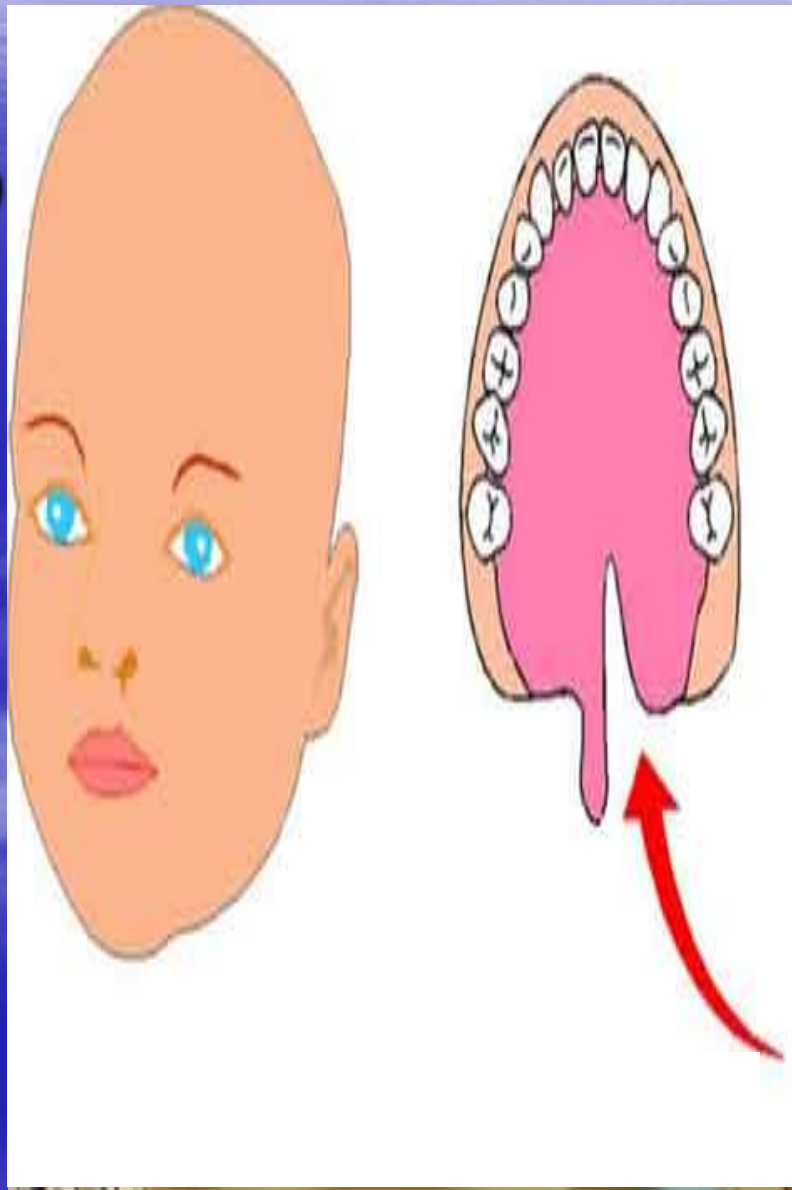
Developmental defect of

Lips and Palate
Oral mucosa
Tongue
Jaw bones



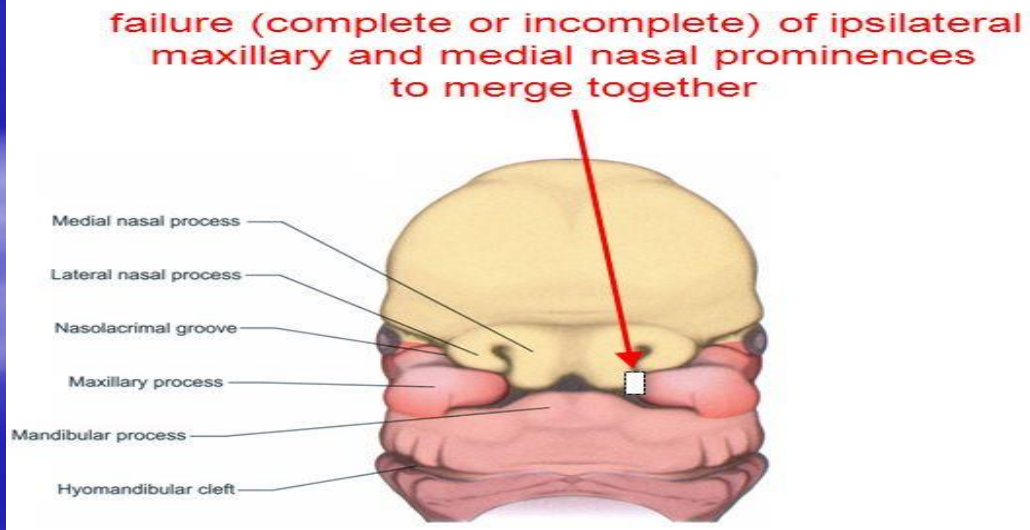
1- Orofacial clefts

A- Cleft lip & palate

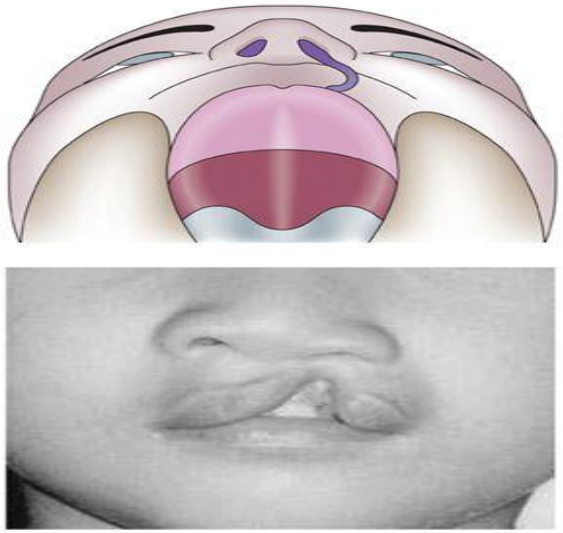
- **A- Cleft Lip:** **A developmental defect characterized by the failure of fusion of median nasal process and maxillary process , into a single structures during emberiogenesis.**

Orofacial Clefts

(Typical Cleft Deformities—Unilateral Cleft Lip [CL])



Modified from Standring (2008)

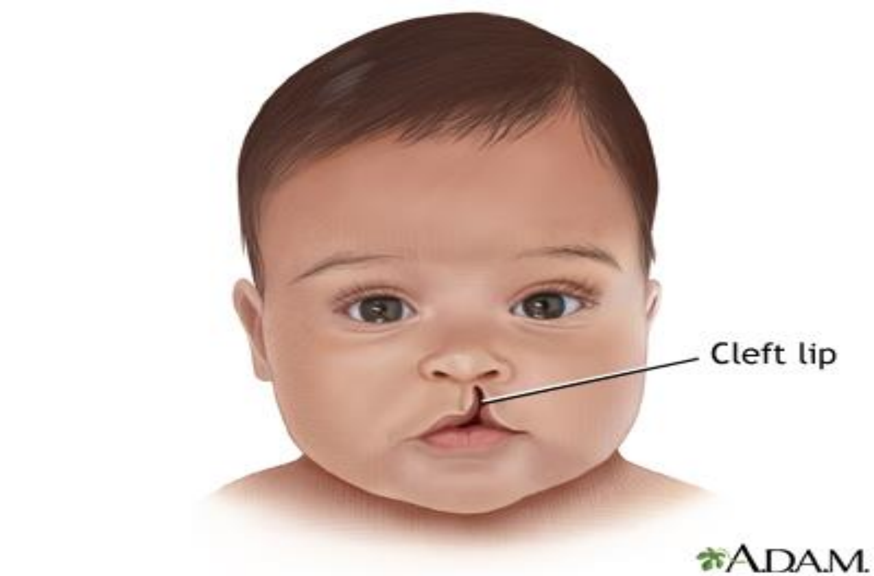


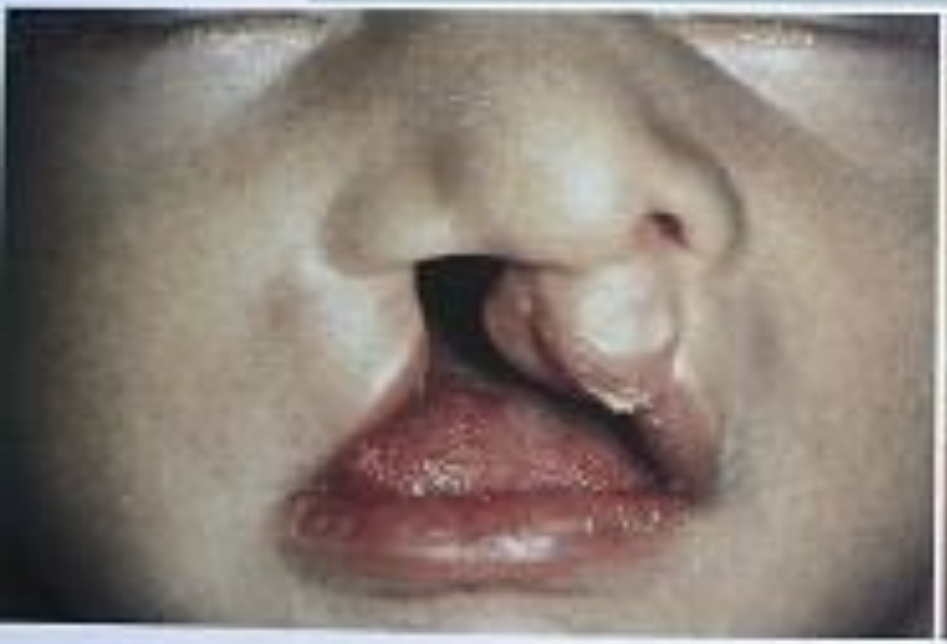
Modified from Dixon (2011)

Classification:

Unilateral, with or without anterior alveolar ridge cleft.

Bilateral, = .





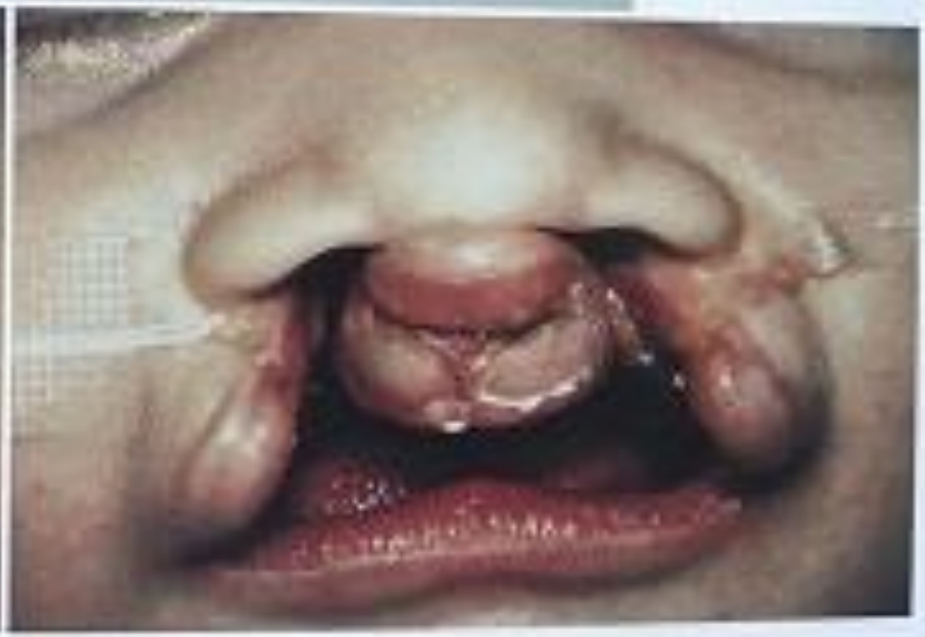


FIGURE 1-58

Cleft lip. A, The common presentations of congenital defects in lip formation. **B,** Unilateral cleft lip and palate. **C,** Bilateral cleft lip and palate. (Courtesy Dr. Ralph A. Latham.)

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- **B-Cleft Palate:** A developmental defect of the palate characterized by a lack of complete fusion of the two lateral portions of the palate, resulting in a communication with the nasal cavity.
- *Emberiogenesis defect*

- Classification:
- *Bifid uvula*
- *Soft palate only*
- *Both soft & hard palate*

