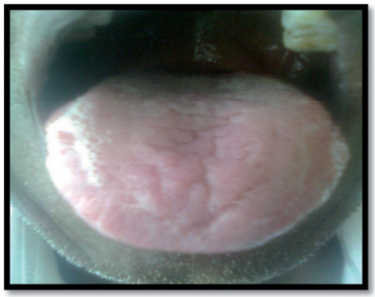
Fissured tongue affects the dorsum and often extends to the lateral borders of the tongue. The depth of the fissures varies but has been noted to be up to 6 mm in diameter. When particularly prominent, the fissures or grooves may be interconnected, separating the tongue dorsum into what may appear to be several lobules. The lesions are usually asymptomatic unless debris is trapped within the fissures or when it occurs in association with geographic tongue.[39]

Prevalence figures vary widely. In an unselected Swedish adult population, a prevalence of 7 % was reported. In a study from Finland, a prevalence of 5.0 % was recorded. The prevalence is much higher among those who are mentally handicapped. In a study of 50 people with Down's syndrome, a prevalence rate of 22% was observed. [4]

Fissured tongue was the most common oral finding in psoriasis cases.[40]

The plicated tongue is one of the signs of the Melkersson-Rosenthal syndrome, the other signs being recurrent swellings of the face or lips with recurrent facial nerve paralysis. Melkersson-Rosenthal syndrome was first described in 1928 by Ernst Gustaf Melkersson with two main features and in 1931 Curt Rosenthal added lingual plicata as the third main feature to the syndrome complex. There has also been a case of a patient with a plicated tongue with bilateral Bell's palsy without facial edema. Fissured tongue is also seen associated with Downs syndrome, Coffin-Lowry syndrome, orofacial digital syndrome and Rabinow’s syndrome. A plicated-like aspect of the tongue has been described in pemphigus vegetans, for which the term *cerebriform tongue* was used.

The author has suggested that most tongue fissuring seen in children should be considered variations in normal anatomy rather than an abnormality.[41,42]



Fissured tongue

Histologic features:

A biopsy is rarely indicated as the clinical findings are characteristic. Histological examination reveals an increase in the thickness of the lamina propria, loss of filiform papillae, hyperplasia of the rete pegs, neutrophilic microabscesses within the epithelium, and a mixed inflammatory infiltrate in the lamina propria. [39]

Treatment:

No definitive therapy or medication is required. If symptomatic, patients are encouraged to brush the dorsum of the tongue and commercially available effervescent mouthwashes or diluted hydrogen peroxide rinses to eliminate debris that may serve as an irritant. Complications are not associated with fissured tongue per se but are noted in association with the manifestations of Melkersson-Rosenthal syndrome. [39]

GEOGRAPHIC TONGUE

Geographic tongue is a benign condition, the cause of which is unknown, and is characterized by the occurrence of one or more smooth areas on the dorsum and the lateral borders of the tongue. In the smooth areas there is an absence of filiform papillae.

Geographic tongue was first described in 1955 and occurs in approximately 3% of the population. The pattern resembles a relief map with mountain ridges, hence the term "geographic". It occurs in all age groups but is more common in adults; the incidence in females is approximately twice that of males.[39]

Often used synonyms are **benign migratory glossitis, erythema migrans, exfoliatio areata linguae et mucosae oris**, and **wandering rash**. The prevalence of geographic tongue can be estimated to be at least 1 % to 2 %, without a distinct preference for race, sex, or age. In a study of 6,090 Iraqi schoolchildren, geographic tongue was observed in 4.3%. In Axell's study in an adult Swedish population, geographic tongue was recorded in more than 8 %.[1]

Patients and their family members may report that the tongue has an abnormal appearance which resembles that of a 'map' or burn injury. The lesions classically wax and wane over time with occasional periods of complete remission, hence the term migratory glossitis. Although generally painless, patients with a geographic tongue may occasionally present with a burning sensation that is noted with hot or spicy foods. Adult patients may occasionally be concerned about a diagnosis of oral cancer, despite reporting that they have noted these lesions over many years.[39]

Etiology:

The etiology of geographic tongue remains unknown. Several related etiologic factors have been proposed, however, none of the suggested causes provide clear-cut evidence of a causal relationship. Some investigators have classified this condition as a congenital anomaly; other researchers have discussed the role of heredity in its development.

Eidelman et al reported that the prevalence of geographic tongue in parent and sibling combinations was significantly higher than in the general population and

concluded it was familial and that heredity plays a significant etiologic role. Other authors have also postulated that a positive family history suggests the possible implication of genetic factors. A study of Greek subjects with geographic tongue indicated an increased frequency of both DR5 and DRW6 antigens when compared to controls. These findings support the theory that genetic factors participate in the pathogenesis of geographic tongue.[43]

In a study of 70,359 Israeli school children, there were 1,380 children with scrotal tongue and 801 with geographic tongue, whereas 391 children had both conditions, which was highly significant.[4]

An association between geographic tongue and fissured tongue has been documented, and a genetic linkage between the two conditions in males has been suggested. The same genes may be responsible for both conditions.[43]

There is a report of the occurrence of geographic tongue in three generations of a large family; in three subjects geographic tongue was associated with hemophilia. In an Australian study of 100 consecutive patients with geographic tongue, there was a significantly high frequency of a history of asthma, eczema and hay fever compared with a control population.[4]

Attempts have been made to demonstrate an association between geographic tongue and various systemic and/or psychological conditions. These conditions include gastrointestinal disorders associated with anemia, Reiter's syndrome, psoriasis, emotional stress, allergies, diabetes, and hormonal disturbances. However, a definitive causal relationship has not yet been established.[43]

It is generally accepted that geographic tongue is not a manifestation of Reiter's syndrome or psoriasis, although some authors prefer to leave that question open, applying the term *annulus migrans* as an inconclusive descriptive term instead. It is very unusual to see geographic tongue and lichen planus simultaneously in the same patient.[4]

Marks et al concluded that a positive association between geographic tongue and atopy exists, and further postulated that geographic tongue and asthma/rhinitis may have a similar pathogenesis. They provided additional support for a genetic basis for geographic tongue by demonstrating an increased incidence of tissue type HLA-B15 in atopic patients with geographic tongue.

Some investigators have suggested that geographic tongue is an oral manifestation of psoriasis. Because of the similarities in the clinical, histologic,

and immunohistochemical findings, these researchers considered psoriasis and geographic stomatitis as related lesions. However, **Espelid et al** failed to show a connection between geographic tongue and psoriasis since HLA-DR was not strongly expressed on the keratinocytes in their samples. **Van der Wal et al** stated an incidentally reported association of geographic stomatitis with psoriasis in a patient did not provide sufficient proof of a common etiologic basis for the two conditions. **Raghoebar et al** concluded that geographic tongue and psoriasis occur coincidentally rather than sharing an etiologic relationship. [43]

Gonzaga HF et al had undertaken a study to investigate human leucocyte antigen (HLA) associations with benign migratory glossitis and psoriasis in Brazilian patients and particularly to determine whether benign migratory glossitis is also associated with HLA-Cw6, the classical association observed in psoriasis. The results showed a highly significant association of Cw6 with both psoriasis and benign migratory glossitis, with this antigen being present in 59.1% of the patients with psoriasis, in 43.8% of the patients with benign migratory glossitis, and in only 12.6% of the controls. Other significant positive associations, although at a lower significance level, were with B13, both in psoriasis and in benign migratory glossitis, and with B17, only in psoriasis. To our knowledge, this is the first report on the association of Cw6 with benign migratory glossitis. We believe that this finding reinforces the concept of a pathogenetic relationship between benign migratory glossitis and psoriasis.[44]

Psychosomatic factors appear to play a significant role in the etiology of geographic tongue. It has been reported that lesions arise in connection with pronounced emotional stress. **Redman et al** found a higher prevalence of geographic tongue in mentally ill patients than in university students. They also noted that when under emotional stress the student group with geographic tongue tended to have more severe lesions. Findings such as these support the possible role of psychological factors in the etiology of geographic tongue.

