

Developmental Disturbances of Oral and Para-oral tissues

Intended Learning Outcomes (ILOs):

By the end of these lectures every student will be able to:

- Recognize the normal development of oral and para-oral tissues
- Identify the developmental disturbances of oral and para-oral tissues
- Determine the pathogenesis of these developmental disturbances
- Describe the clinical significance of these developmental disturbances

Normal development of face and lips:

Nasal process descends from frontal bone forming 2 lateral nasal processes and one medial nasal process which is longer. Fusion occur between the lateral nasal processes and maxillary processes on both sides starting from the inner canthus of the eye and the median nasal process fuse with 2 maxillary process bilaterally to form the upper lip. The 2 mandibular process fuse in midline to form the lower lip.

Developmental disturbances of the face

I- Oblique facial cleft

This is a developmental cleft start from the inner canthus of the eye to the ala of the nose or upper lip. It is due to failure of fusion between maxillary and lateral and medial nasal processes. It is usually unilateral but may be bilateral in about 20% of cases.

II- Transverse facial cleft:

This is a cleft running from the angle of the mouth towards the ear. It is due to failure of fusion between the maxillary and mandibular processes, and may be unilateral or bilateral, partial or complete (rare), extending from the angle of the mouth to the ear.

III- Macrostomia and Microstomia:

Macrostomia: it is an excessively large mouth. It is due to premature arrest of fusion between maxillary and mandibular processes bilaterally.

Microstomia: it is an abnormally small mouth. It is due to excessive fusion between maxillary and mandibular processes.

IV- Facial Hemihypertrophy:

This is a congenital malformation in which one half of the face and jaws is abnormally larger than the normal. This may be limited to the face and jaws or may be combined with enlargement of one half of the whole body. It is of unknown etiology but vascular or heredity factor have been suggested. The teeth on the affected side are abnormally large from the canines to first molars and roots are abnormally large. The tongue is also enlarged on the affected side. There is early shedding of the deciduous teeth with early eruption of the permanent successors on the affected side.

V- Facial Hemihypotrophy:

This is a congenital malformation in which one half of the face and jaws is abnormally smaller than normal. It is of unknown etiology but vascular or heredity factors have been suggested.

VI- Treacher Collins Syndrome (Mandibulofacial Dysostosis):

It is a hereditary disorder transmitted through genes. The gene may be completely expressed showing all abnormalities or incompletely expressed where the deformity is less extensive.

Clinical manifestations:

- 1- Anti-mongoloid slant of eyes
- 2- Notch of the outer third of the lower eyelids, associated with absence of eyelashes of the medial 2/3 of the lower lid.
- 3- Underdevelopment of the facial bones, especially the zygomatic bones and the mandible.
- 4- Malformation of the external ears.
- 5- Macrostomia.
- 6- High arched palate.
- 7- Abnormal dentition with malocclusion.

VII- Pierre Robin Syndrome (Mandibular dysostosis with glossoptosis)

It is a syndrome characterized by hypoplasia of the mandible with falling back of the tongue until obstructing the laryngeal inlet and causing cyanosis and asphyxia.

Clinical manifestations:

1. Mandible is relatively smaller than normal.
2. The angle between ramus and body of the mandible is at right angle or less in contrast to the normal obtuse angle of newborn infants.

The explanation of this disorder:

During intrauterine life the fetus is in abnormal position where there is a back pressure from the sternum on the mandible. If this occurs before the mandible is completely calcified, it may be forced posteriorly causing the observed right angle. This also prevents descent of the tongue, which interferes with meeting and fusion of the palatine processes, causing a cleft palate, which is usually U-shaped. The condition is improved after birth where there is relief of the back pressure from the sternum on the mandible leading the angle to be right or slight obtuse instead of acute and this proves the mechanical cause of this condition.

Developmental Disturbances of the Lip**I- Congenital Lip pits:**

These are congenital blind epithelial-lined small shallow depressions observed more common on lower lip around the midline or at the corner of the mouth. They may represent a less severe expression of the genetic defect leading to cleft lip formation.