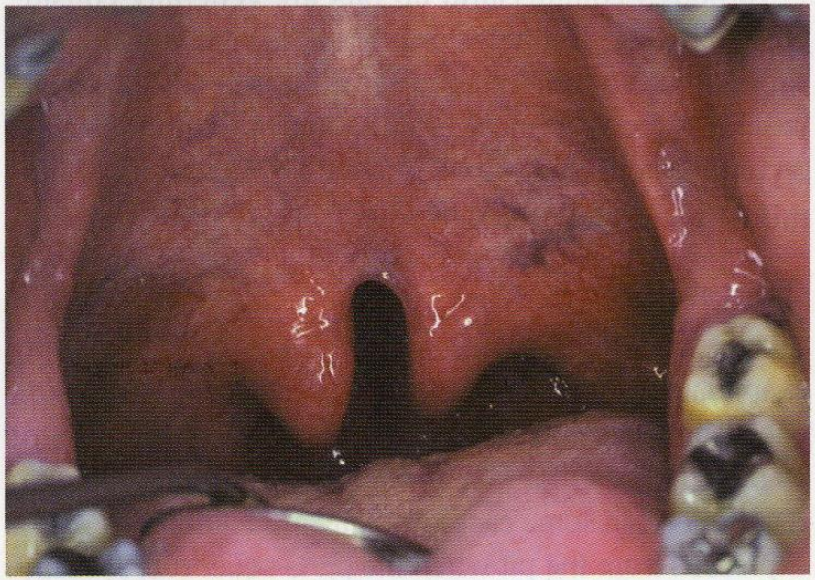
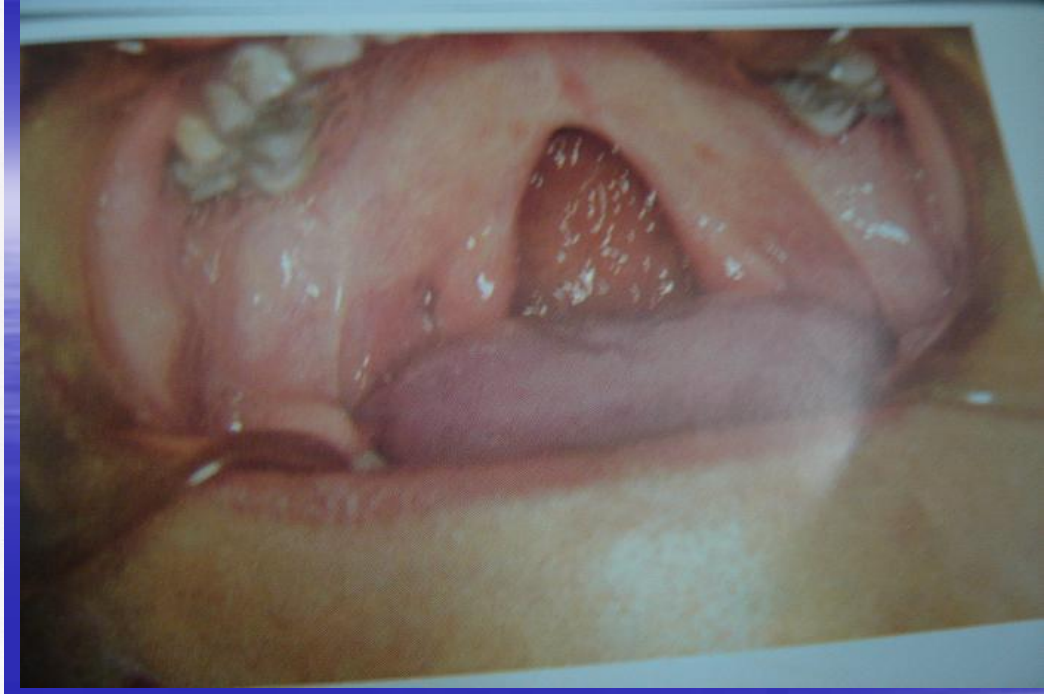
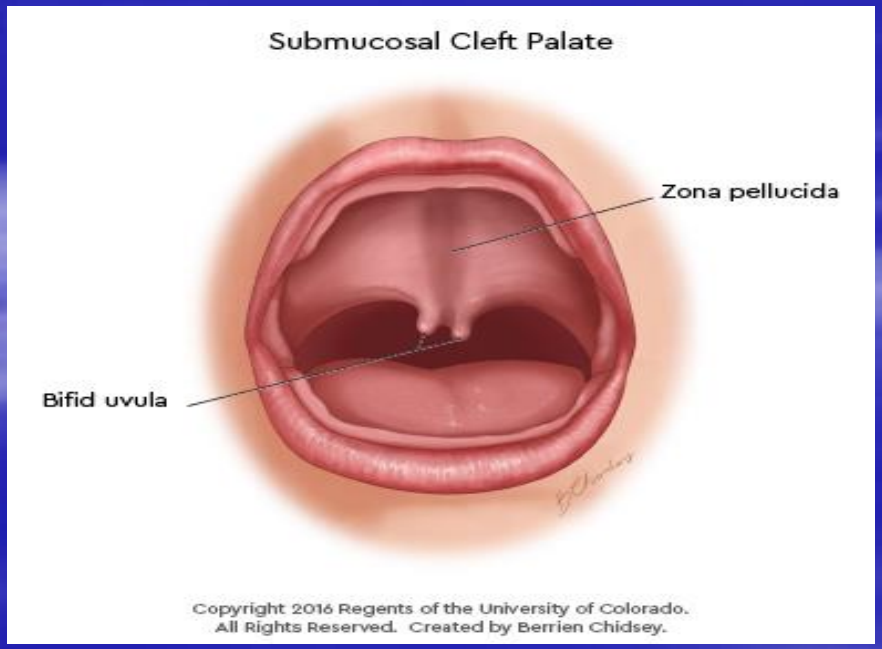


Figure 1-3 ◆ **Bifid uvula.**





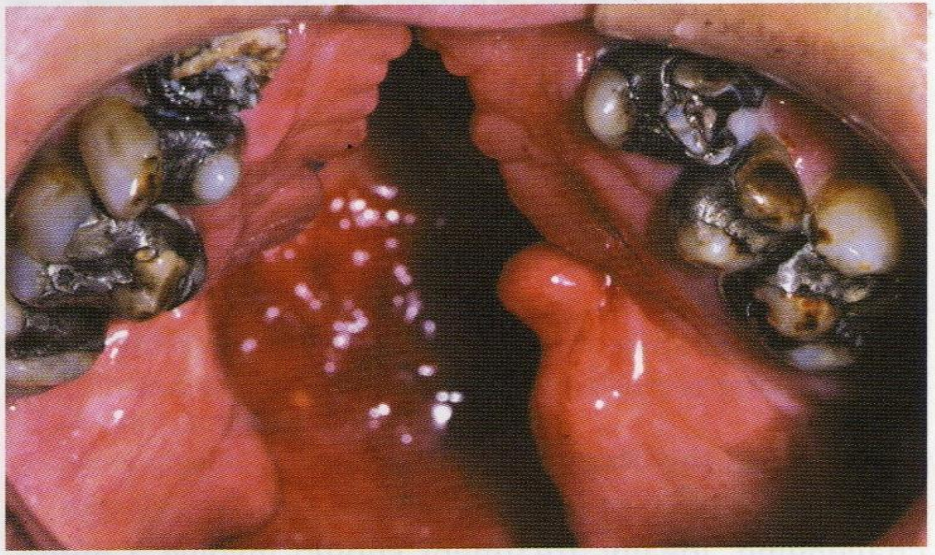


Figure 1-2 ◆ **Cleft palate.** Palatal defect resulting in communication with the nasal cavity.

C- Combined cleft lip and palate

Types:-

- Unilateral (complete or incomplete)

-Cleft palate with bilateral cleft lip(complete or incomplete)

Etiology :-

C.L & C.P involve both hereditary and environmental factors.

1-Hereditary :-

-Polygenic mean several different genes acting together.

-Every one carry genetic liability for clefts, if combined liability of parents exceed minimum threshold dose celfting occur.

2-Environmental:-

a. Nutritional factors or excess vitamin A & .a
   Riboflavin deficiency.
b. Physiological, emotional, or traumatic stress.
c. Ischemia to area.
d. Mechanical obstruction of enlarged tongue .
e. Substances e.g :- alcohol, drugs, toxins.
f. Infection.

Clinically:-

-Cleft lip with or without cleft palate occur in 1:1000 birth.

-80% of cleft palate unilateral, left side mainly.

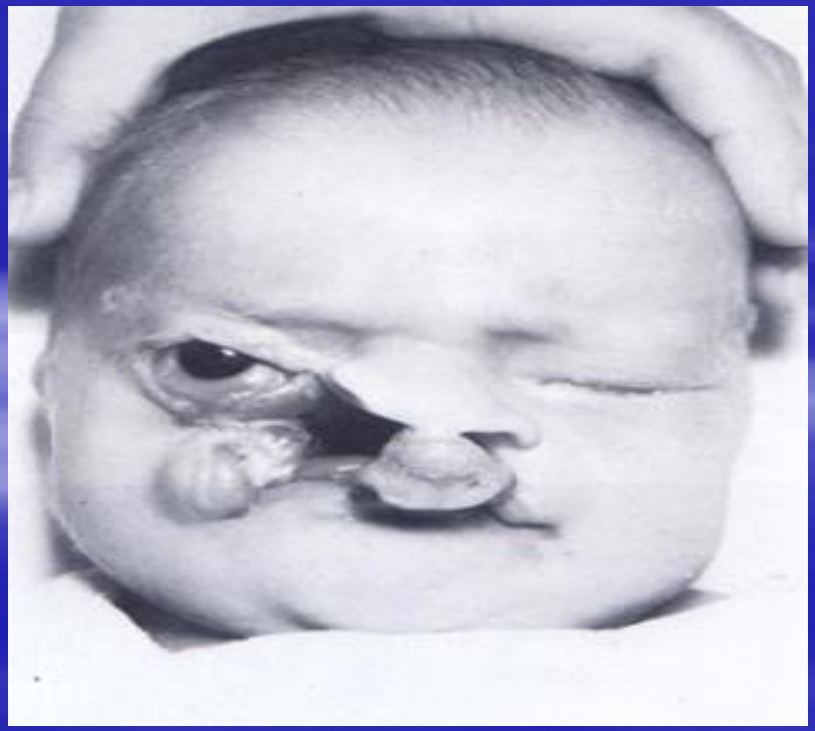
-More common in male, isolated cleft palate common in female.

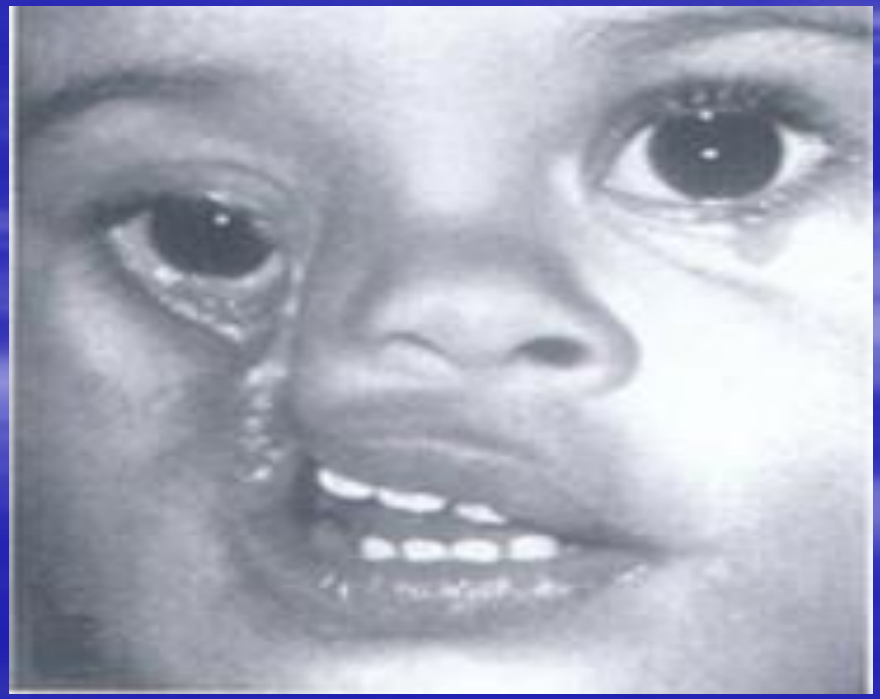
-Alveolus clefting mainly between lateral incisors & canine.

-Complete cleft lip extends to nostril, incomplete cleft lip not reach to nose. -

B- Oblique facial cleft:

- Failure of fusion of lateral nasal process with maxillary process.
- It extends from upprr lip to the eye, & always associated with cleft palate.

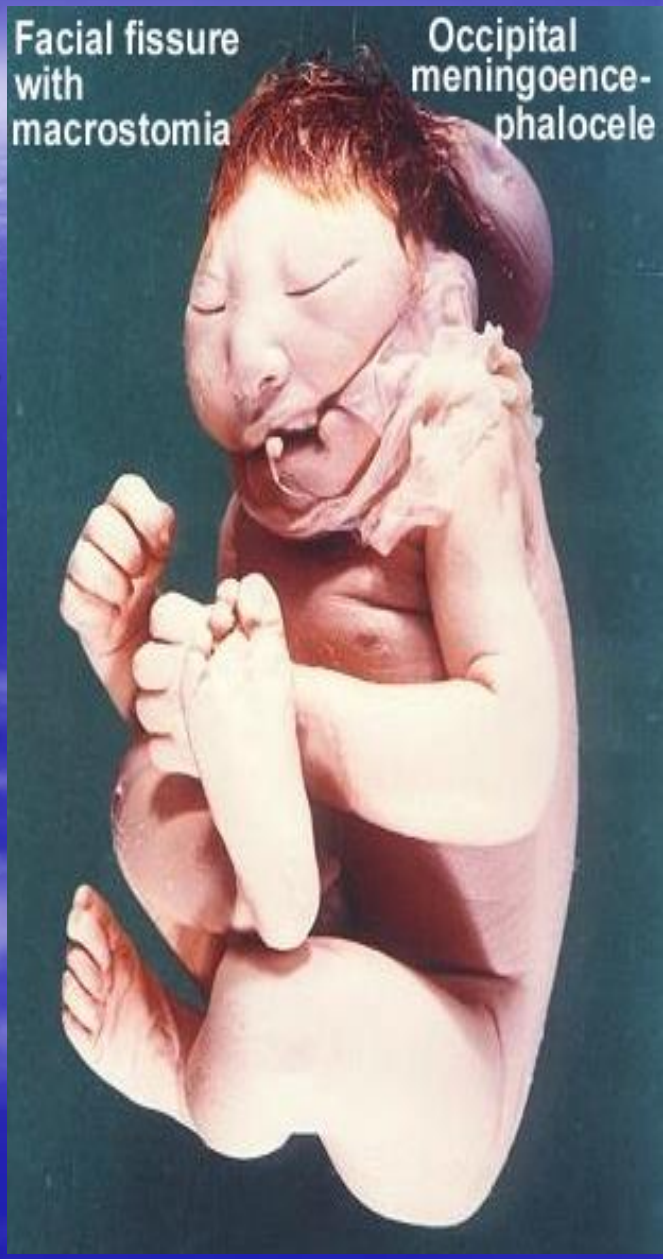




C- Lateral facial cleft:

- Failure of fusion of maxillary & mandibular process.

- Either unilateral or bilateral extending from the commissure toward the ear , resulting in **Macrostomia .**



2- Congenital lip pits:-

-Developmental defect involve :-

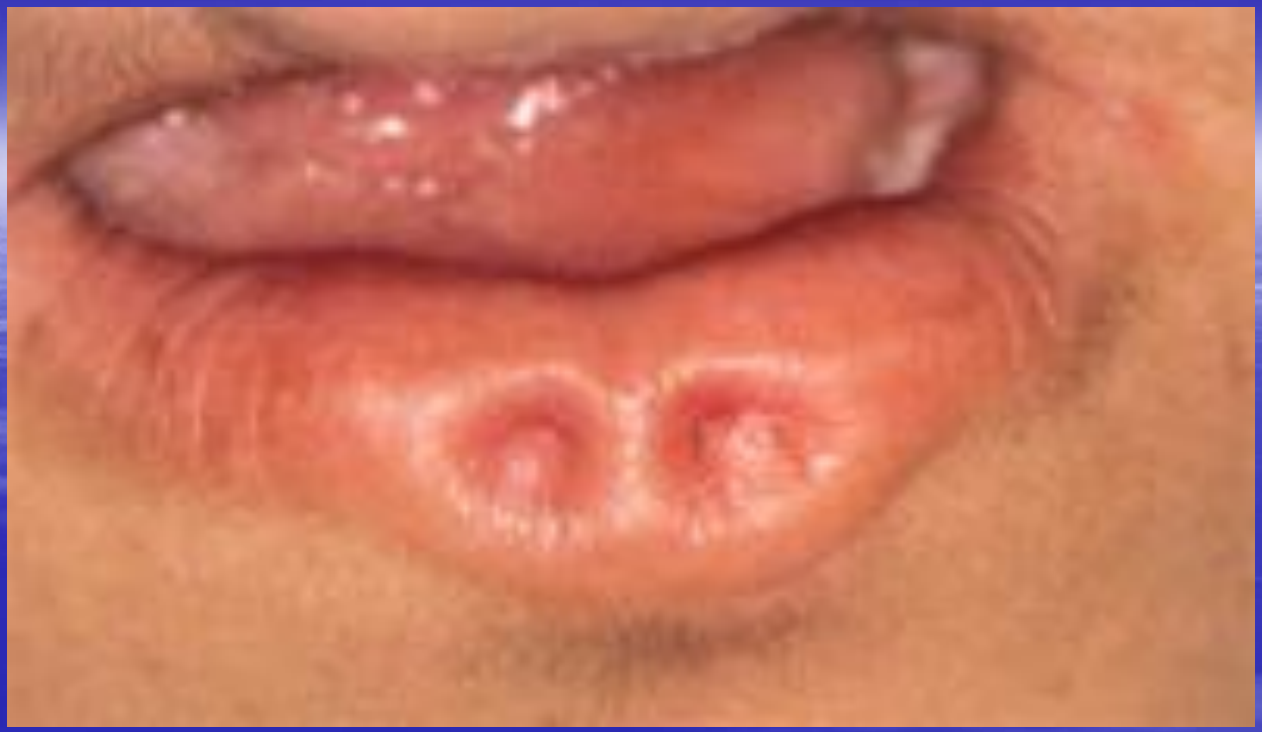
1-Paramedian portion of vermillion of lower lip and upper lip **(paramedial lip pits),** usually bilateral, lower lip.