Wysocky et al studied the relationship between geographic tongue and diabetes and found a fourfold increase in the prevalence of geographic tongue in the diabetic group. However, a recent study by **Guggenheimer et al** reported no significant correlation between geographic tongue and insulin-dependent diabetes mellitus. [43]

In a study of 775 children up to 2 years of age, the association between geographic tongue, seborrheic dermatitis, and spasmodic bronchitis was seen to be present.[4]

Clinical features:

George du Toit et al reported that the disease is characterized by the occurrence of one or more smooth areas on the dorsum and lateral borders of the tongue. The top layer of the 'skin' of the tongue is unevenly shed leading to the classic manifestation of an area of erythema, with atrophy of the filiform papillae of the tongue, surrounded by a serpiginous, white hyperkeratotic border and degeneration of the overlying mucosa. The tongue exhibits a well-demarcated area of erythema, primarily affecting the dorsum, and often extending to involve the lateral borders of the tongue. [39]

The smooth areas heal spontaneously in a matter of hours, days, or even weeks and may in some cases persist for a much longer time. New lesions may develop at other sites on the tongue or may appear after a considerable interval of time. The condition is usually asymptomatic, but in some patients a slight discomfort may be recorded, for instance, when spicy food is eaten. As mentioned before, the geographic tongue may occur superimposed on a plicated tongue. Similar lesions may occur elsewhere on the oral mucosa, and these are sometimes referred to as **ectopic geographic tongue**, or **geographic stomatitis**.[1]

Hume has proposed the following classification.[4]

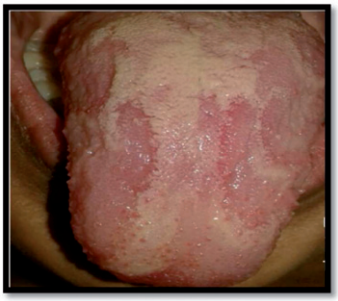
- Type 1 - Lesions confined to the tongue with both active and remission phases; no other lesions elsewhere in the oral cavity.
- Type 2 - As in type 1 but with similar lesions elsewhere in the mouth.
- Type 3 - Lesions of the tongue that are not typical and that may or may not be accompanied by lesions elsewhere in the mouth. The atypical tongue lesions consist of two forms:
    - Fixed form - A few areas of the tongue are affected, but no movement of these areas is observed. They may disappear only to recur at the same area.
    - Abortive forms - These forms start as yellow-white patches but disappear before acquiring the typical appearance of a

geographic lesion.

- Type 4 - No tongue lesions are seen but geographic areas are present elsewhere in the mouth (ectopic geographic tongue). Movement of these areas may or may not be marked.

The diagnosis of geographic tongue rarely needs histopathologic confirmation. White opaque thickenings with a geographic tongue -like appearance may be a manifestation of pachyonychia congenita.[4]

George du Toit et al showed that there is no loss of the sense of taste, or dexterity of the tongue. There is, however, a measurable decrease in the tongue's sense of touch. Importantly, most people with geographic tongue are otherwise healthy.[39]



Geographic tongue

Histopathology:

The microscopic appearance is characterized by numerous polymorphonuclear leukocytes that migrate through the epithelium and give rise to the formation of spongiform pustules or microabscesses, called **Munro's abscesses**, in the upper epithelial layers. The stratum spinosum may be thickened and edematous. In the smooth, red areas the papillae of the connective tissue may reach close to the surface mimicking to some extent the microscopic appearance of psoriasis. The lamina propria usually shows a mild lymphoplasma-cellular infiltrate. Occasionally a few eosinophils may be encountered.[4]

Chronic inflammatory cells can be seen in variable numbers within the stroma and silver or PAS staining will often demonstrate candida hyphae or spores in the superficial layers of the epithelium. There is no liquefaction degeneration of the basal cells, as seen in lichenoid lesions, and there is no ulceration except in cases of Reiter's syndrome.[26]

In a report from France, the histopathologic findings were divided into five consecutive stages, ranging from the changes that can be seen in the periphery of the lesion, with a clinically normal aspect through the center of the lesion, to the regenerating and finally normal-appearing mucosa again. In the literature the electron microscopic findings of a few cases have been reported. A striking intimacy between sensory nerve processes and the basal lamina of the epithelium has been observed.[4]

Treatment:

George du Toit et al have reported that geographic tongue lesions heal spontaneously, and although benign, this condition may last for years and often recurs. Although no treatment is generally recommended, several have been tried, including topical Retin-A and treatments for thrush. Patients who experience pain and burning may experience relief when treated with antihistamines.[39]

In a Swedish study the successful use of local application of a solution of 7 % salicylic acid in 70 % alcohol has been reported. The solution was applied with cotton pellets for about 10 seconds along the borders between affected and un-affected areas, followed by rinsing the mouth with water. The procedure was carried out twice with a two-minute interval and was repeated daily for a maximum of eight consecutive days until the symptoms disappeared. No side effects were noted. Good results have also been claimed in three patients who topically applied 0.1 % tretinoin.

Reassuring the patient that he or she is not suffering from a malignancy commonly suffices. Very few follow-up studies have been reported. In a mean observation period of 2.8 years, it was shown that migratory glossitis is a more or less persistent condition that does not undergo malignant change.[4]

LINGUAL THYROID

Lingual thyroid is defined as the presence of thyroid tissue in the midline at the base of the tongue anywhere between the circumvallate papillae and the epiglottis.[45]

Lingual thyroid is a condition in which thyroid tissue is found in the foramen caecum area of the tongue. A statistical incidence of lingual thyroid is not available. Estimates from large clinics indicate that its occurrence may be 1: 3000 in patients with thyroid disease. Until 1972 a total number of 373 cases had been collected from world literature.[4]

Microscopic examination of human tongues removed at autopsy reveals that despite the absence of a clinically apparent thyroid nodule, as many as 10% exhibit remnants of thyroid tissue within the tongue.[46]

Embryologic aspects:

The thyroid gland originates as a midline endodermal outgrowth at the junction of the dorsal anterior two thirds and the base of the tongue in the region of the future foramen caecum. From there, the thyroid tissue normally descends through the tongue and cervical tissue to reach its final position in the region of the larynx. However, when this migration fails, persistence of thyroid tissue is found in the tongue.[3]

This persistence may result in the formation of a thyroglossal duct cyst or a lingual thyroid. In a series of 15 patients with lingual thyroid, three had thyroglossal cysts excised in earlier life. Thus lingual thyroid is considered as a defect of embryologic development.[4]

The presence of remnants of thyroid tissue in the tongue is well documented. In a study of 200 consecutive routine autopsies, the presence of ectopic lingual thyroid tissues was recorded in 10% of the individuals, and this tissue was located primarily in the region of the foramen caecum. In most instances the thyroid tissue was arranged in a microfollicular adenomatous pattern, rarely appearing as mature thyroid tissue. The size of the ectopic tissue ranged from a few acini to a mass of 1 cm. There were no differences between the sexes. In a study that focused on the foramen caecum area of 100 cadaver tongues, no thyroid remnants were found.[4]

Clinical aspects:

Lingual thyroid is a rare anomaly with a reported incidence of 1 in 3000 of the cases seen with overall prevalence of 1 in 100,000. Of all ectopic thyroids 90% are found to be lingual thyroids.[45]