II- Congenital Lip Fistula:

This is a channel connecting the minor salivary glands in the lip with the external environment. They may be around midline more commonly on the lower lip or at the corner of the mouth. To differentiate between lip pit and fistula; on pressure, there is saliva oozing from the fistula.

III. Cleft lip:**Etiology**

Congenital cleft lip is hereditary or due to environmental factors such as hypervitaminosis A that affect the fetus in the intrauterine life.

Acquired clefts that occur after birth due to trauma, infection, tumor or other causes are not on the line of fusion and not on the direction of fusion.

Clinical features

Cleft lip may be unilateral or bilateral and may be partial as a notch of the lower border of the lip or complete cleft extending from the floor of the nose to the lower border of the lip. Median clefts of lower lip occur at the site of fusion between two mandibular processes.

IV. Cheilitis Glandularis:

This is a condition of unknown etiology in which the lip is enlarged, firm and everted exposing the openings of the labial mucous glands which become enlarged and the orifices of their ducts become inflamed and dilated giving the labial mucosa a red appearance.

The condition affects males more than females, and is painless. Malignant transformation has been reported.

V. Cheilitis Granulomatosa:

It is a disorder characterized by a chronic, firm swelling, affecting usually the upper lip, but swelling of the lower lip is sometimes seen. This may be associated with **Melkerson-Rosenthal Syndrome** who also suffers from of facial paralysis and fissured tongue.

VI. Peutz-Jeghers syndrome (hereditary intestinal polyposis syndrome):

This is a hereditary condition, in which there is intestinal polyposis associated with circum-oral, circum-nasal and circum-ocular pigmentation, as well as pigmentation of the mucous membrane of the mouth and the skin of the dorsal aspects of the hands and feet. The polyps are manifested by gastrointestinal obstruction and troubles. They present no tendency for malignant transformation.

Developmental Disturbances of the Palate

Clefts are classified into pre-alveolar, alveolar and post-alveolar according to their occurrence in the lip, alveolus or palate respectively.

I. Alveolar cleft:

Cleft of the alveolus may be unilateral or bilateral. It also may be complete, incomplete or mixed. The clefts pass through the region of the upper lateral incisor which may be absent or may develop medial or lateral to the cleft or may be doubled (supplemental teeth).

II. Cleft palate

Normal fusion extends forwards from the incisive foramen to uvula. So if there is failure of fusion; cleft may involve uvula or soft palate and uvula or hard palate, soft palate and uvula.

III. Combination of clefts:

All combinations may be found since union of embryonic processes starts in the region of the incisive foramen; proceeds forward into the lip and backwards into the palate. Cleft may be a complete unilateral cleft extending from the uvula to the incisive foramen in the midline then deviating to one side and meeting the alveolar process in the area of the lateral incisor and then into the lip or cleft may be a complete bilateral extending forward bilaterally from the incisive foramen to the alveolus and lip.

IV. Torus palatinus:

This is a developmental disturbance, frequently hereditary appearing as a single or multiple bony swellings at the midline of the palate. They may be flat, fusiform or lobulated. The covering mucosa is intact but thin. It is composed either of compact bone, or of a core of cancellous bone covered by a layer of compact bone. Its clinical significance is rocking of complete denture.

Developmental Disturbances of the Jaws

I. Agnathia

Very rare developmental disorder in which there may be complete or partial absence of either jaw. Partial agnathia is in the form of absence of ramus, condylar or coronoid processes on one or both sides.

II. Micrognathia

It is a small jaw and may affect either upper or lower jaws. Micrognathia may be true or apparent, when the jaw is in post-normal relationship to the base of the skull. It may be classified into congenital and acquired forms. The congenital form is usually hereditary, while the acquired form is usually due to trauma or infection destroying the condylar growth center at an early age.

III. Macrognathia

This means an abnormally large jaw which may be true or apparent according to the relation to the base of the skull. Cases of macrognathia may be hereditary or acquired as Paget's disease of bone or Acromegaly.

Developmental Disturbances of the Tongue

I. Aglossia

Congenital absence of the tongue is very rare. It is due to failure of development of the two lateral lingual processes and their subsequent fusion with the tuberculum impar. However, there may be a small nodular elevation in the middle of the floor of the mouth representing the tuberculum impar.