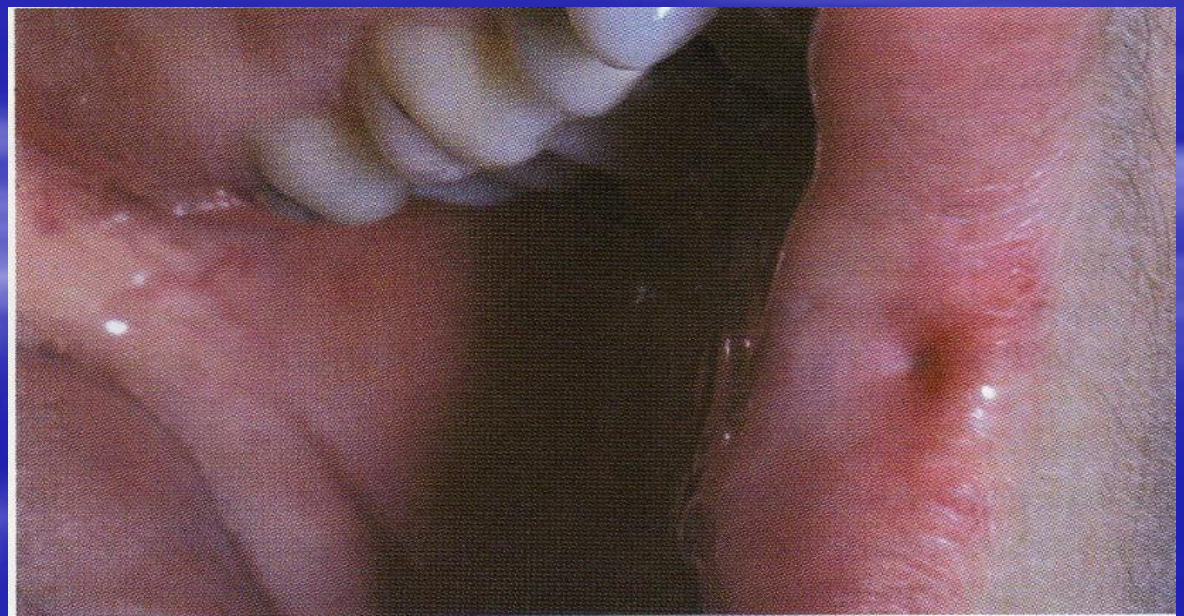
2-Labial commissure area **(commissural lip pits)** , may be unilateral or bilateral , on the corner of the mouth on the vermillion border .

It present either as:

a. congenital mucosal invaginaton → blind tract .

b. dilated ectopic salivary duct (mucous secretion)

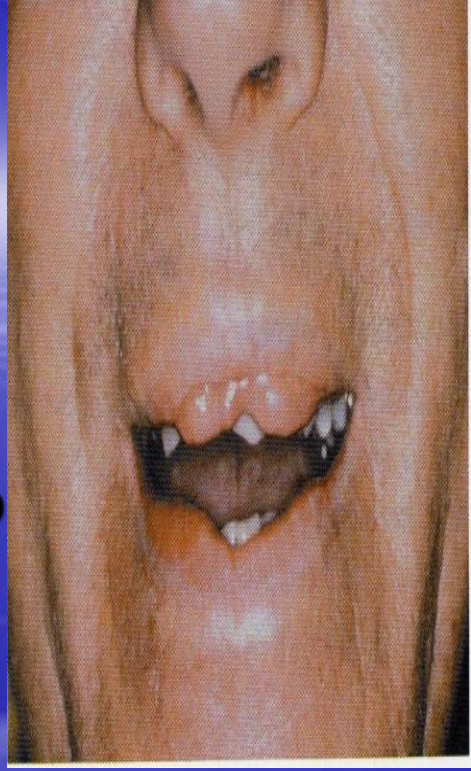




3- Douple lip

Developmental anomaly.

Horizontal fold of a redundant mucosal tissue. Usually on inner aspect of upper lip



Most often congenital in nature.

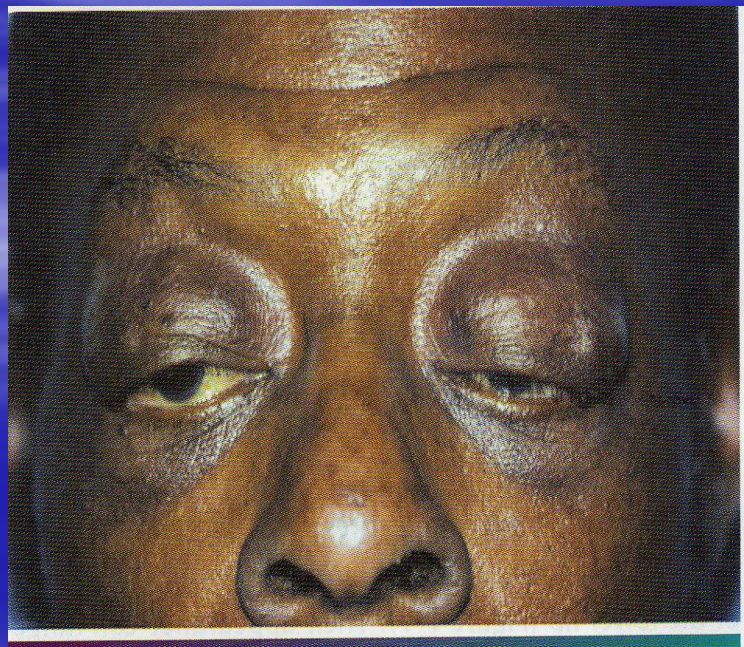
Aquired one is seen in:

Ascher syndrome

-Non-toxic goiter

-Edema of upper eye lid

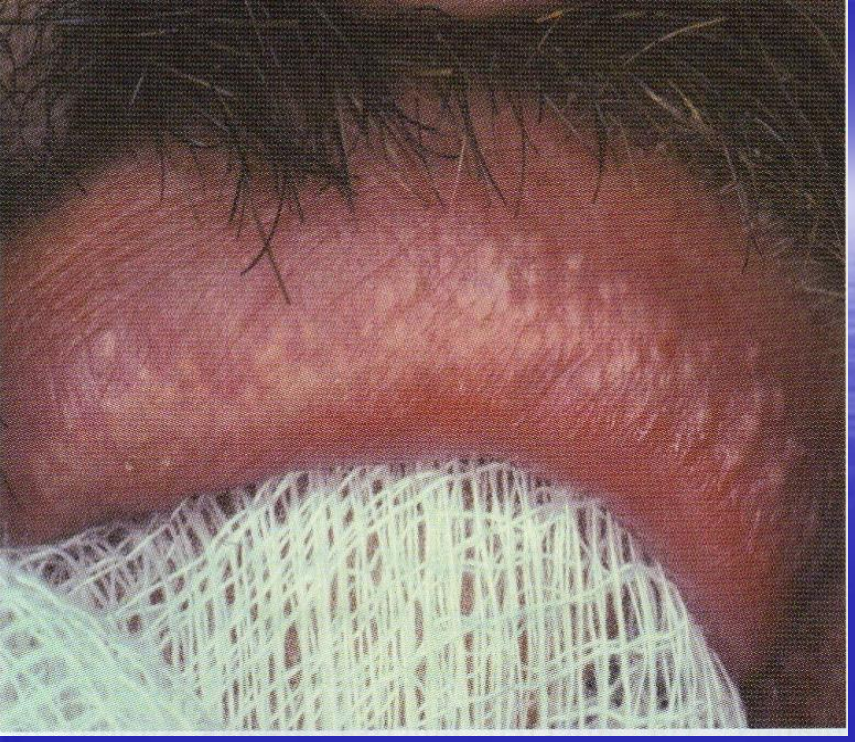
-Aquired double lip

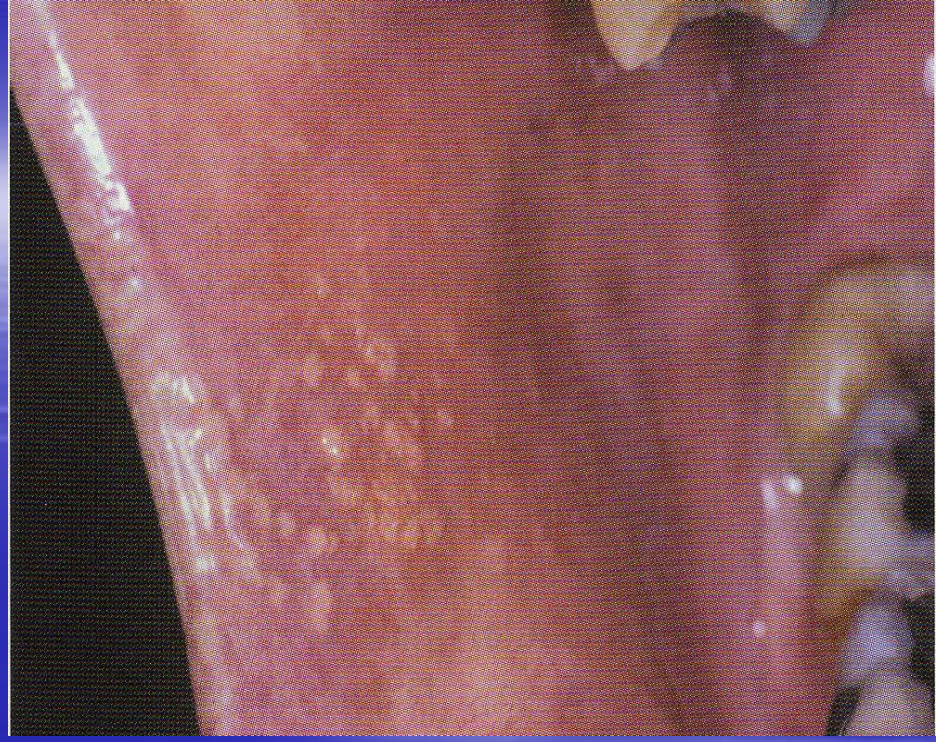


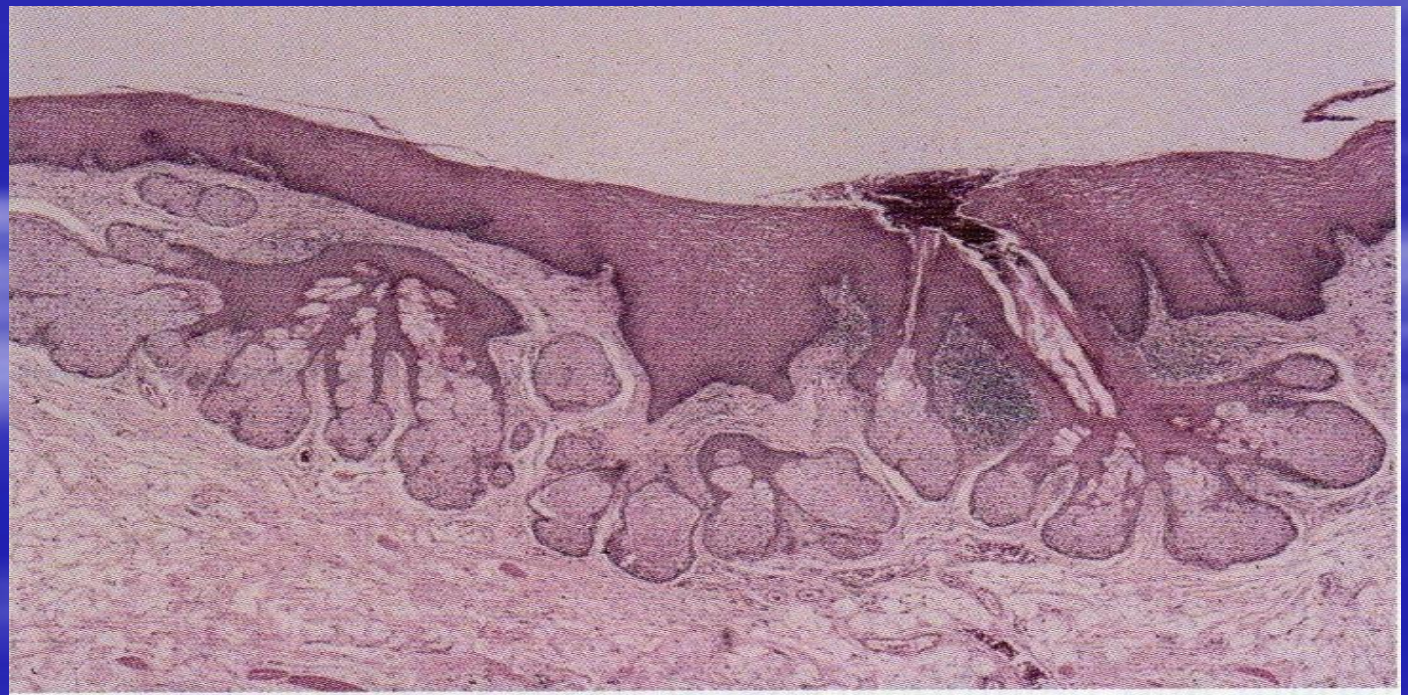
2-Developmental defect of oral mucosa:-

A. fordyce granules :-

- Ectopic presentation of sebaceous gland within oral cavity.
- Different location, mainly on buccal mucosa (bilaterally) .
- Presents as a multiple, small, yellowish spots of **1-2 mm** in diameter (milia-like) .
- It is a normal anatomic variation,seen in **>80%** of population.
- Common in adult than children → hormonal factors.