A lingual thyroid nodule is far more common in females and most commonly becomes clinically apparent during puberty and adolescence.[2] The female to male ratio being about 4:1, is perhaps the result of hormonal influences. The age of onset ranges from birth to the sixth decade, with a peak in the second decade. There are no racial or geographic differences.[4]

It presents as a 2 to 3 cm smooth sessile mass located on the midposterior dorsum of the tongue, in the region of the foramen caecum. The chief symptoms are dysphagia, dysphonia, dyspnea, and a feeling of tightness in the area.[2] The incidence of lingual thyroid is approximately 10%. The overlying mucosa may show an increased vascularity. Hemorrhage may occur.[1]

However, most lingual thyroid nodules are asymptomatic. In as many as 70 % of all patients with lingual thyroids, this lingual thyroid tissue is the only functioning thyroid tissue present. Therefore, a thyroid scan is mandatory before a biopsy is done in this particular region of the tongue. Due to the low sensitivity and specificity of usual ^{99m}Tc scintigraphy, all patients with suspected ectopic thyroid tissue should be examined by radioactive iodine imaging.[4]

Hypothyroidism has been reported in up to 33% of patients. Many authors say that lingual thyroid enlargement is a secondary phenomenon, compensating for thyroid hypofunction. Interestingly, as many as 75% of patients with infantile hypothyroidism have some ectopic thyroid tissue.[22]

Carcinoma arising in a lingual thyroid is even more unusual with fewer than 30 cases reported in the literature.[47]

Clinical differential diagnosis:

The clinical differential diagnosis includes hypertrophied lingual tonsil, thyroglossal duct cyst, salivary gland tumor, and squamous cell carcinoma.[4]

Histopathology:

The histopathologic findings of lingual thyroid tissue are similar to those of cervical thyroid tissue.[1] Most cases are composed of normal mature thyroid tissue

although embryonic or fetal thyroid tissue may also be seen.[2] Most detailed histologic studies have shown that lingual thyroid tissue characteristically has an incomplete or poorly defined capsule. A variable amount of inflammatory cells may be present. The chance of malignant degeneration in a lingual thyroid is rather small. These are usually papillary adenocarcinomas, but follicular carcinomas have been described as well. [4]

Diagnosis:

Diagnosis is best established by thyroid scan using iodine isotopes or technetium-99m. Computed tomography (CT) and magnetic resonance imaging (MRI) can be helpful in delineating the size and extent of the lesion. Biopsy is often avoided because of the risk of hemorrhage and because the mass may represent the patient's only functioning thyroid tissue. In some cases incisional biopsy may be needed to confirm the diagnosis or to rule out malignant changes.[22]

Management:

Treatment of lingual thyroid is not always necessary and depends largely on the complaints. Most patients are in euthyroid state, with normal T_3, T_4 determinations. The serum TSH concentration may be increased. [4]

Some patients show hypothyroidism. Hyperfunction is exceptional. When the mass is causing functional impairment, suppressive doses of thyroid hormones may be sufficient. If not, total excision may be unavoidable. This usually can be done via an intraoral approach, which may require midline splitting of the tongue. Successful auto-transplantation of lingual thyroid into the neck or abdominal muscles has been reported.

Radio - iodine is contraindicated in women of childbearing age because of the risk of teratogenesis, and in children, because of risk of carcinogenesis. Radioiodine is therefore used only when the patient is unfit for anesthesia or refuses surgery. [48]

LINGUAL VARICES

Varicosities are abnormally dilated and tortuous veins. They are relatively trivial but common vascular malformations when seen in the oral mucosa. Age appears to be an important etiologic factor as they are common in older adults. Varices involving the ventral aspect of the tongue are common developmental abnormalities. Less frequently varices occur in other areas of the mouth.[49]

The literature provides a variety of names for the condition observed such as sublingual varices, phlebectasia linguae, caviar lesions, and lingual varicosities. They are benign and acquired. No direct association was established between varicosities and other specific organic diseases. Varicosities in the younger age groups might indicate premature physiologic change.[50]

Clinical features:

The most common type of oral varicosity is the sublingual varix, which occurs in two-thirds of people older than 60 years of age. Sublingual varicosities classically present as multiple blue-purple, elevated or popular blebs on the ventral and lateral border of the tongue. They are typically blue and blanch with compression. The lesions are usually asymptomatic, except in rare instances when secondary thrombosis occurs.[22] Thrombosis, which is insignificant in these lesions occasionally occurs, giving them a firm texture.[50]

Ettinger and Mandersen confirmed that the incidence of varicosities increases with age. Koscard and coworkers concluded that diminished elastic support to capillaries, associated with age, allowed the capillaries to dilate and varicosities to form. Elastic fibres are less numerous in gingiva than in more mobile mucosa such as cheek, floor of mouth, and ventral surface of tongue. They are more numerous in the skin of adult than in skin of infants.[51]

Less frequently, solitary varices occur in other areas of the mouth, especially the lips and buccal mucosa. These isolated varicosities often are first noticed after they have become thrombosed. Clinically, a thrombosed varix presents as a firm, nontender, blue- purple nodule that may feel like a piece of buckshot beneath the mucosal surface.[22]

Histopathologic features:

Microscopic examination of a varix reveals a dilated vein, the wall of which shows little smooth muscle and poorly developed elastic tissue. If secondary thrombosis has occurred, then the lumen may contain concentrically layered zones of platelets and erythrocytes (lines of Zahn). The clot can undergo organization via granulation tissue, with subsequent recanalization. Older thrombi may exhibit dystrophic calcification, resulting in formation of a phlebolith (phlebo = vein; lith = stone).[22]

Treatment and prognosis:

Sublingual varicosities are typically asymptomatic, and no treatment is indicated.[22] No treatment is required for a venous varix unless it is frequently traumatized or is cosmetically objectionable.[50]

LINGUAL POLYP

A polyp is a circumscribed, sessile or pedunculated lesion. The majority of polyps of the oral and lingual mucosa are fibroepithelial overgrowths, often designated as fibromas. A lingual polyp based on a developmental disturbance is a rarity. [4]

Clinical features:

A case of a lingual polyp that caused birth asphyxia has been reported in the literature**.** Immediately after birth the neonate became cyanosed**.** Inspection of the upper airway showed a pedunculated tumor arising from the base of the tongue and obstructing the pharynx. The pedicle was ligated and the tumor excised immediately. The lesion measured 18 x 13 mm and consisted of smooth muscle bundles and fibrous tissue containing adipose tissue and mucus-producing salivary glands.[4, 52]

In the literature a case of an 18-month-old infant with a hamartomatous lesion at the base of the tongue consisting of neural tissue, smooth muscles, and striated muscles was encountered. In another case, a polyp located at the base of the tongue was causing dysphagia in a 64 year old man. [4]

The lesion was of unknown duration and may have been of a congenital or developmental nature, although heavy smoking was suggested by the author as the possible cause. An anterior teratomatous lingual polyp has been described as well. [53]

Histopathological features:

The tumor measured 18 x 13 x 13 mm and consisted of smooth muscle bundles and fibrous tissue containing nerve bundles, blood vessels, lymphatics, adipose tissue, and glands and ducts similar to mucus-producing salivary glands. The nodule was covered with keratinising stratified squamous epithelium containing pilosebaceous follicles and one definite hair.[52]

Management:

Treatment consists of surgical removal. Histopathologic examination should be performed routinely, even in the case of a small, harmless looking lingual polyp. [4]

THYROGLOSSAL TRACT CYST (THYROGLOSSAL DUCT CYST)