Clinical significance: impairment of mastication, deglutition and speech.

II. Microglossia

It is a rare congenital defect in which the tongue is smaller than normal.

III. Macroglossia

It is a large tongue which may be congenital or acquired. The congenital may be due to lymphangioma, haemangioma or fibrous as well as muscular hypertrophy. While, acquired macroglossia is observed in acromegaly and in lymphangioma that results from injury. Acquired macroglossia may also result from tumor as papilloma, lipoma, fibroma, carcinoma or sarcoma as well as due to infection as actinomycosis, syphilis or tuberculosis. To differentiate between congenital and acquired macroglossia; in the acquired type; the edges are scalloped where they fit against the teeth and interdental spaces while the edge of congenital macroglossia is smooth border and when teeth erupted they become spaced and labially inclined.

IV. Ankyloglossia:

This is a developmental defect in which the tongue is of limited movement. It may be **complete ankyloglossia** due to failure of separation of the tongue from the floor of the mouth or **partial ankyloglossia** due to an abnormally short lingual frenum or attached near the tip of the tongue. It may be **superior ankyloglossia** which is associated with cleft palate in which the dorsum of the tongue is joined by a membrane to one side of the cleft. On the other hand, **acquired ankyloglossia** may be due to infection, trauma, malignant neoplasm or radiation involving the floor of the mouth and the ventral surface of the tongue.

Clinical significance: If the condition is severe, speech may be disturbed.

A more serious condition is **hypermobility** of the tongue due to an abnormally long lingual frenum which allow the tongue to slip back and obstruct the airway causing death by suffocation.

V. Cleft tongue (bifid tongue)

This is a congenital defect in which complete or partial cleft (fissure) of the tongue is present running antero-posteriorly from the tip backwards. The condition is due to an arrest of development resulting in failure of the two lateral lingual masses to completely fuse with each other in front of the tuberculum impar.

VI. Median Rhomboidal Glossitis:

The condition is due to persistence of the tuberculum impar on the dorsum of the tongue resulting from failure of the two lateral lingual elevations to completely submerge it before fusing with each other.

It appears as a reddish, depapillated, raised, rounded or ovoid smooth, painless elevation situated on the dorsum of the tongue anterior to the foramen caecum (patients should be assured that it is a congenital condition and does not present any harm such as malignancy).

The term median rhomboid glossitis is a misnomer, since it is rarely rhomboidal in outline and it is not of an inflammatory condition but it is developmental and superimposed with inflammation.

VII. Lingual thyroid nodule:

Development of the thyroid gland involves migration of epithelial cells (thyroglossal tract) from the foramen caecum of the tongue to the neck where it is differentiated into thyroid secreting tissue. Deficient migration may occur at the area of the foramen caecum leading to thyroid tissue being restricted to its site of origin giving rise to lingual thyroid. It is considered as an **ECTOPIC** condition. It appears as a nodule slightly elevated from the surface of the dorsal surface of the tongue, it is usually yellowish white.

It may transform to malignant tumor (adenocarcinoma). Verification of the presence of normal thyroid tissue in the neck should be done by scanning the neck through injection of radioactive iodine. If it is present in its normal site, removal of the thyroid nodule is advisable. If there is absence of thyroid gland, then implantation of the thyroid nodule as endocrine gland is performed in any vascular area in the body.

VIII. Geographic tongue (glossitis areata exfoliativa, erythema migrans, benign migratory glossitis):

It is a condition of unknown etiology which mostly affects young unhealthy children, though sometimes observed in older individuals, especially women under stress.

Clinical appearance:

There are circular or oval red patches on the anterior two-thirds of the tongue due to disappearance of the filiform papillae, but the fungiform papillae may be retained and appear as red dots. There is slightly elevated yellowish border due to hypertrophy of filiform papillae. When a patch reaches the lateral border of the tongue, it disappears and when an advancing margin of one patch meets that of another, one of them may recede while the other continues to advance or they may coalesce. Each patch has a life history of about one week after which it disappears, only to reappear at another area.