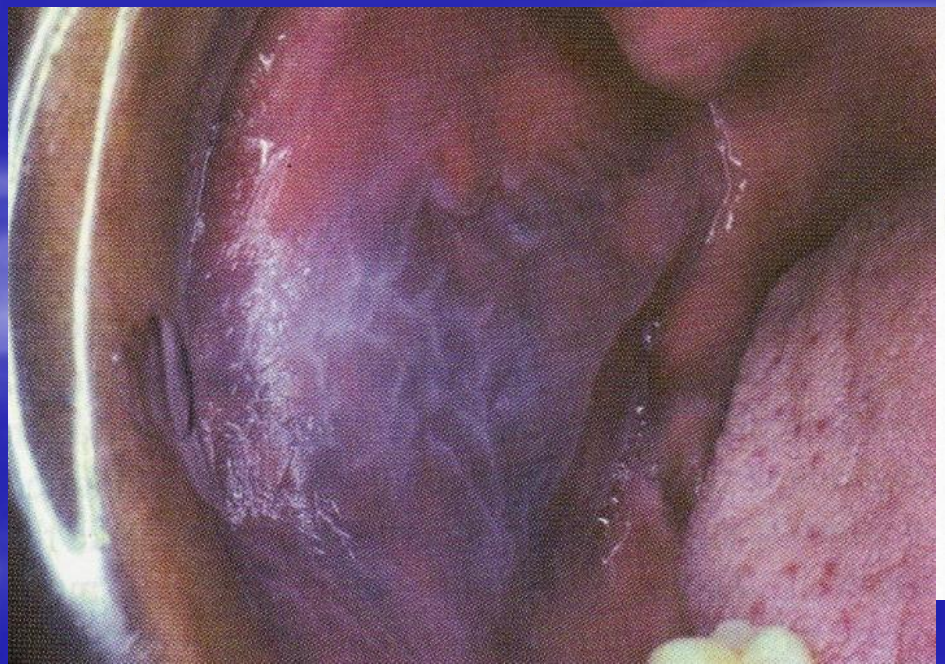
B. leukoedema:-

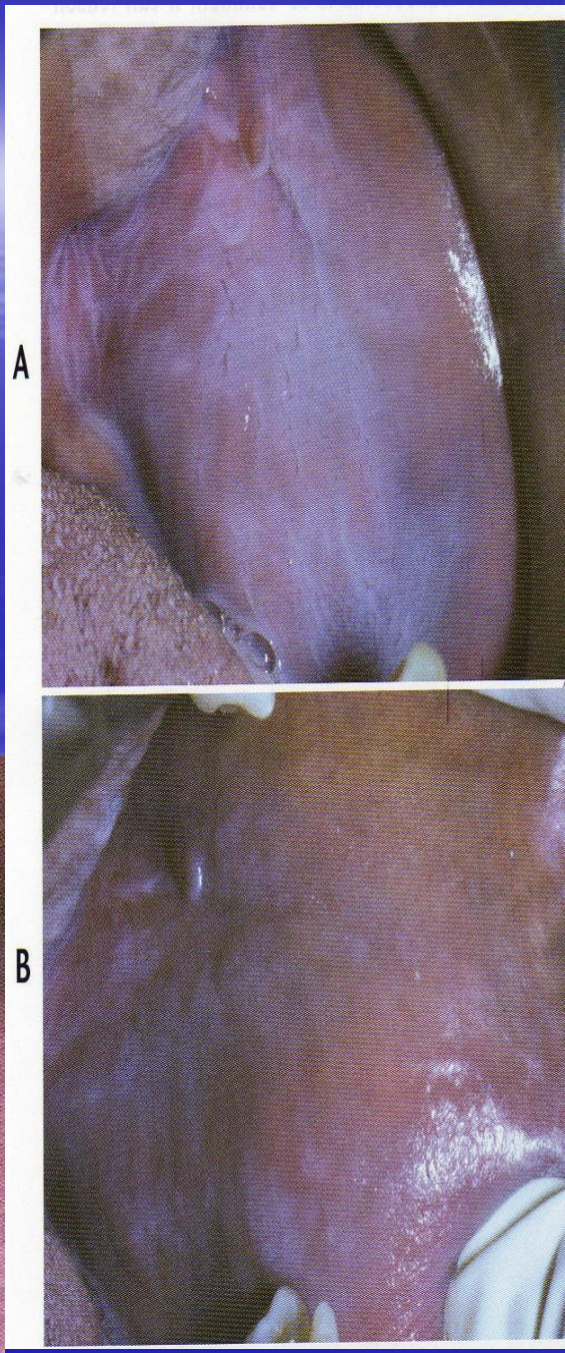
-A symptomatic, diffuse, translucent, grayish –white , filmy appearance ,on bilateral aspect of buccal mucosa.

-On stretching → reduced appearance.

-In blacks more than in white

-Unknown etiology, not a disease (normal variation).

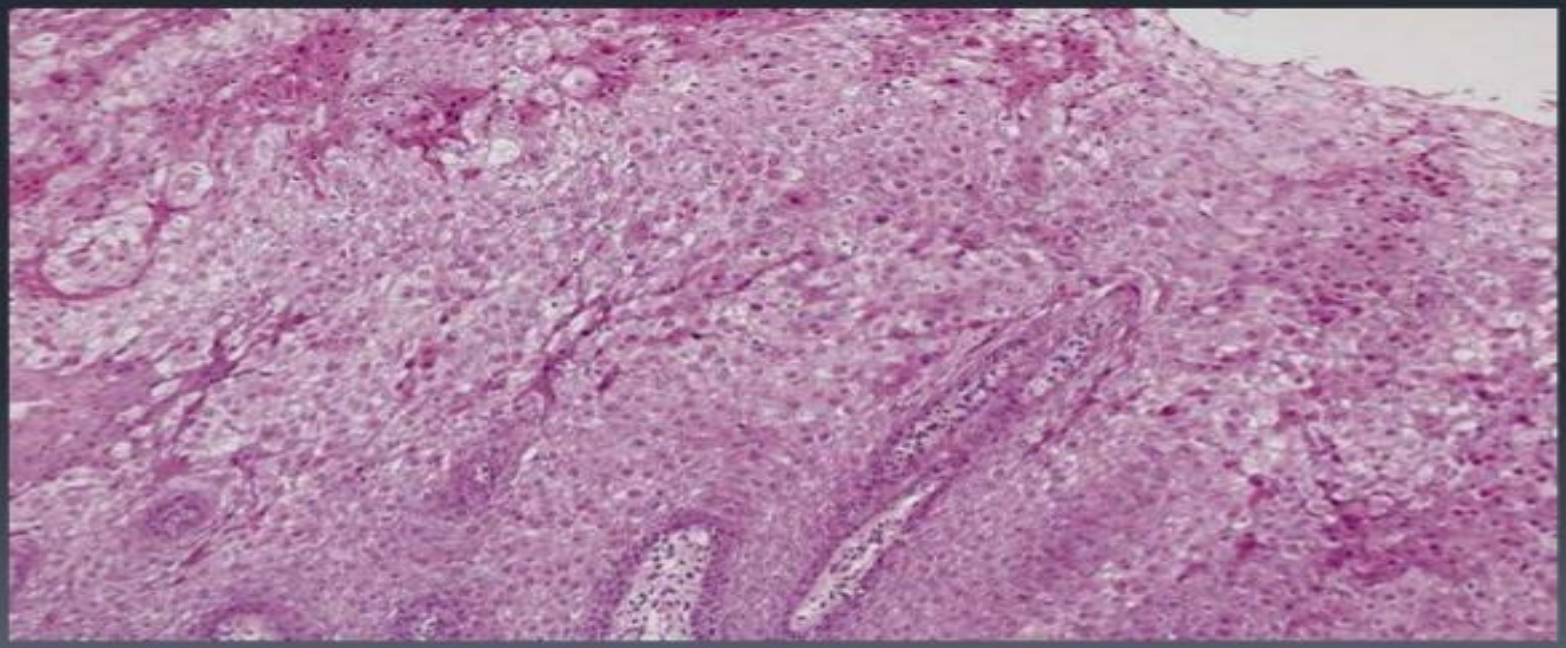




Hstologically:

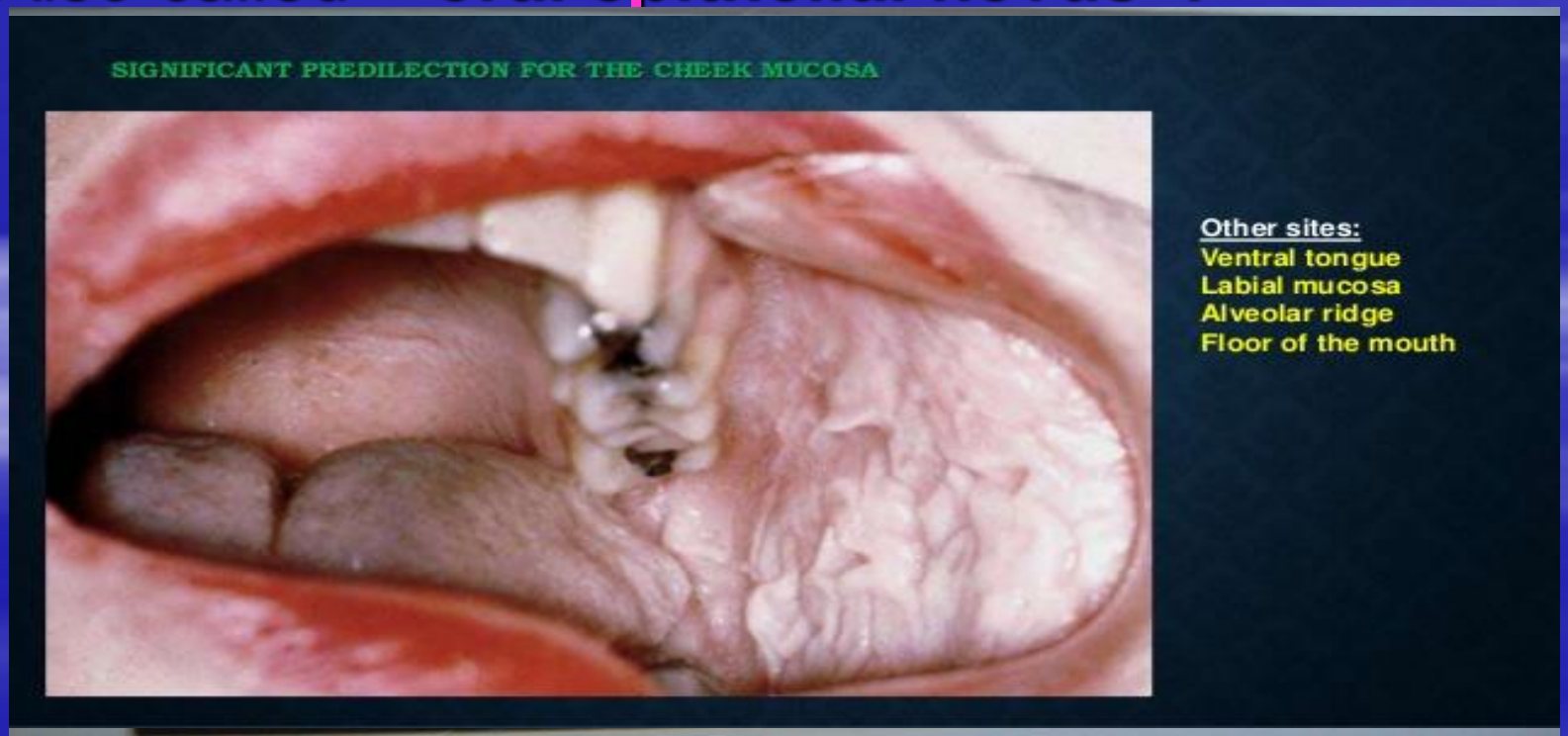
Thickening of epithelium with intracellular oedema of the spinous cell layer

Histopathology: a mild to moderate hyperparakeratosis, acanthosis , and intracellular edema of the spinous cells are shrunken (pyknotic).

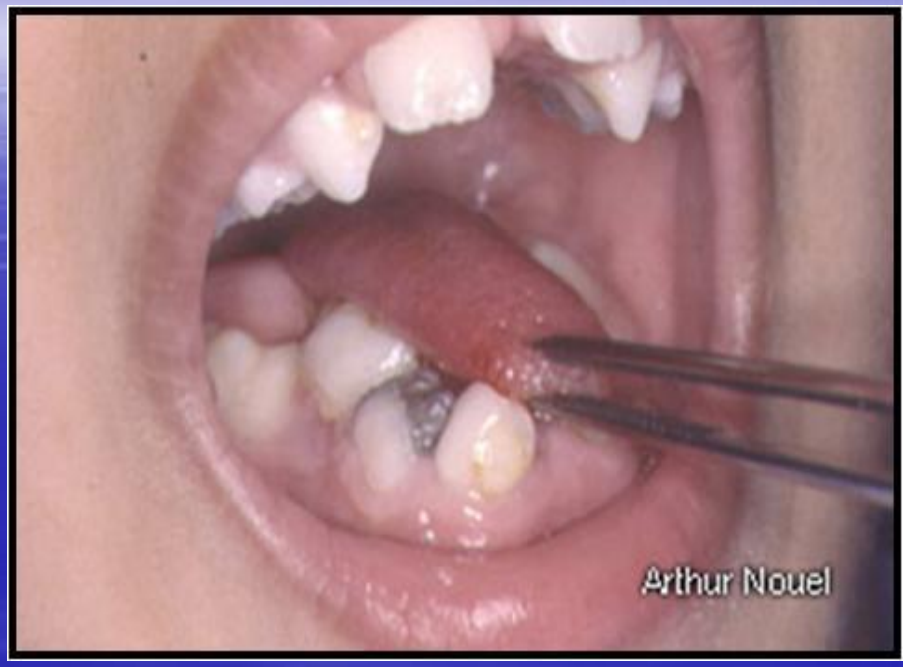


C. White sponge nevus :-

- Autosomal dominant hereditary condition.
- Asymptomatic, reassurance of patient.
-Whitish thickening and folding involving entire oral mucosa.
-Also called **“oral epithelial nevus”.**



3-Developmental defects of tongue:-



A- Microglossia

-Abnormal small tongue

-Uncommon , unknown cause

-Associated with **oromandibular –limb hypogenesis syndrome**.