Thyroglossal tract cysts arising from remnants of the embryonic thyroglossal duct are found in the midline anywhere from the foramen caecum of the tongue to the sternum.[37] Thyroglossal duct cyst is the most common congenital anomaly that arises from the remnants of the thyroglossal duct and occurs in 7% of the adult population. [54]

Embryologically**,** as the thyroid decends from the base of the tongue to its cervical location, it brings with it a tract of epithelial tissue (thyroglossal tract) that normally involutes in the 10th week of gestation. However the remnants may remain, giving rise to cyst formation in the base of the tongue.[3]

Clinical features:

The cyst may appear extra-orally in the midline or intra-orally in the base of the tongue, producing symptoms of dysphagia. The thyroglossal duct cyst is usually diagnosed in childhood. [4]

Only 2% are located in the mouth. Intraoral lesions arise either in the floor of the mouth or at the foramen caecum.[55]

Thyroglossal tract cysts in the region of the tongue appear as dome-shaped compressible lesions. The swelling usually develops slowly and is asymptomatic. Neoplasms, including carcinomas, have been reported to occur in these cysts. Duvie (1985) reported that thyroglossal cysts are not as rare in the tropics as previously suggested in the literature.[37]

A proportion of the thyroglossal duct cyst has an associated fistula. The cysts are usually in the midline and produce soft, movable, sometimes fluctuant, sometimes tender swellings. Occasionally, they may be located laterally. Classically, they move when the patient swallows or protrudes the tongue. If they are located high in the tract they may cause dysphonia.[55]

Thyroglossal cysts arising in and around the tongue will present clinical features similar to the lingual thyroid nodule, mesenchymal neoplasms, and other cysts such as the epidermoid and dermoid cysts.[37]

Histologic features:

Microscopically, a cyst lined by stratified squamous, columnar, or respiratory epithelium is observed, often in association with follicles of glandular thyroid epithelium.[37]

Mucous cells may be present in the cystic lining and seromucous glands in the wall (Wampler et al, 1978) particularly if the cysts are located in the lingual area. In the same region lymphoid tissue with prominent germinal centres may also be seen.[55]

Carcinoma arising in the thyroglossal duct cyst was first described by Brentano in 1911, and since then about 215 cases have been reported in the literature. The incidence of papillary carcinoma arising in the thyroglossal duct cyst is < 1% and most of these tumors arise from the ectopic thyroid tissue within the cyst. Papillary carcinoma is the most common type being reported. The prognosis of papillary carcinoma arising in a thyroglossal cyst is similar to that of papillary carcinoma of the thyroid gland having cure rates in excess of 95%. [54]

Malignant change has occasionally been observed and it is reported that thyroid carcinoma may arise in approximately 1.5% of lesions. [56]

A review of literature on the subject also revealed nine cases in which squamous cell carcinoma had developed. [57]

Differential diagnosis:

The differential diagnosis involves dermoid cyst, epidermoid cyst, branchial cleft cyst, lymph nodes, lymphangioma, and thyroid pathology. [58]

Treatment:

Thyroglossal tract cysts should be surgically removed along with the tract.[37] Before surgical removal, a thyroid scan should be done to exclude a possible lingual thyroid. It is often necessary to remove a part of the hyoid bone as well. This procedure is called the Sistrunk operation and is strongly recommended to minimize the chance of recurrence.[4]

DERMOID CYST

Some authors use the term dermoid cyst just clinically, making a histologic subdivision in dermoid cyst-dermoid type, dermoid cyst-epidermoid type, and a third type, dermoid cyst-teratoid type. When gastric or intestinal mucosa is present, such a cyst is classified as a gastric-mucosal cyst. The dermoid cyst, entrapment of epithelium during the embryonic phase is the most likely explanation. [4]

The origins of dermoid cysts are embryological. It has been suggested that dermoid cysts are derived from epithelial debris or nests that are trapped during the midline closure of the first and second brachial arches. Shafer et al attributed the non – epithelial structures that are found in the dermoid cyst, to the entrapment of totipotent cells in the midline during the closure of the first and second arches-a view also shared by Ettinger and Manderson – who however, differentiated dermoid cyst into congenital and acquired. The congenital type arose during the developmental fusion in the body, while the acquired type resulted from some previous injury that drove epithelial cells into the dermis.[59]

Clinical features:

Epidermoid and dermoid cysts usually occur in the floor of the mouth and seldom in the tongue itself. Just a few cases have been reported of lingual involvement. The clinical aspect is not characteristic and consists merely of a cystic swelling of the tongue, which may result in macroglossia. In some cases a sinus tract cyst is present. The head and neck lesions account for 7% of all dermoid cysts and of these 23% are located in the floor of the mouth, but their proximity to the tongue may make it difficult to distinguish the exact location. They occur as rare entities in the midline of tongue.[4]

The lesion is usually located in the body of the tongue more anterior than the thyroglossal duct cyst, but it otherwise is indistinguishable except on histological examination.[4]

Histopathology:

The lining of an epidermoid cyst usually consists of stratified squamous epithelium without adnexal structures and without a muscularis mucosa. In some cases respiratory epithelial metaplasias have been reported. When

adnexal structures, such as sebaceous glands, are present, a diagnosis of dermoid cyst is made. When gastric mucosa is present, the cyst is classified as a gastric mucosal cyst. According to some authors, the presence of a wall of smooth muscles is strong evidence of an alimentary or gastrointestinal tract cyst. [4]

Diagnosis:

In fine needle aspiration cytology contents of the cyst are often keratinous, caesous, sebaceous, or purulent with hair, nails, fat globules, cholestene and even cartilage.

Diagnostic imaging of the lesion includes MRIs, CT scans, and ultrasonography; contrast medium radiographs may be helpful in delineating the extent of the lesion. [60]

Differential diagnosis:

This includes ranula, blockage of a submandibular salivary duct, cystic hygroma, thyroglossal duct cyst, branchial cleft cyst and a detached bronchogenic cyst.[59]

Treatment:

The only effective treatment for sublingual dermoid is surgery, consisting of complete enucleation.[59]

Recurrence is uncommon. Malignant transformation into squamous cell carcinoma has been reported only rarely.[22]

CHORISTOMA OF TONGUE

Choristoma, as defined by **Chou et al**, is a tumor-like growth of otherwise normal cells in an abnormal location.[61]

Choristoma (aberrant rest or heterotopic tissue) is defined as histologically normal tissue proliferation which is not normally found in the anatomic site of proliferation. If the ectopic tissue contains elements from more than one germ layer, they have traditionally been called as teratoma.[62]

Choristoma with proliferation of chondroid tissue is known as cartilaginous choristoma. Cartilaginous and osseous choristomas of oral soft tissue are rare lesions. [62]

Oral glial choristoma is generally reported in the palatopharyngeal complex area of infants and children without gender and ethnic susceptibilities. The tongue is the second most frequently reported site. [63]

Origin:

This extra skeletal proliferation of bone and cartilage in oral and maxillofacial soft tissue reflects the multipotential nature of primitive mesenchymal cells in this region. Usually it is developmental in origin; some of these proliferations seem to occur as a result of local trauma. [62]

Cartilaginous choristoma represents a clinical and pathologic condition of debatable origin. Several hypotheses have been proposed to explain the occurrence of choristoma of tongue. These include:

1. Origin from cartilaginous embryonic rests,
2. Metaplastic chondroid tissue,
3. Derived from pluripotent cells,
4. Neoplasm or Teratoma with preponderance of cartilage and
5. Mixed salivary gland tumor with predominance of cartilage. [62]

The 'origin from embryonic rests' theory postulates that the lesion originates from heterotopic cartilage remnants from any of the first four branchial arches. It is believed that multipotential cells are misplaced during development and sequestered in the tongue. This theory explains the wide distribution of cartilaginous choristomas within the tongue.