IX. Hairy tongue (Black hairy tongue):

It is a condition of unknown etiology in which there is extensive proliferation of the filiform papillae. There may be an actual hypertrophy of the papillae or there is impaired desquamation. The hypertrophic papillae may cover the whole dorsum of the tongue or only a part of it. They vary in color from yellowish to brown or black. The color is due to some extrinsic factor from foods or drugs or from chromogenic bacteria.

X. Fissured tongue

This is a condition in which the tongue presents symmetrical or semi-symmetrical grooves on its dorsal surface. This is not a developmental condition. It may appear at any time. **Causes** may be chronic trauma, vitamin deficiency, tobacco smoking, psychological stress or infections such as syphilis.

Clinical appearance

The pattern of fissures may be transverse, cribriform or foliaceous or irregular and the depths of the grooves are devoid of papillae thus may be irritated with food constituents leading to inflammation. The individual who suffers from fissured tongue is advised; while brushing teeth; to clean his/her tongue with soft tooth brush. On microscopic examination the epithelium is frequently seen to be hyperplastic with a chronic inflammatory cell infiltrate.

Developmental Disturbances of the Oral Mucosa**I. Fordyce Granules:**

This is an **ECTOPIC** condition in which sebaceous glands develop in the oral mucosa which is normally devoid of them as sebaceous glands normally develop in skin in relation to hair follicles.

Clinical appearance:

Single or multiple pinhead-sized spots appear symmetrically located most frequently in the buccal mucosa in the molar region, but they have been observed also on the mucous side of the lips. The covering mucosa may be smooth or slightly elevated.

II. White Spongy Naevus (white folded gingivostomatitis):

Naevus is hamartoma in skin or mucous memberane.

This is a hereditary condition of the oral mucosa which is sometimes congenital, appears in early childhood and persists for life.

Clinical appearance:

It is grayish-white spongy areas of the oral mucosa with folds and it is soft upon palpation. It often exhibits a symmetric wavy pattern. The entire oral mucosa may be involved, but the most frequently affected regions are the buccal mucosa, the floor of the mouth and the ventral surface of the tongue.

Histopathology:

Prickle cell layer shows acanthosis (increase in number of layers) with inter- and intra-cellular oedema or vacuolation giving the **basket-weave** appearance with surface hyperkeratosis.

III. Bohn's nodules

These are small, discrete, whitish swellings occurring in the gingivae of infants representing small cysts that arise from degeneration of remnants of the dental lamina.

Developmental Disturbances of Salivary Glands

I. Aplasia

It is absence of one or more major salivary glands.

Clinical significance

Xerostomia (dry mouth) develops.

II. Atresia

There is congenital absence or occlusion of one or more ducts of the major salivary glands. It results from failure of canalization of the proximal part of the epithelial salivary gland after the distal part has differentiated into salivary gland tissue.

III. Aberrancy

This is an **ECTOPIC** condition in which normal salivary gland tissue develops at an anatomically abnormal position. Example: **latent bone cyst (developmental lingual mandibular salivary gland inclusion cyst or depression; Steffen's bone cyst; static bone cyst)** in which part of the submandibular, or more rarely sublingual, salivary gland develop in a bony cavity or depression on the lingual surface of the mandible and maintaining its connection with its parent gland. The condition is discovered on rotein radiographic examination, and is asymptomatic.

Radiographic appearance

Rounded or oval radiolucency present in the molar area below inferior alveolar canal.

General Developmental Disturbances

I. Mongolism:

This is a developmental disturbance due to trisomy 21 (presence of three copies of chromosome 21 instead of two). It occurs with increasing frequency with advancing maternal age at the time of conception. It is characterized by a mentality retarded individual together with multiple congenital malformations of the heart and joints. There is a flat facial profile with mongoloid slant of the eyes. There is midface bony hypoplasia, the frontal and sphenoidal sinuses are absent. The maxillary sinus is hypoplastic. Macroglossia and prognathism may also present.

II. Oxycephaly:

This is a rare deformity of the skull, which is due to premature closure of the fontanelles and sutures. The skull is characterized by a wedge-shaped tower-like appearance. The upper jaw is poorly formed with crowding of the teeth and a high arched palate.