Limb abnormalities → Hypodactylia

(Absence of digit)

B. Macroglossia

-Abnormal large tongue

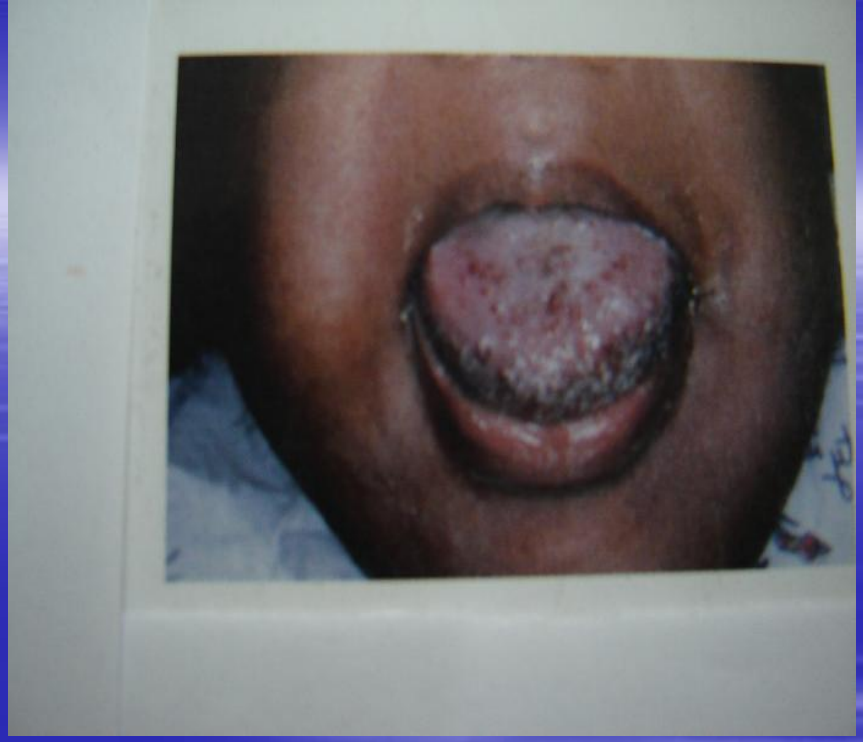
-Either

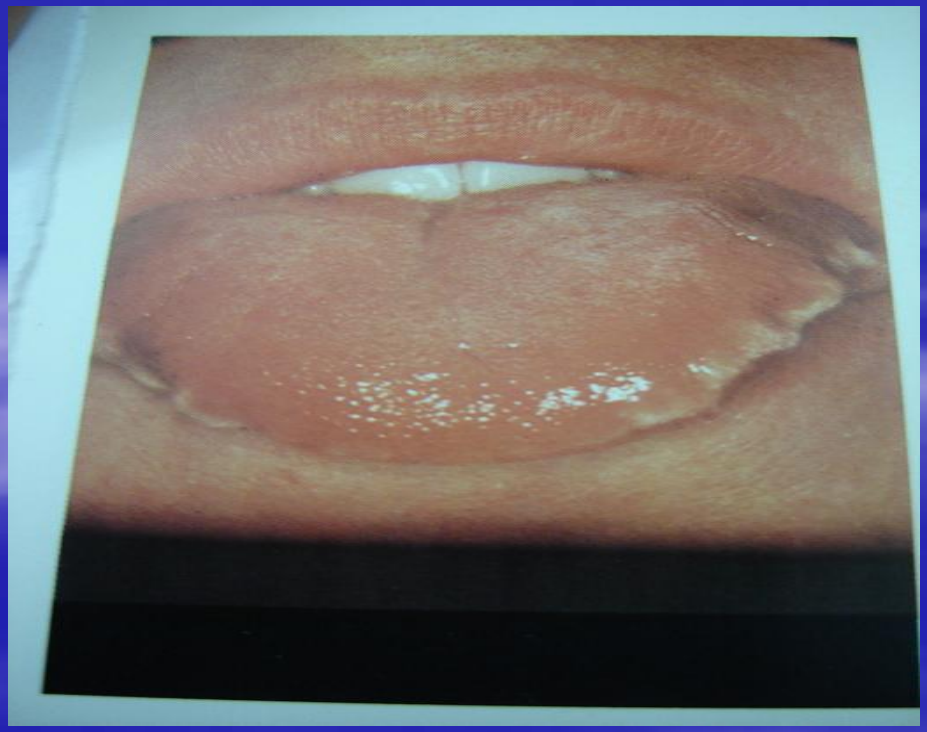
1-Congenital :-as in Down's syndrome,Hemangioma, Lymphangioma

2-Acquired :-as in Acromegaly, Amyloidosis ,Cancer

-Macroglossia manifested :-Noisy braething,Drolling of saliva,difficulty in eating and lisping speech.

Clinically :- patient with open bite and mandibular prognathism, crenated lateral border of tongue.





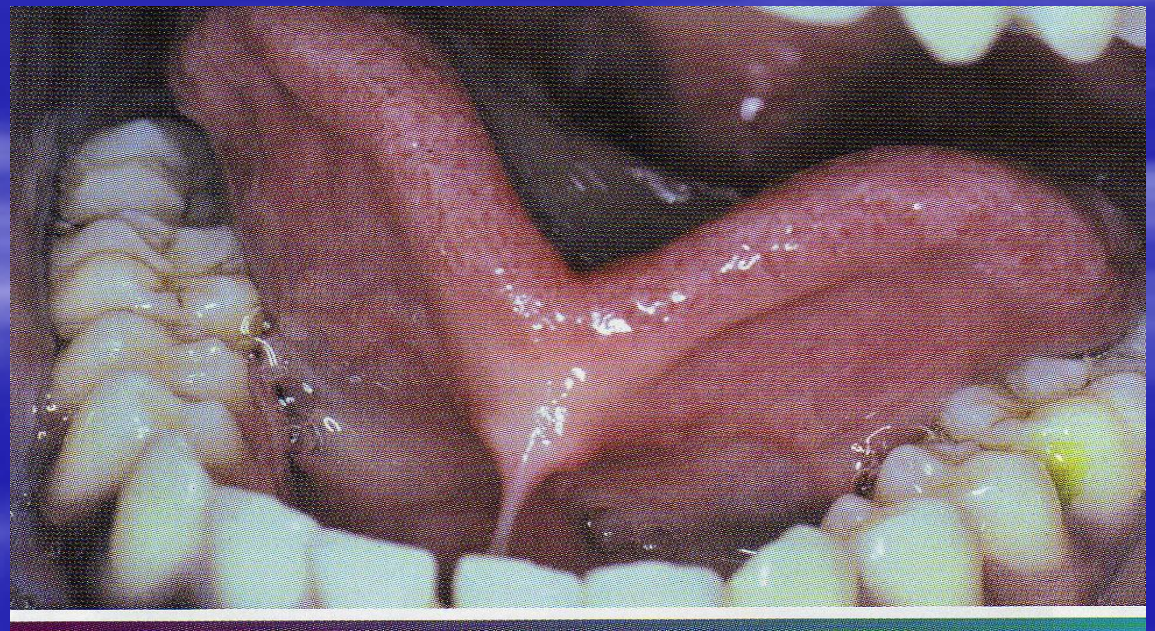
Macroglossia

C. Ankylglosia (Tongue tie)

-Developmental anomalies

-Short ,thick lingual frenum lead to limitation of tongue movement and impaired speech.

-High mucogingival frenum attachment lead to gingival and periodontal disease (locally).



D. lingual thyroid nodule

-Accessory accumulation of thyroid tissue (functional) within the body of posterior tongue.

-Thyroid remnant in region of thyroid gland origin.

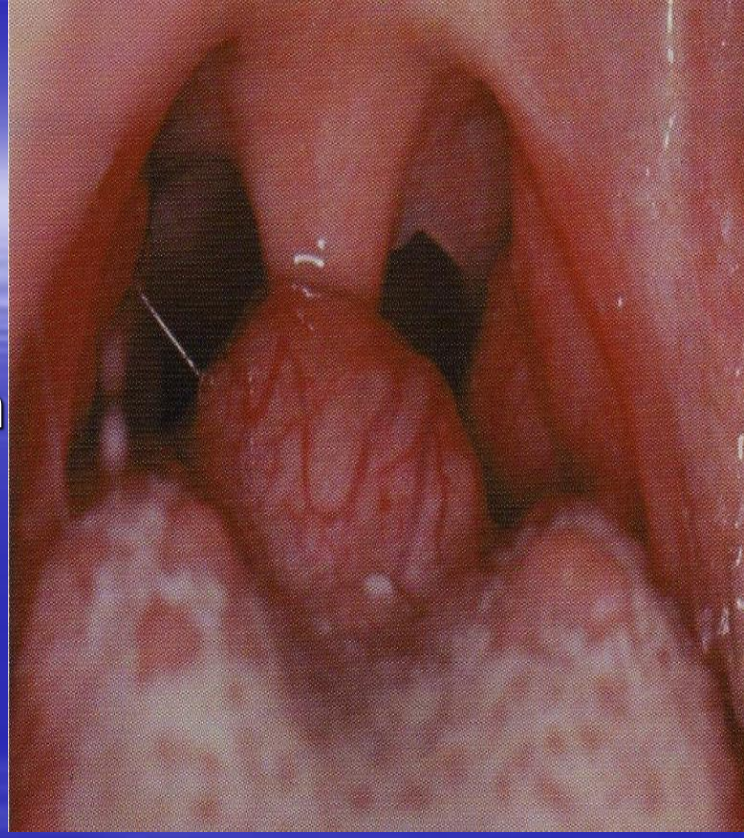
-Rare, common in female during puberty & adolescence.

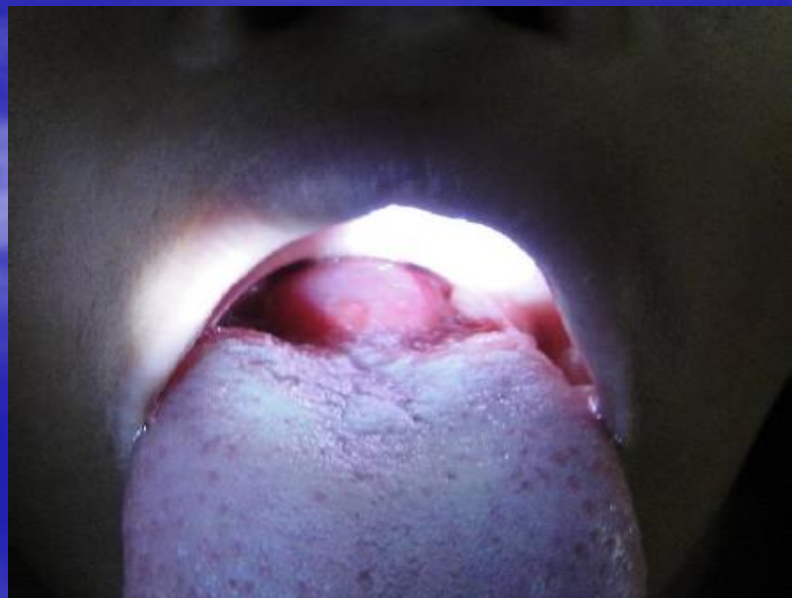
- 2-3 cm smooth, sessile mass on mid- posterior dorsum of tongue, in the foramen caecum region.

-Symptoms:-

Dysphagia,

Dysphonia, Dyspnea, Hypothyroidism



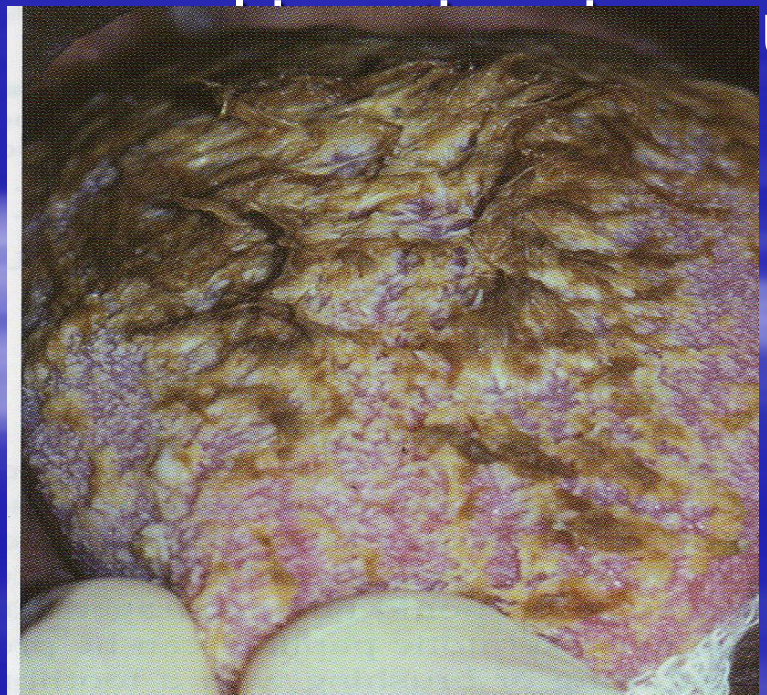


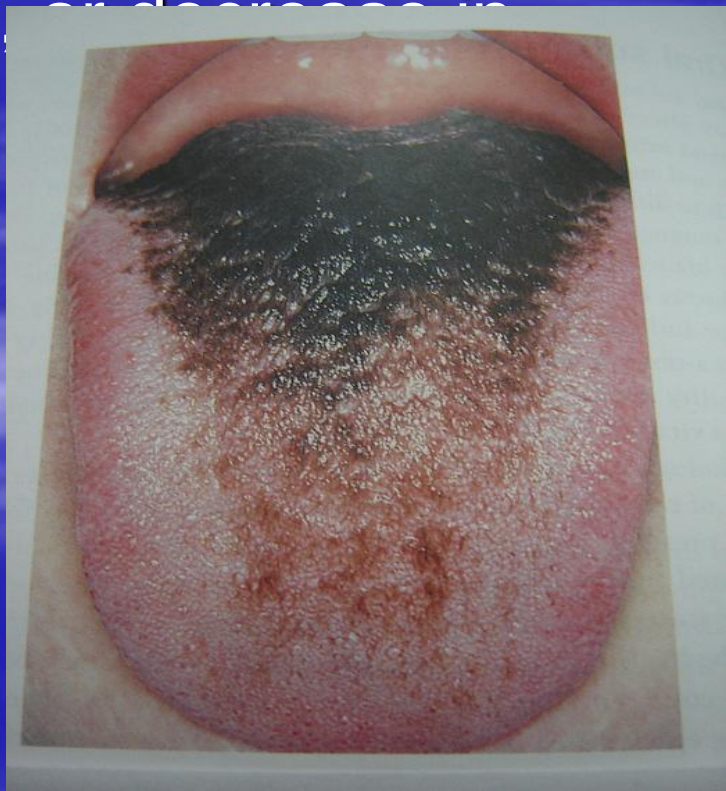
E. Black hairy tongue

-Marked keratine accumulation on the filliform papillae on dorsal tongue → Hair- like appenrance.

-Patient complain of gag reflex and bad taste.

-An increase in keratine production, or decrease in uamation.





Associated factors:-

1.Antibiotic therapy.

2. Poor oral hygiene.

3. Oxidizing mouth wash.

4. Bacteria and fungi over growth.

Clinically:-

- Elongated filliform papillae with brown, yellow or black pigment, due to over growth of pigment-producing bacteria or fungi .

Patient complain of gagging or bad taste. -

Treatment:- Periodic scraping of papillae.