Another possible embryonic origin is from remnants of Meckel's cartilage. [1] Origin from embryonic rests is one of the most probable explanations, because of the close proximity to foramen cecum. In the complex embryologic development, multipotential cells may become embedded and produce unusual proliferative lesions some years later. [64]

The metaplastic theory involves the differentiation of pluripotential mesenchymal cells into chondrocytes or cartilaginous metaplasia of the connective tissue. This metaplasia is probably stimulated by trauma or chronic inflammation. Some investigators consider the lesions located on the dorsum of the tongue as choristomas, whereas those on the lateral border of the tongue are metaplastic. [64] The metaplastic theory can be supported by the fact that some reported cases follow trauma or episodes of chronic inflammation. This type of cartilage producing neoplasm is usually seen on the edentulous alveolar ridge of a denture wearer, and is known as Cutright tumor. However cartilaginous choristoma can occur within various areas of tongue, with greatest predilection for the middle dorsal aspect. [62]

The embryological disturbance related to the pathogenesis of glial choristoma located in the tongue seems to be different from brain heterotopias in other sites. The nasal and palatopharyngeal lesions may arise from herniation of neural tissues through an arrested closure in the osseous cranium or by an initial overgrowth of the developing neural tube preventing closure of the cranial opening. On the other hand, the glial choristoma of the tongue may have a different pathogenesis. According to the current theory, the tongue musculature is derived embryologically from occipital myotomes that differentiate into myofibroblasts that migrate to the stomatodeum. Lingual heterotopic glial tissue masses might develop from a nest of pluripotent cells, which become separated before complete fusion of the neural tube, and integrate within those migrating myoblasts to finally reach the tongue.[63]

Clinical features:

Choristoma is a lesion characterized by the presence of tissue not normally found in a site. The most frequent site to be involved is dorsum of tongue followed by lateral border and ventral surface. In the tongue, choristoma may consist of proliferation of bone, cartilage, fat, neural and glial tissue, thyroid gland, respiratory, gastric or intestinal mucosa. Weitzner et al preferred the term cartilaginous choristoma because it indicates the true nature of this lesion in the

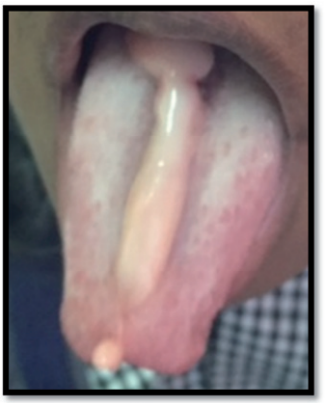
tongue.[64] Cartilaginous choristomas of oral soft tissue were first described in 1913. They occur most frequently in tongue and less commonly in other sites in the oral cavity. They may exist in a pure form or as mixed lesion like lipocartilaginous choristoma or osteocartilaginous choristoma. [62]

Clinically they present as a painless firm nodule and may produce local dysfunction. They occur most frequently in tongue and less commonly in other sites, such as buccal mucosa, soft palate, and gingiva.[62]

Till date 24 pure cartilaginous choristomas of the tongue have been reported in the English language literature. [62]

Centscheff, in his autopsy study found islands of hyaline cartilage in the tongues of nine out of twenty-seven autopsies performed. Among them three were newborn, one was a young child and five were adults. [62]

The osteochondromatous type is the least common type of choristoma of the tongue. The lesion presents as a round, lobulated, firm to hard, movable or fixed, variably elevated, lesion about 1 cm in diameter (range 0.3 to 5.0 cm). The osseous choristoma is frequently located posterior to the circumvallate papillae or close to foramen caecum. It also may be located in the buccal mucosa or soft palate. The cartilaginous choristoma does not produce any specific symptoms. The lesion may be present for an average of 8.2 years. Osteocartilaginous choristomas have a benign clinical behaviour. [64]



Choristoma of tongue

Histopathology:

H & E stained sections of cartilaginous choriostoma show mature chondrocytes arranged in a lobular pattern within a hyaline matrix enclosed by a fibrous perichondrium. [61]

Histologically, diverse histopathologic features are noted in most of the cases of oral glial choristoma. Some reports mention only the presence of neuroglial tissue without further description. Pure neuroglial tissue intermixed with neuron cells, with or without choroids plexus and clefts lined by ependymal- type epithelium are described. In reported cases neuron axons intermixed with cells resembling astrocytes, oligodendroglial and microglial cells have been observed. The neuroglial tissue is intensely positive for GFAP and S-100 and weakly positive for NSE and vimentin.[63]

Differential diagnosis:

The differential diagnoses of cartilaginous choristoma are pleomorphic adenoma, soft tissue chondroma, traumatic chondroid neoplasm and ectomesenchymal chondromyxoid tumor (ECT). Pleomorphic adenomas contain both epithelial and mesenchymal components in characteristic chondromyxoid background. Traumatic chondromatous metaplasia or Cutright tumor is unusual in this site. The ECT is a distinctly uncommon neoplasm that develops mainly in the anterior dorsal tongue. Microscopy of this tumor shows lobular proliferation of ovoid and fusiform cells with occasional foci of atypia in a chondromyxoid background with infiltrating margins. Absence of these features excluded ECT. [62]

Treatment:

The treatment of the lesion is surgical excision. [62]

MIDLINE FISTULA

A fistula can be defined as a pathologic tract in the tissues, usually lined by epithelium. The cause can be either of an inflammatory or, as is nearly always the case in a midline fistula of the tongue, of a developmental nature. A midline fistula of the tongue is an exceptional developmental defect, with just an occasional case being reported. [4]

Clinical features:

The fistula reported in a 19 year old man was located in the anterior two-thirds of the tongue from which a small quantity of yellow material could be expressed and was believed to be the result of malunion of the two lateral lingual swellings during embryologic development. [66]

Other cases encountered of a midline fistula of the tongue: One was in a 26-year-old woman in the anterior part of the dorsal surface; the other was an incidental finding at autopsy in a 55-year-old man. In the latter case the fistula was located just anteriorly to the foramen cecum. The tract was lined by nonkeratinizing stratified squamous epithelium. [4]

Treatment:

The fistula was excised after the tongue had been split in the midline and the tract completely separated from the surrounding musculature.[66]

LYMPHANGIOMA

Lymphangioma is a benign lesion, characterized by proliferation of lymphatic vessels. It is a hamartoma rather than a neoplasm. Some authors use the term lymphangioma circumscriptum. In some cases it is difficult to make the distinction between a lymphangioma and a hemangioma, in which case the term lymphangio-hemangioma, or just angiomatosis, may be used. [4]

Lymphangioma of the tongue was first described by Virchow in 1854 and in 1872 Krester hypothesized that hygromas were derived from lymphatic tissue. Occurrence in the oral cavity is rather rare, the tongue being the site of predilection.[67]

Lymphangioma is generally considered to be a disease of childhood during which time there is an active lymphatic growth. Lymphangiomas can occur from birth to the first 2 years of life. The predominant site is head and neck region and this may be related to the anatomic locations of the embryologic sacs.

Classification:

Watson and McCarthy classified lymphangioma based upon their 41 cases

1) Simple lymphangioma
2) Cavernous lymphangioma
3) Cellular or hypertrophic lymphangioma
4) Diffuse systemic lymphangioma
5) Cystic lymphangioma or hygroma [26]

It can be classified into 3 subtypes

1. Lymphangioma simplex, which consists of capillary sized lymphatic channels
2. Cavernous lymphangioma, with dilated lymphatic channels
3. Cystic lymphangioma composed of large, macroscopic cystic spaces. [22]

Pathogenesis:

Two major theories have been proposed to explain the origin of lymphangiomas.