F. Fissured tongue :-

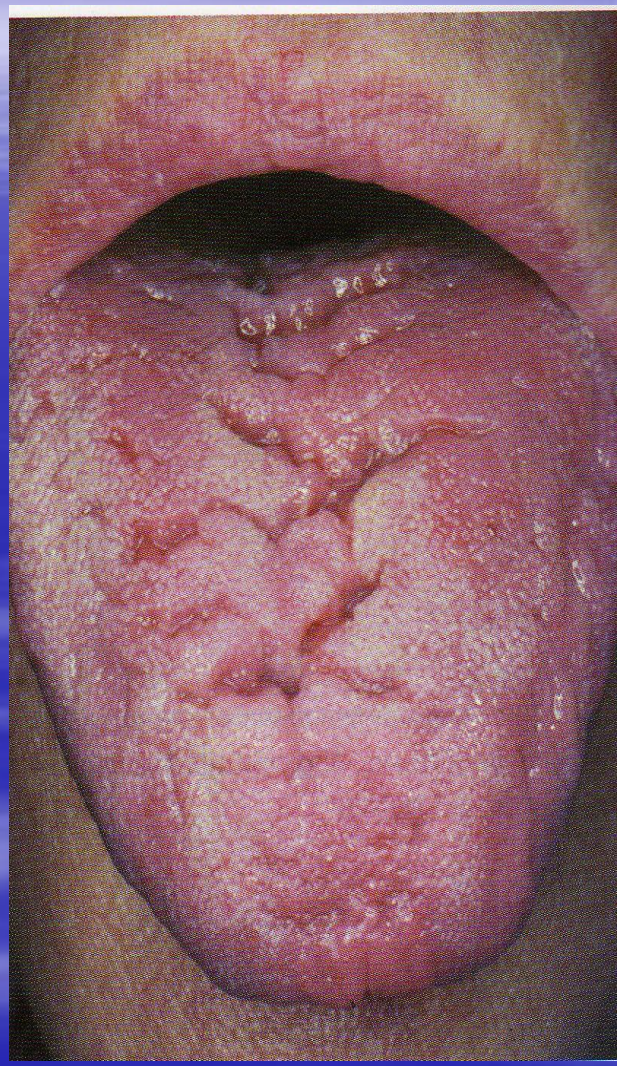
-Numerous grooves or fissure presents on dorsal tongue surface.

-Uncertain cause, relatively common.

-An association between fissure tongue and geographical tongue

.

- **Treatment**:- improvement of oral hygiene , antibiotic therapy to treat infection result from accumulation of bacteria and food debris in fissures.

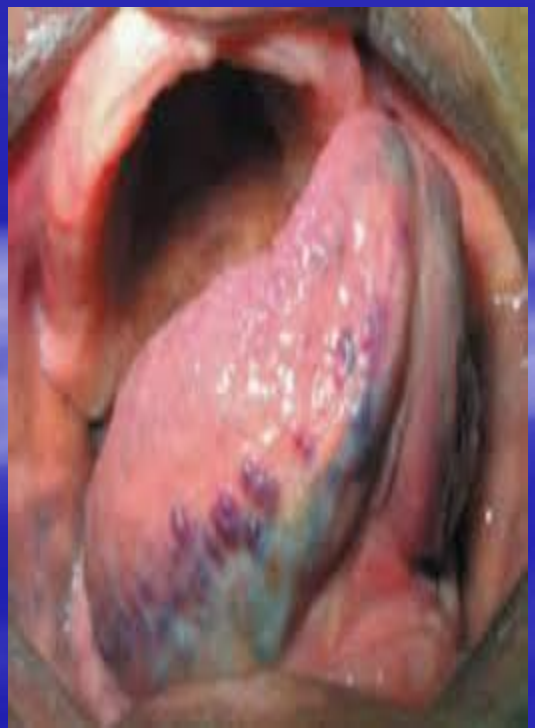


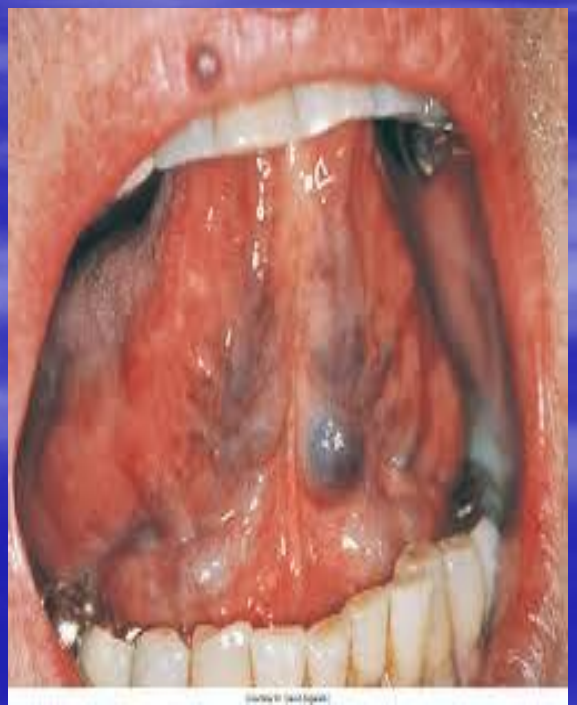
G. Lingual varicosities (varices)

-Dilated tortuous veins on ventral tongue surface.

-Increase with age (prominent).

-Asymptomatic, unless secondary thrombus is formed.





H. Geographic tongue (= Benign migratory glossitis, Erythema migran)

-Large, red, atrophic patches in tongue with white, slightly raised border.

-Patches resolves in days-weeks & the papillae will regenerate.

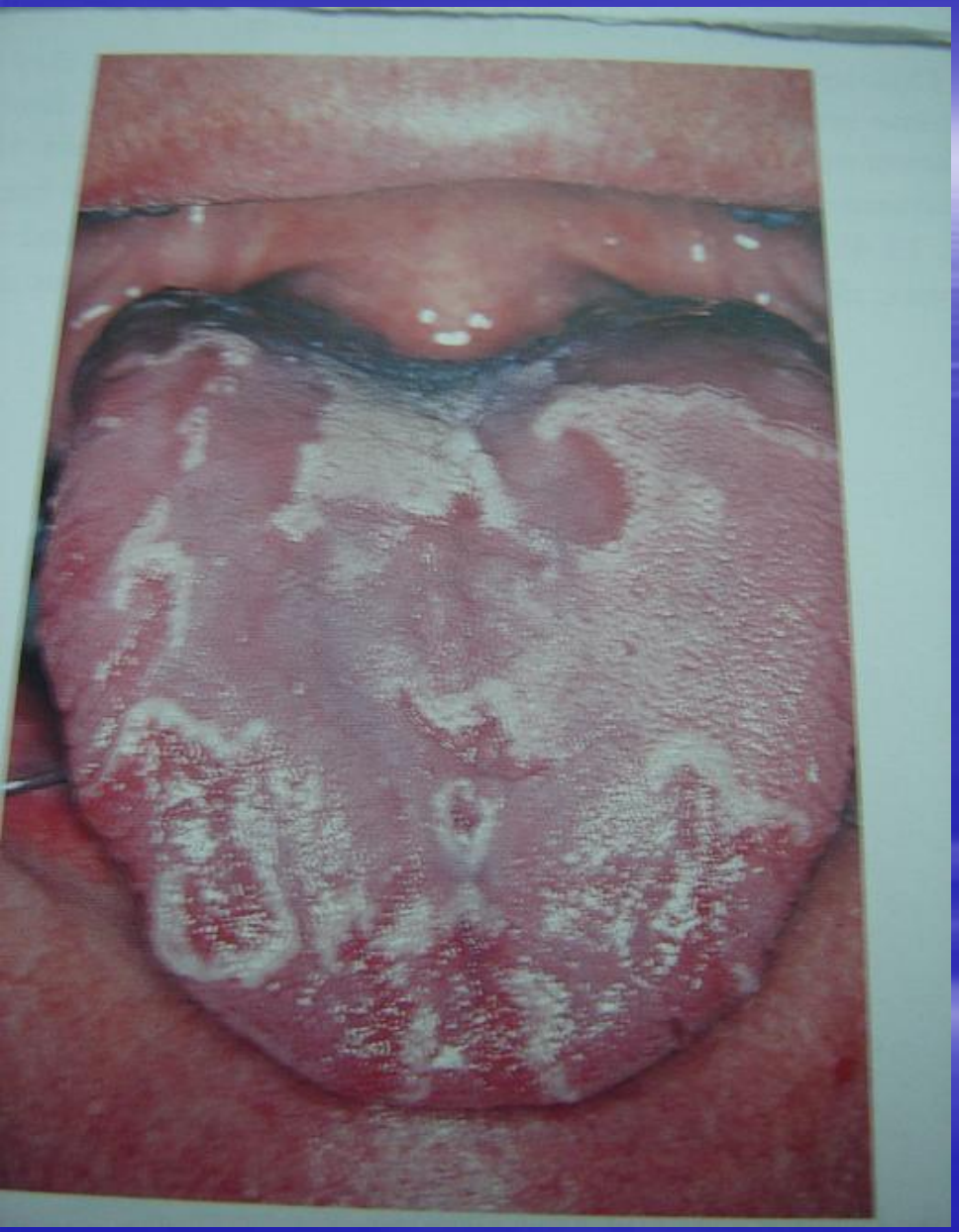
-Multiple lesions on the dorsal tongue.

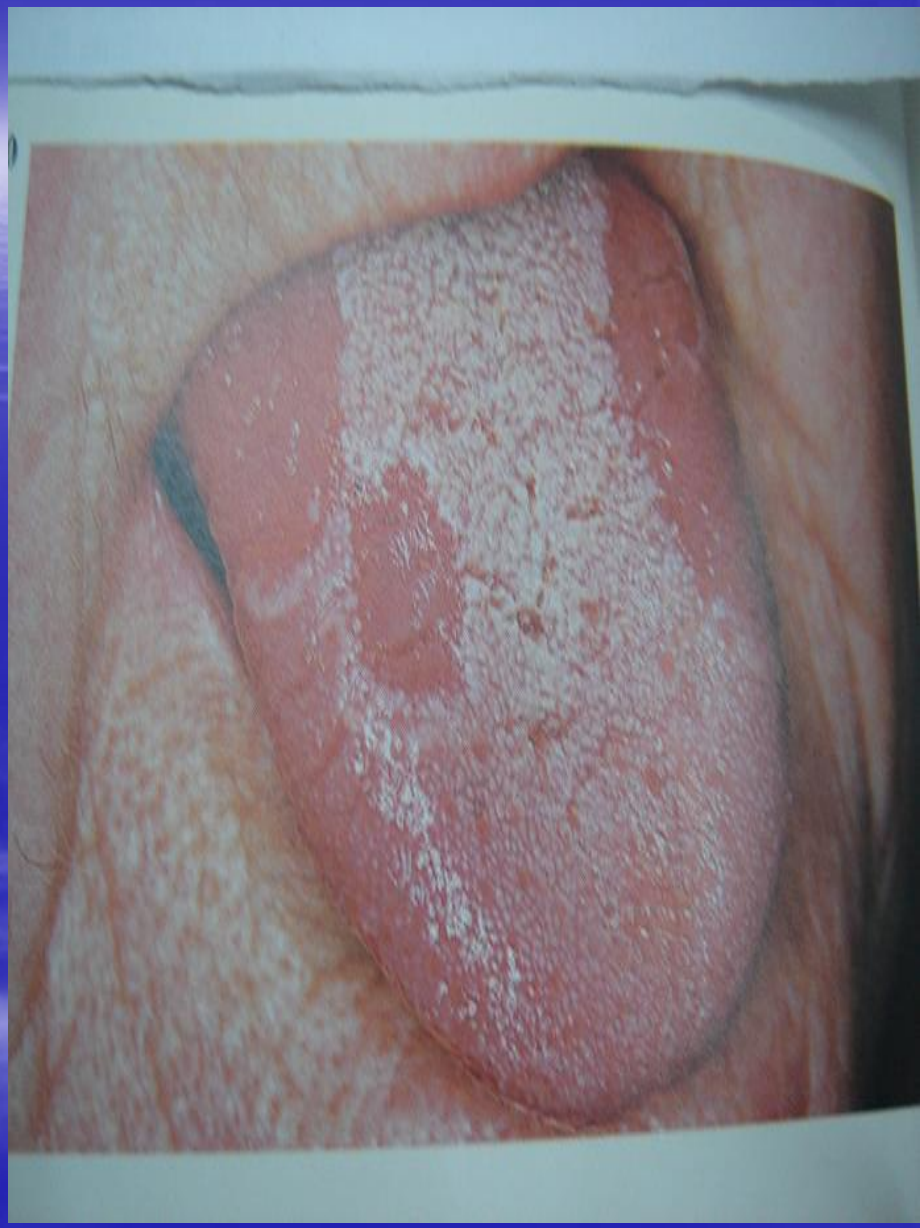
-Red area → no filliform papillae, White area → hyperatrophy of papillae .

-It is a recuurent lesion, so it appear as migrate from area to area.

-Important ,,because it may confused with a premalignant and malignant lesion.

(complete benign lesion).





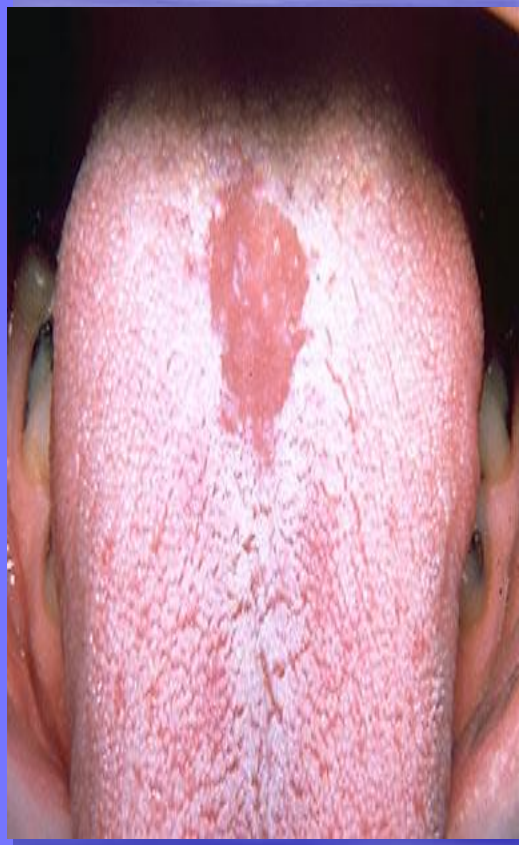
Geographic tongue

i-Median rhomboid glossitis

(Posterior lingual papillary atrophy)

(Glossal central papillary atrophy)

- The embryonic tongue is formed by two lateral processes (lingual tubercles) meeting in the midline and fusing above a central structure from the first and second branchial arches, (tuberculum impar)
- The posterior dorsal point of fusion is occasionally defective, leaving a rhomboid-shaped, smooth erythematous mucosa lacking in papillae or taste buds.
- This median rhomboid glossitis is a focal area of susceptibility to recurring of chronic atrophic candidiasis, prompting a recent movement toward the use of posterior midline atrophic candidiasis as a more appropriate diagnostic term.



4-Develapmental defects of jaw bones

A. Micrognathia "very small jaw"

Either mandible or maxilla , or both jaws.

It result in :-1.Posterior tongue displacement

2.Airway obstruction

3.Dental problems lead to

-difficult mouth opening

-difficult brushing

- periodontal disease

It may be associated with other developmental defect like in **‘‘Pierre Robin’s sequence’’**

Cleft palate ,Micrognathia ,Glossoptosis

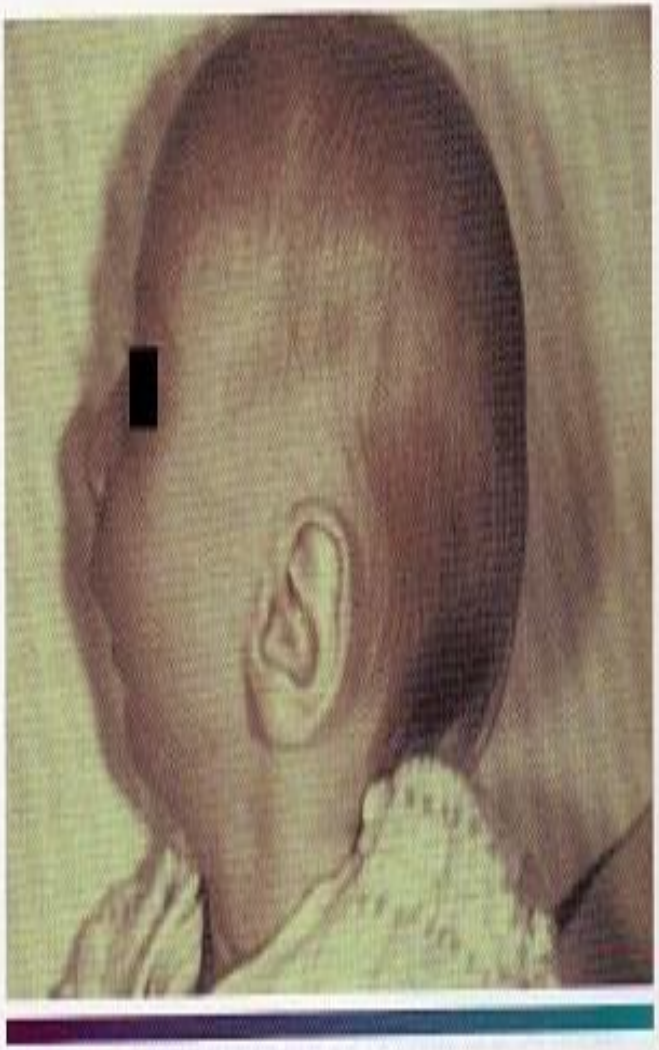
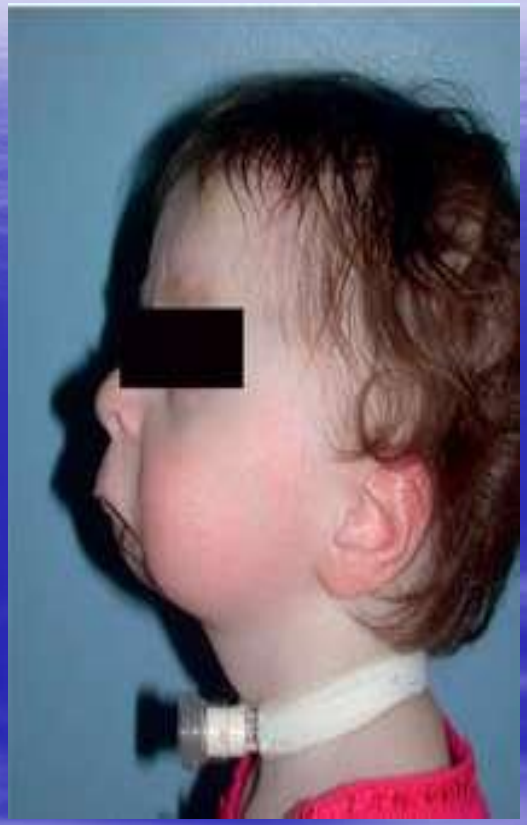
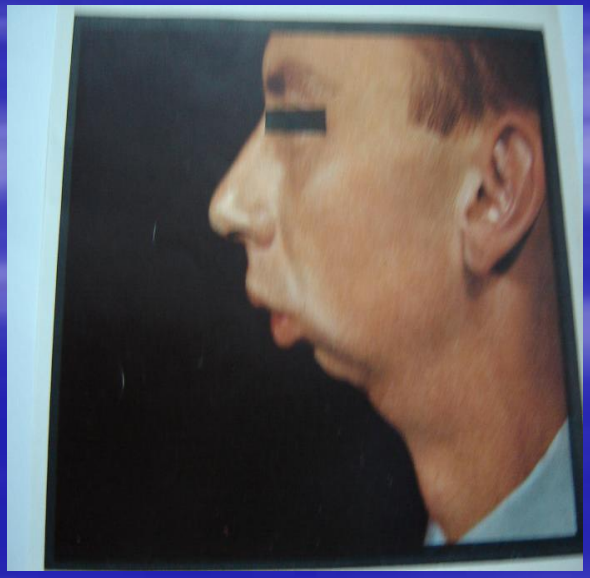


Figure 1-5 • **Pierre Robin sequence.** Micrognathic mandible in an infant with cleft palate. (Courtesy of Dr. Robert Gorlin.)





B. Macrognathia "large jaw"

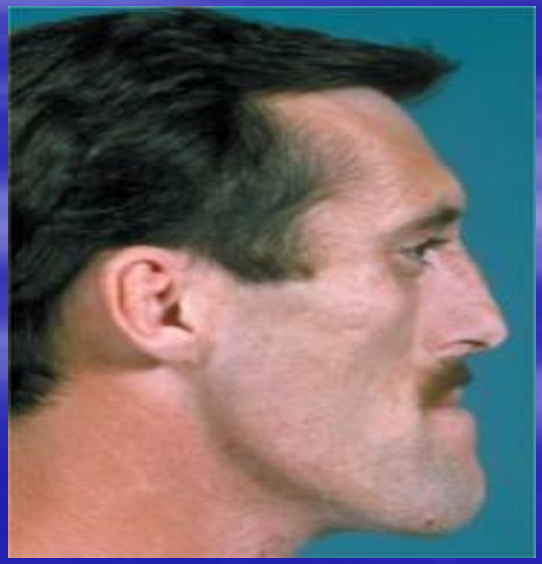
Local causes :- 1- Fibrous dysplasia of bone.

2- Reactive bone tumor.

3- odontogenic cyst and tumor.

systemic cause:- 1-Acromegaly

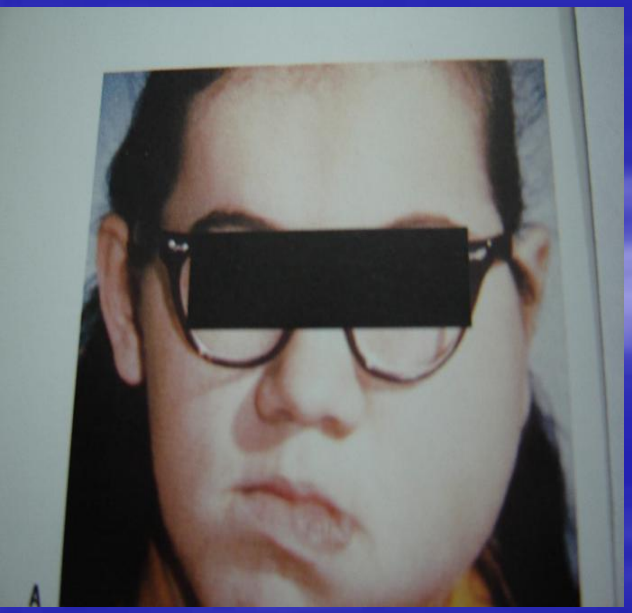
2-Paget's disease.



C. Hemifacial hypertrophy :-

-Significant unilateral enlargement of face as a result of increase neurovascular supply of the affected side.

-Asymmetry of face: Unilateral enlargement of facial tissue, bones, & teeth → malocclusion & deviation of affected side to un affected one.



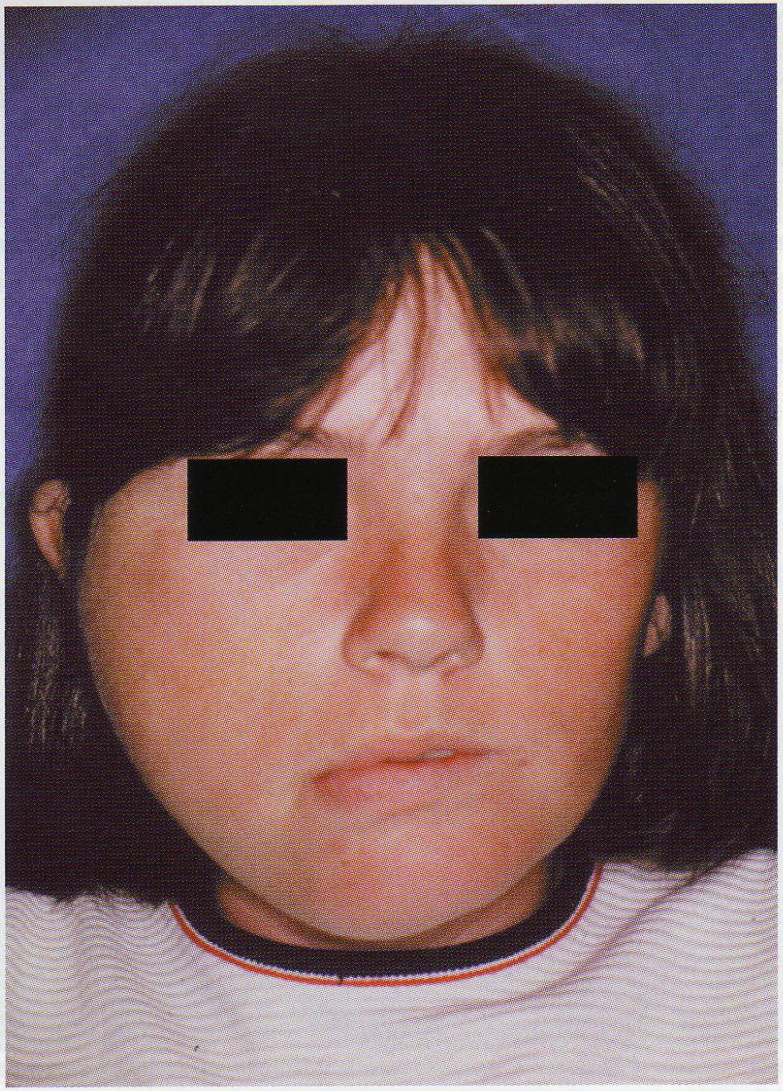


Figure 1-77 • Hemihyperplasia. Enlargement of the right side of the face. (Courtesy of Dr. George Blozis.)

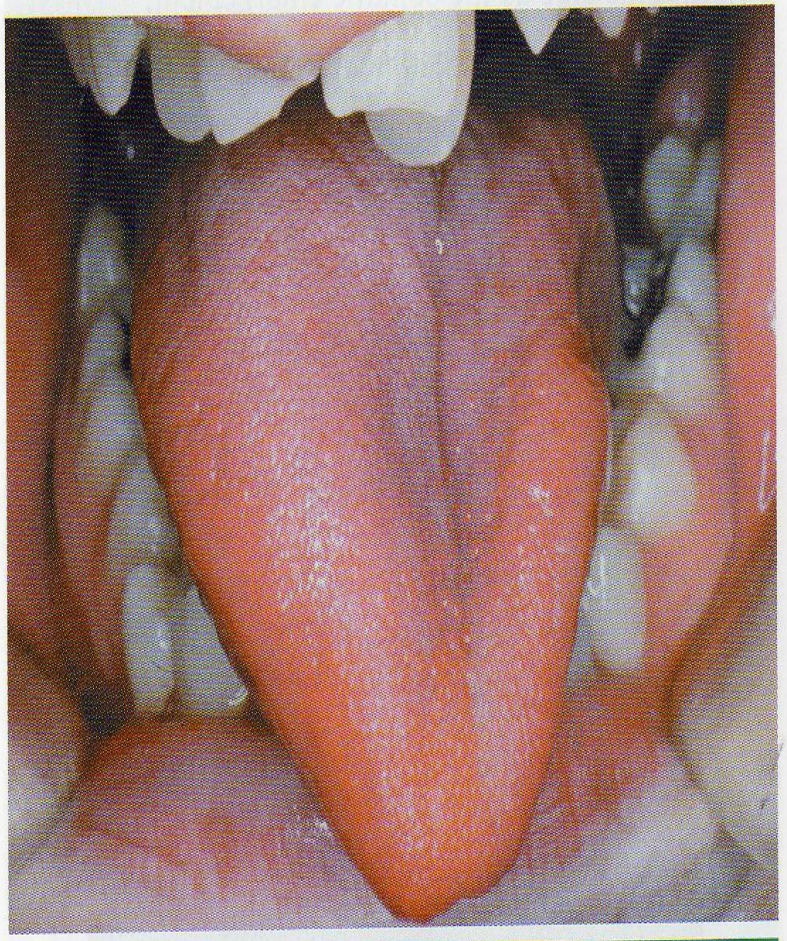


Figure 1-78 • Hemihyperplasia. Same patient as depicted in Figure 1-77, with associated enlargement of the right half of the tongue. (Courtesy of Dr. George Blozis.)

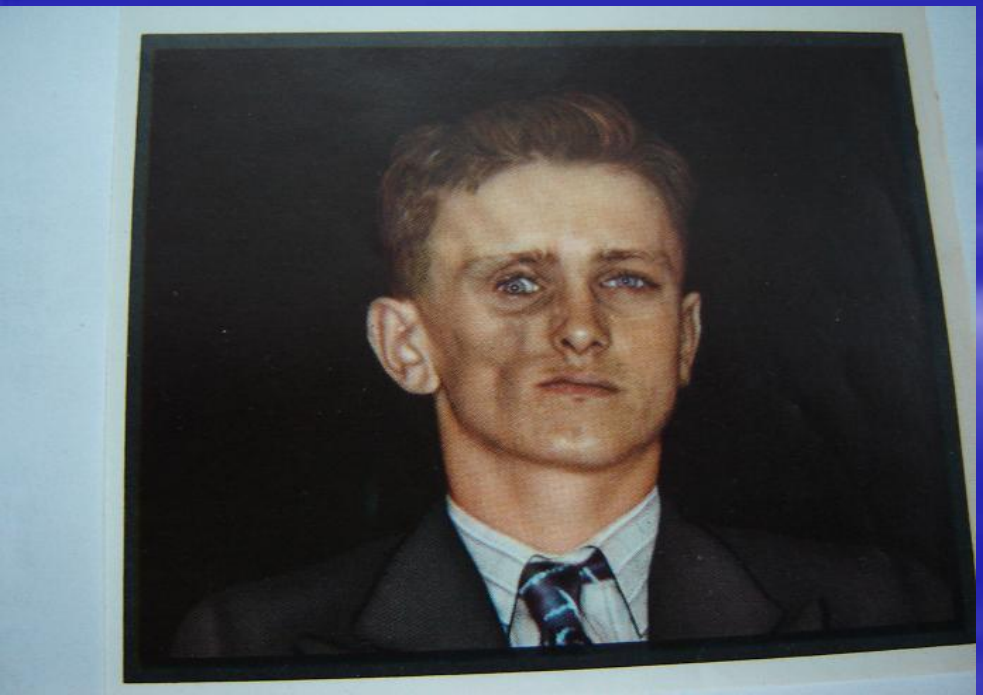
D.Hemifacial atrophy

-Un common, regenerative condition characterized :

1.Atrophic change affected one side of the face.