The first theory is that the lymphatic system develops from five primitive sacs arising from the venous system. Concerning the head and the neck, endothelial outpouchings from the jugular sac spread centrifugally to form the lymphatic system. Another theory proposes that the lymphatic system develops from mesenchymal clefts in the venous plexus reticulum and spreads centripetally towards the jugular sac. Finally, lymphangiomas develop from congenital obstructions or sequestrations of the primitive lymphatic enlargement.[68]

Based on various hypotheses concerning lymphangiogenesis published in the literature, different putative mechanisms of lymphangioma development are discussed including failure of the lymphatic system to connect with or separate from the venous system, abnormal budding of the lymphatic system from the cardinal vein, or acquired processes such as traumata, infections, chronic inflammations, and obstructions. Increasingly, the possible influence of lymphangiogenic growth factors on the development of lymphangiomas is being discussed. The proved expression of different growth factors in the endothelium of lymphangiomas leads to new hypotheses regarding the pathogenesis of lymphangiomas. Thus, further studies on lymphangiogenesis and the development of lymphangiomas will have to clarify as to whether lymphangiomas are true malformations or neoplastic in nature. [69]

Clinical features:

In a series of 46 cases of lymphangioma of the tongue, 75% of patients were less than 20 years of age; a number of cases were diagnosed at birth, and sex predilection not observed.[6]

The majority of lymphangiomas are located on the dorsal surface of the anterior part of the tongue. The size may vary from pinhead dimensions to massive lesions involving the entire tongue and surrounding structures. The typical lymphangioma is characterized by irregular nodularity of the surface of the tongue with gray and pink grapelike projections.[4]

Litzow and Lash reviewed a series of 46 cases of lymphangioma of tongue and pointed out that the anterior dorsal part of the tongue was most frequently affected. The superficial lesion is manifested as a papillary lesion which may be of the same color as the surrounding mucosa or of a slightly reddish hue.[26]

Usually, the lesions present superficially as a pebbly, vesicle-like feature with so-called ‘frog-egg’ or ‘tapioca-pudding’ appearance. [70]

The deeper lesions appear as diffuse nodules or masses without any significant change in the surface texture or color. The irregular nodularity of the surface of the tongue with gray and pink projections is the commonest sign of the disease, and when associated with macroglossia, is pathognomonic of lymphangioma. [26]

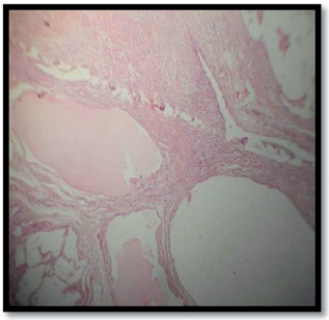
Most lingual lymphangiomas are simple or cavernous lymphangiomas that occur on the dorsum of the tongue. Secondary hemorrhage in lymphangiomas is not a rare occurrence. Cystic hygroma is a large extension of the same type of lesion. It is more common in the neck. When it is located in the upper part of the neck, it has a tendency to invade and involve the floor of the mouth and the tongue.[4]

A history of recurrent swellings can be considered more or less diagnostic of lymphangioma. Inflammatory edema may occur secondary to upper respiratory tract infections. In severe cases of long duration, an open-bite malocclusion may occur. Speech and mastication may be hampered as well.[4]

Histopathology:

The lymphangioma consists of multiple, interwining lymph vessels in a loose fibrovascular stroma. Cavernous lymphangioma is the most common type consisting of numerous dilated lymphatics; a single layer of endothelial cells with flattened, occasionally plump nuclei and containing lymph. Those vessels just beneath the surface epithelium tend to fill or replace the connective tissue papillae, perhaps producing a papillary surface change. The superficial lesion has little or no fibrous stroma separating it from the overlying epithelium. Occasionally the lesion may be filled with blood, a mixed haemangiolymphangioma. Occasional lesions demonstrate proliferation of lymphatic channels with another connective tissue component, primarily smooth muscle cells called lymphangiomyoma.

Histologically, endothelial-lined spaces are found in the connective tissue. In some cases the spaces contain elements of blood, making the distinction between lymphangioma and hemangioma difficult or even impossible. Usually, a lymphangioma has a cavernous architecture like many hemangiomas. There is no true encapsulation, and quite often the proliferation of lymphatic vessels spreads diffusely into the surrounding soft tissues. [4]



Lymphangioma

Differential diagnosis:

Lymphangiomata may be similar to a number of oral lesions including haemangioma, teratoma, lingual thyroid, dermoid cyst, thyroglossal duct cyst, heterotopic gastric mucosal cyst, and granular cell tumour. [70]

Management:

During inflammatory episodes the combined use of corticosteroids and antibiotics may be required. Small lesions can be excised. In extensive symptomatic lymphangiomas, surgical removal of the bulk of the lesions seems to be the only possible treatment.

Complete surgical excision is difficult, particularly in the mouth area, because of the multiple finger-like projections extending into the adjacent tissues. Steroids and antibiotics should be administered during the surgical and post surgical periods. Recurrences are not uncommon.

In the past radon seeds were used occasionally. Today, however, radiotherapy for this benign condition is never applied because of possible adverse changes to the tissues. [4]

CONGENITAL HEMANGIOMA AND CONGENITAL VASCULAR MALFORMATION

A hemangioma of the soft tissues is either a neoplasm or a malformation. Actually, most of the hemangiomas in the oral cavity are of developmental nature. In some instances, lesions seem to be a mixture of hemangioma and lymphangioma, resulting in such terms as hemangio-lymphangioma and angiomatosis. Hemangioma-like lesions may also be a manifestation of Kaposi's sarcoma. Above the age of 40 years, solitary or multiple, bluish, hemangioma-like changes may occur in the oral and lingual mucosa as the result of widening veins. [4]

Etiology:

The cause of vasoformative tumors is unknown. One hypothesis postulates that a placental cell, such as the trophoblast, may be the cell of origin for hemangiomas. Therefore, hemangiomas may arise secondary to some event in utero. However, the relationship between hemangiomas and placental tissues needs further investigation.[26]

Pathophysiology:

Vascular malformations need to be understood in terms of their embryology and development. The classic sequence of events usually falls into three stages.

- The undifferentiated capillary network stage
- The retiform developmental stage
- The final developmental stage

In the undifferentiated capillary network stage, the primitive mesenshyme is nourished by an interlacing system of blood spaces without distinguishable arterial and venous channels. Separate venous and arterial stems appear on either side of the capillary network in the retiform development stage. The retiform developmental stage begins at about 48 hours of embryonic development. The final developmental stage begins at two months of development and involves the gradual replacement of the immature plexiform network by the mature vascular channels. [26]

The more common capillary hemangioma represents an arrest in the development of the mesenchyme primordia in the undifferentiated capillary network stage. As differentiation progresses, primitive vessels penetrate deeper into the subcutaneous

layer, the muscle, or the bone tissue and give rise to capillary hemangiomas. Termination of development in the retiform developmental stage may produce venous, arterial, or capillary malformations because this stage is characterized by an establishment of venous, arterial, and capillary systems. In the final developmental stage, the maturation of the venous and lymphatic systems predominates. Aberrations in this mature stage of development result in venous malformations and lymphangiomas.[26]

Takahashi has outlined a number of cellular markers that distinguish the phases of hemangiomas; these markers include tissue metalloproteinase (TIMP-1), bFGF, proliferating cell nuclear antigen, type IV, VEGF, and urokinase. [26]

Classification: The hemangioma can be classified into two types:

1. The congenital hemangioma: This is a common vascular tumor of infancy that gradually involutes during adolescence.
2. Vascular malformation: This is present at birth but never regresses. This type occurs in the tongue. [3]

Watson and Mc Carthy

1. Capillary hemangioma
2. Cavernous hemangioma
3. Angioblastic/hypertrophic hemangioma
4. Racemose hemangioma
5. Diffuse systemic hemangioma
6. Metastasizing hemangioma
7. Nevus vinosus or Portwine stain
8. Hereditary hemorrhagic telengiectasis [26]

Mulliken (1982)

1. Capillary
2. Cavernous
3. Capillary-cavernous/mixed

Waner and Suen (1999)

1. Superficial
2. Deep [26]

Biological classification

Hemangioma	Malformation
Proliferative phase	Capillary
Involutive phase	Lymphatic
	Venous
	Arterial
	Combined

Clinical features:

Congenital hemangiomas of the oral cavity show a strong preference for occurrence in the tongue and the floor of the mouth. A hemangioma of the tongue may affect just a part of the tongue or the entire tongue, producing macroglossia. Rather rarely the base of the tongue is involved. In some cases multicentric hemangiomas are present. The color of a hemangioma may vary from bluish to purple or fiery red. The texture of the mucosa may be more or less unchanged, showing just an increased vascularity on the surface, but in other cases the appearance is pebbly.[4]

Lingual hemangiomas pose distressing problems to the patients, producing cosmetic deformity, recurrent hemorrhage, and functional problems with speaking, deglutition, and mastication.[71]

Pain is not a prominent feature, except in cases of traumatization or secondary inflammation. In severe cases there may be loss of mobility of the tongue. Bleeding, either spontaneously or due to mechanical irritation, can be a serious problem.[4]

Hemangioma of the base of tongue is rare. Qureshi Sajid S et al reported a case of hemangioma of base of tongue extending to the supraglottis, which necessitated an extended supraglottic laryngectomy. [71]

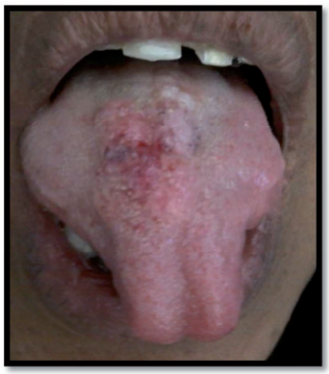
Brown et al reported a case of mixed capillary and cavernous hemangioma of tongue in a 76 year old woman, which was present since birth and was

asymptomatic for more than 50 years. The patient had a dislocated jaw presumably due to mass effect of hemangioma, which started growing after 56 years. This hemangioma rapidly enlarged after an unrelated operation under spinal anesthesia causing severe functional and cosmetic deformity, for which surgical treatment was required. [71]

Syndromes associated with haemangioma are

- Rendu-Osler Weber Syndrome
- Sturge-Weber-Dimitri Syndrome
- Kasabach- Merritt Syndrome
- Maffucci Syndrome
- Von Hippel- Lindau Syndrome
- Kilppel- Trenaunay- Weber Syndrome [26]

A hemangioma or just telangiectasia may also be part of a generalized condition such as hereditary hemorrhagic telangiectasia (Rendu-Osler-Weber syndrome). The spider like telangiectasia usually first manifests during adolescence. The lingual lesions rarely cause any discomfort. In encephalo- trigeminal angiomatosis (Sturge-Weber disease), the tongue is usually not involved, in the rare angiokeratoma corporis diffusum universale (Anderson-Fabry disease), lingual involvement has been reported occasionally. Also, a vascular malformation of the tongue has been described in a patient with von Willebrand's disease. In most instances the diagnosis of hemangioma is based on clinical judgment.[4]



Hemangioma of tongue

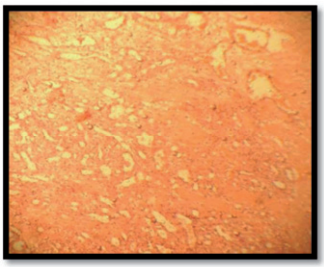
Histopathology:

Congenital hemangiomas are composed of abundant capillary spaces lined by endothelium without muscular support. Congenital vascular malformations may consist not only of capillaries, but also of venous, arteriolar, and lymphatic channels. Lesions may be of purely one type of vessels, or they may be combinations of two or more. [49]

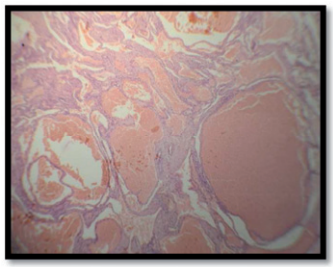
A hemangioma may consist of numerous irregular, blood-filled spaces lined by endothelial cells and surrounded by connective tissue. When a large number of proliferating endothelial cells lining small capillaries are present, the lesion is referred to as a capillary hemangioma. [4]

In the case of large dilated blood sinuses, the term cavernous hemangioma is applied. The cavernous hemangioma and a lymphangioma may occasionally be indistinguishable from each other, both clinically and histologically. The cavernous type tends to be somewhat more circumscribed than the capillary one. However, a true capsule is absent. There are no signs of cellular or nuclear pleomorphism. Mitoses are rare. In case of ulceration and, thereby, the presence of inflammatory cells, it may be impossible to distinguish a hemangioma from a pyogenic granuloma. [4]

When hemangiomas undergo regression, extensive sclerosing can occur, especially in the central portion of the lesion. It is debatable whether such a lesion should be classified as a separate entity by applying the term sclerosing hemangioma. In long-standing cases, thrombus formation may take place in a hemangioma, followed by calcification. Such concrements are called phleboliths. [4]



Capillary hemangioma



Cavernous hemangioma

Diagnosis:

The characteristic and extent of lesions situated in the deep portion of the tongue, such as its base, or submucosal lesions can be recognized only on cross-sectional CT scan or MRI. Hemangiomas usually appear as a well-demarcated enhancing mass often containing phleboliths on CT scan. MRI shows the lesion as a solid mass with isointense or slightly high signal intensity to muscle on T1-weighted images and heterogeneous signal intensity on T2-weighted images. Post contrast T1-weighted imaging commonly demonstrates prominent enhancement. Either CT scan or MRI should be done before a biopsy. [71]

Management:

The treatment of congenital vascular anomalies is based on an understanding of the clinical behavior and natural history of individual lesions.

The majority of hemangiomas does not require treatment and regress spontaneously during childhood. In the case of a large, persisting, or even growing hemangioma, therapeutic management is a difficult problem. [72]

A conservative approach seems justified, taking care of limited areas that produce bleeding. In these large and diffuse lesions the use of cryosurgery or laser therapy does not seem to be effective. Injection of sclerosing agents was advocated years ago but is rarely used today. Even injection of boiling water has been mentioned in the past

Corticosteroids and interferon form the first line of treatment. Vincristine and bleomycin are considered for problematic hemangiomas in infants, which fail to respond to steroids.

The technique of selective percutaneous embolization before surgery has emerged as a valuable adjunct to surgery. Radiotherapy should be avoided at any rate because of the possible late adverse consequences of that therapy. [72]

FORDYCE'S SPOTS

Fordyce's spots, or Fordyce's condition, are names given to ectopic sebaceous glands occurring in the oral mucosa. Fordyce first described this condition in 1896. It is a harmless condition of a developmental nature occurring in almost all adults, although it is somewhat more frequent in males than in females. The prevalence reaches a peak at 20 to 29 years of age. No function can be assigned to the ectopic sebaceous glands.[4]

Clinical features:

The sebaceous glands, clinically appearing as yellowish, slightly elevated granules, are usually found in the buccal mucosa and upper vermilion border with a more or less symmetrical distribution. They are asymptomatic. The diagnosis is based on clinical judgment.

Occurrence of Fordyce's spots in the tongue is exceptional. The lesions in the tongue have been described as a solitary dome-shaped nodule from a few millimeters up to 2 cm in diameter on the midline dorsum of the tongue. [4]

A 28-year-old dentist noticed a 9x5mm yellow submucosal plaque in the midlateral dorsum of his tongue. After a period of observation an excisional biopsy was performed. The histopathologic examination of the specimen revealed normal sebaceous glands opening onto the tongue surface. This occurrence of ectopic sebaceous glands in the tongue is notably rare in contrast with their frequent occurrence in the lips and buccal mucosa.[73]
Kovero O noticed the presence of a sebaceous gland in the dorsal surface of the tongue. [74]

Intraoral sebaceous carcinoma (SC) is a rare tumor in the oral cavity, thought to arise from malignant transformation of oral sebaceous glands. There are only six cases of intraoral SC that have been reported in the English language literature.[75]

Histopathology:

Microscopically, lobules of sebaceous glands are aggregated around and adjacent to excretory ducts. The heteropic glands are well formed and appear normal.[49]

Differential diagnosis:

Large numbers of lobules coalescing into a definitely elevated mass may be called benign sebaceous hyperplasia, and occasional small keratin-filled pseudocysts may be seen and must be differentiated from epidermoid cyst or dermoid cyst with sebaceous adnexa. The pathologist must be careful to differentiate such lesions from salivary neoplasms with sebaceous cells, such as sebaceous lymphadenoma and sebaceous adenoma, and their malignant counterpart's sebaceous lymphadenocarcinoma and sebaceous carcinoma. [76]

Management:

No treatment is required. [4] Inflamed glands can be treated topically with clindamycin. When surgically excised, recurrence does not occur. Neoplastic transformation is very rare but has been reported.

TERATOMA

Teratomas are defined as true neoplasms composed of multiple tissues foreign to the site from which they originate.[77]
Teratoma arising from the tongue is very rare and a literature search showed only 14 other reported cases since it was first reported in 1966. [78]

Teratomas are composed of tissues of all 3 germ layers and show varying degrees of differentiation. These tumors arise from pluripotent stem cells and ectopic embryonic nongerm cells. Teratomas of the head and neck are rarely encountered and account for less than 5% of reported cases. Teratomas of the tongue are extremely rare, with only a few cases reported. [79]

Embryologically, they are thought to occur as a result of displacement of cells from normal tissue during fetal life. In the tongue, they are thought to occur as a result of a displacement of cells from the tuberculum impar. [79]

Classification:

Teratomas have been classified into four subtypes. These are:

1. Dermoids, which contain tissues of mesodermal and ectodermal origin;
2. Teratoids, which contain tissue from three primary germ layers but are poorly-differentiated;
3. True teratomas, which are similar to teratoids but differentiated into recognisable tissues histologically; and
4. Epignathi, which are also tridermal in origin, but differentiated into recognisable organs, sometimes with limbs or even a visible second foetus. [78]

Clinical features:

Teratomas of the head and neck are uncommon; they typically emerge during the neonatal period and are associated with airway obstruction and high infant mortality rate. However, teratomas of the tongue rarely produce respiratory difficulties. [79]

Teratomas can be detected prenatally. Teratomas of the oral cavity may be associated with polyhydramnios in utero, secondary to foetal pharyngeal obstruction. Tongue teratoma usually gives rise to feeding difficulties rather than airway problems. Tongue teratoma may also present with elevated maternal serum alpha-foetal protein. This elevation may be due to the presence of abundant extramedullary haematopoietic liver tissues. Most congenital teratomas are benign. Teratomas of the head and neck that occur in adults are usually malignant. [78]

In the literature a case of a gliomatous teratoma occurring in the tongue of a newborn has been reported. [6]

Embryologically, teratomas of the head and neck are thought to occur as a result of a displacement of cells during gestation. Miller and Owens speculate that teratomas arising from the tongue are exceedingly rare with only a few reported cases in the literature. In a review of 12 reported teratomas of the tongue, it was found that all presented at birth, with some measuring up to 12 cm in diameter, a dimension reached by this reported case.

Teratomas are composed of various tissues of ectodermal, endodermal, and mesodermal origin, and may exhibit variable levels of maturity. Neuroectodermal tissues dominated the currently described case, although all 3 germ cell layers were represented. Head and neck teratoma in adults is usually malignant; however, most congenital teratomas are benign, despite the frequent presence of histopathologically immature components. Immature tissues do not necessarily denote a malignancy in this context. However, the malignant potential of a teratoma of the tongue is not fully defined, given the paucity of reported cases and limited follow-up data. On the basis of the available literature, the case described in this report was classified as a benign immature teratoma.

Histopathology:

The histology of that case showed a variety of tissue elements, such as cartilage, bone, mucus-secreting glands, sebaceous glands, stratified squamous epithelium, and hair follicles in a cystic lesion. [6]

Histopathological examination of reported cases of teratomas of the tongue showed that the tumours were all benign in character, with no evidence of malignancy despite histological immaturity. [78]

According to the classification of dermoid cysts presented by Meyer in 1955, that case also can be classified as a teratoid-type dermoid cyst. Strictly speaking, the case of heterotopic brain tissue, found in the tongue of a 4-day-old infant, is not a teratoma since only central nervous tissue elements without endodermal or mesodermal components were present. [6]

Differential diagnosis of tongue lesions includes thyroglossal duct cyst, lingual thyroid, lymphangioma, hemangioma, dermoid cyst, granular cell myoblastoma and heterotopic gastric mucosal cyst. Computerized tomography is useful in differential diagnosis and defining extent of disease. [80]

Treatment:

The aim of surgical management was to remove the diseased mass as well as to provide good airways in the long term. Retrospectively, the tumor would have been better removed with CO2 laser**.**

Definitive treatment of teratoma of the tongue is complete surgical excision. Surgery is curative and the prognosis is excellent. Recurrence has not been reported. [78]

CONCLUSION

The oral cavity is the site of many neoplasms, reactive processes, infections and manifestations of systemic disease. The primary objectives of an oral cavity examination are to distinguish between health and disease and to recognize normal anatomic structures and their variations. The tongue shows a wide degree of clinical changes in both systemic and local disease processes. It has been used as an indicator of health for centuries.

The inspection of tongue was first described in the third and fifth century BC and was believed as important as the palpation of pulses. The tongue changes are believed to be closely associated with disease.

The development of the tongue in the embryo reveals its close relationship with the origin of human organism from the three germ layers ectoderm, mesoderm and endoderm. Thus, the tongue is an ontogenic key organ for understanding the morphological and physiological intercommunication within the body. Moreover, the human tongue is an essential element of human culture in general. Languages and intellectual conceptions as well as the entirety of verbal terminology start with the tongue; furthermore, the human tongue represents an important diagnostic medium and is a valuable instrument of examination for well trained physician. A skillful diagnosis of the tongue structures incorporates an essential supplement of a medical diagnosis, Western or Chinese, as far as an integration of the whole individual and his or her disease is concerned.

Clinical appearance of the tongue in health and disease can be evaluated through its colour, size, position, movement, muscle toxicity and anatomical landmarks. However, recent studies have shown that it is not only the digestive system that can reflect on the tongue, but also a variety of other disease, both systemic and local in origin.

Considerable advances have been made in the categorization of disease of the tongue and significant progresses have been made in the recognition of lesions and the knowledge about their behaviour and management. The current state of knowledge of the clinical, etiological, histopathologic and therapeutic aspects is analysed.

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