2.Mouth and nose deviated to the defective side.

3.Overlying skin presented with dark pigmentation.



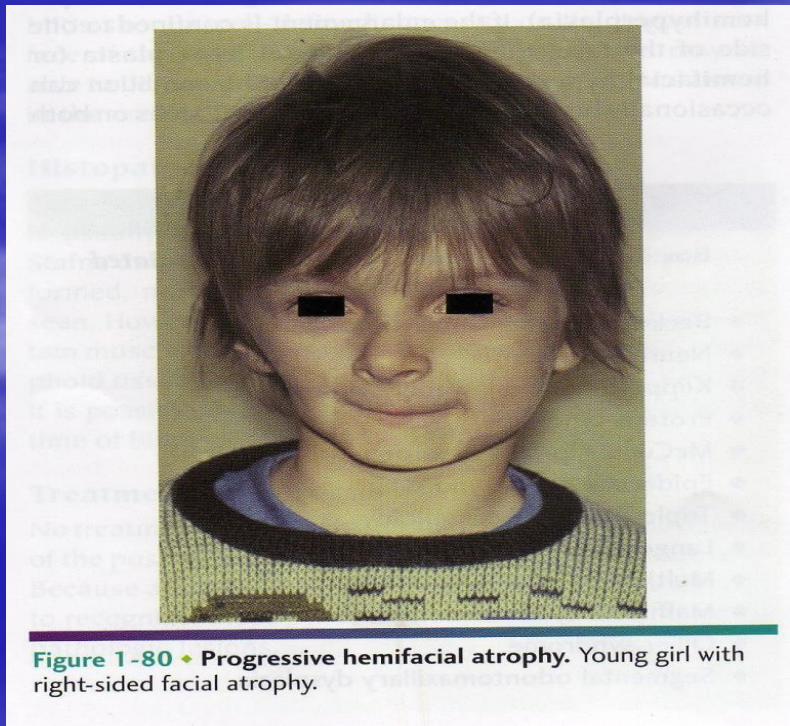


Figure 1-80 • **Progressive hemifacial atrophy.** Young girl with right-sided facial atrophy.

E. Bony Exostosis

Localized bony protuberance arising from normal cortical plate

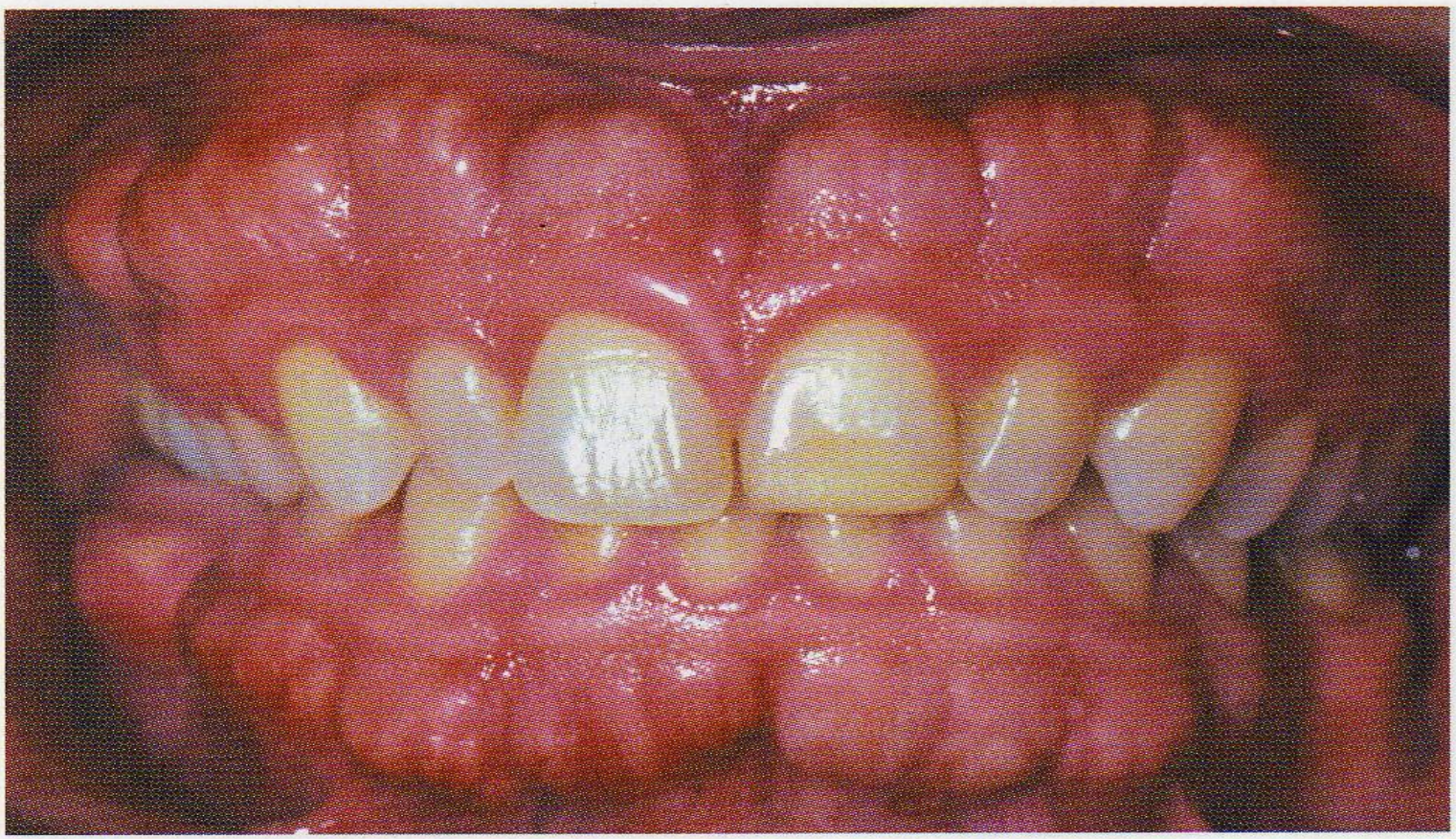


Figure 1-35 • Exostoses. Multiple buccal exostoses of the maxillary and mandibular alveolar ridges.

1.Torus palatinus :-

- Common in midline of vault of palate.
- Classified according to their morphology into:-

a.Flat torus , which has broad base

b.Spindle torus, appears as a midline ridge

c.Nodular torus, appears as a multiple protuberances.

d.Lobular torus , appears as a lobulated mass arise from single

se.

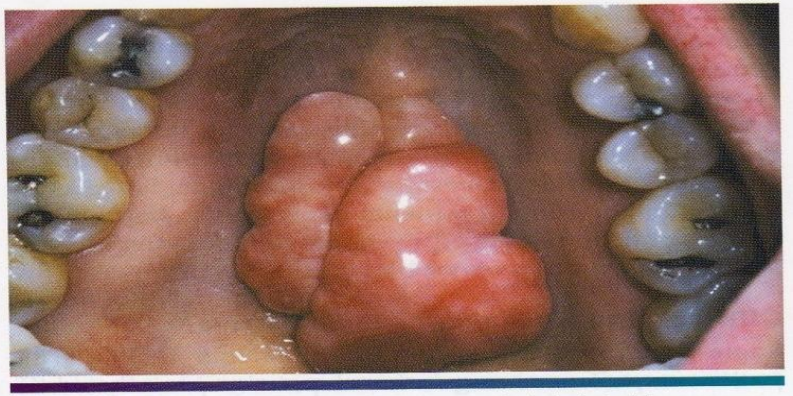


Figure 1-40 • **Torus palatinus.** Asymmetric, lobulated bony mass.

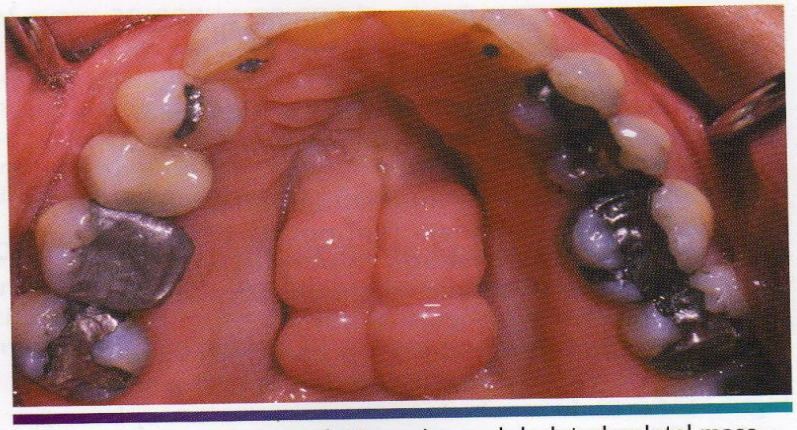


Figure 1-39 • **Torus palatinus.** Large, lobulated palatal mass.

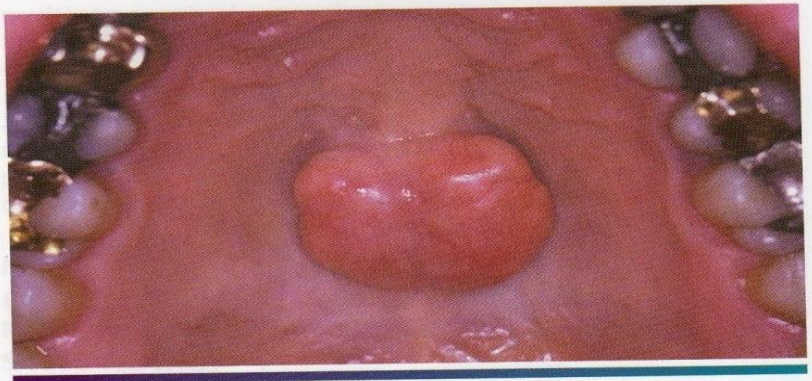


Figure 1-38 • **Torus palatinus.** Midline bony nodule of the palatal vault.

2- Tours mandibulares:-

-Bony protuberance along lingual aspect of mandible, above mylohyoid line, in premolar region.

-Bilateral in 90% of cases or as a single.

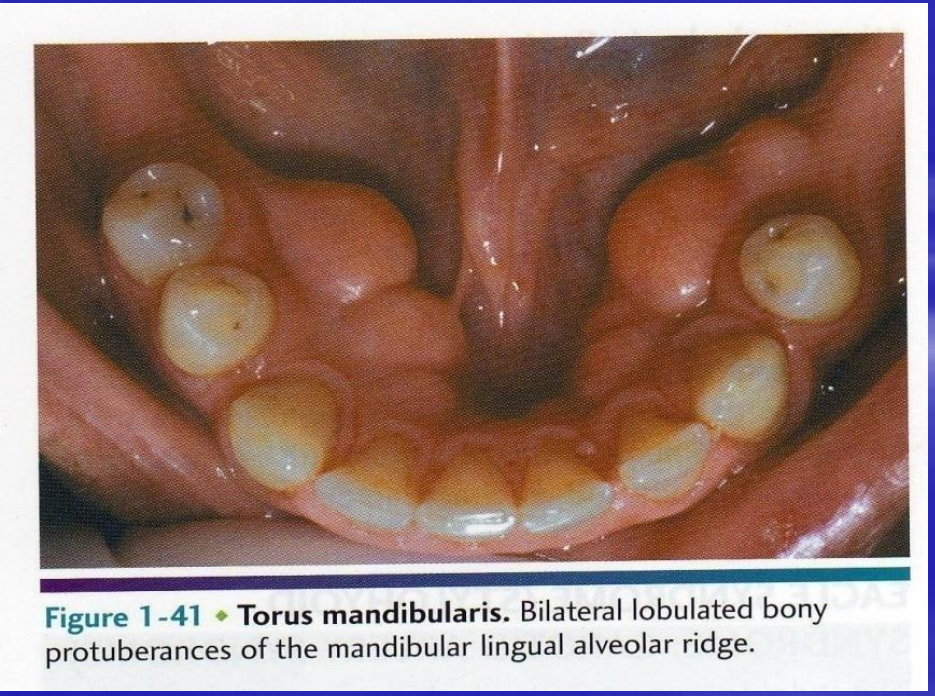


Figure 1-41 • **Torus mandibularis.** Bilateral lobulated bony protuberances of the mandibular lingual alveolar ridge.

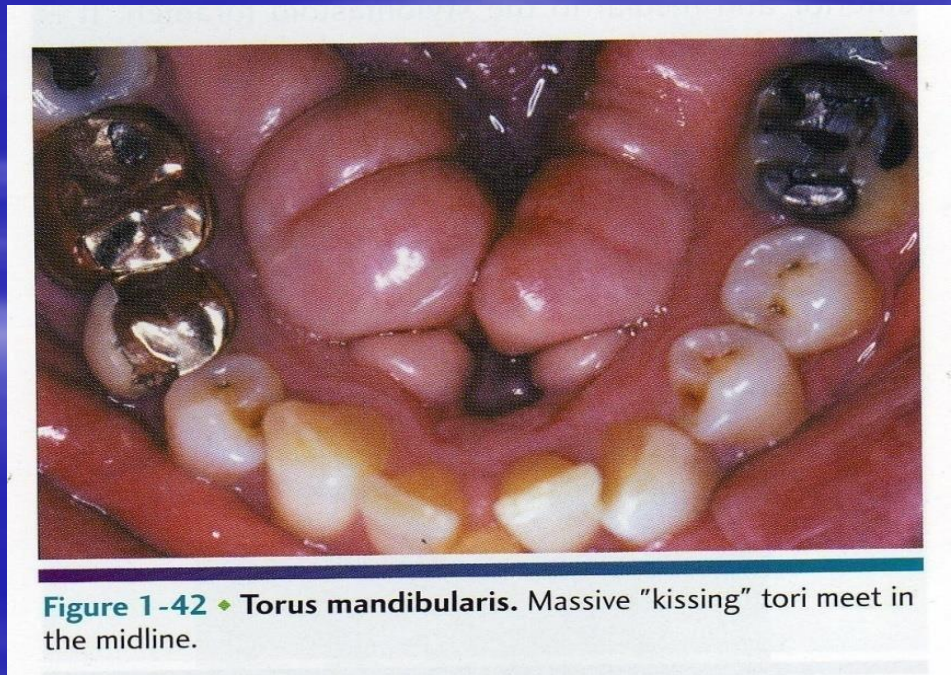


Figure 1-42 • **Torus mandibularis.** Massive "kissing" tori meet in the midline.

F- Clidocranial dysplasia (dysostosis)

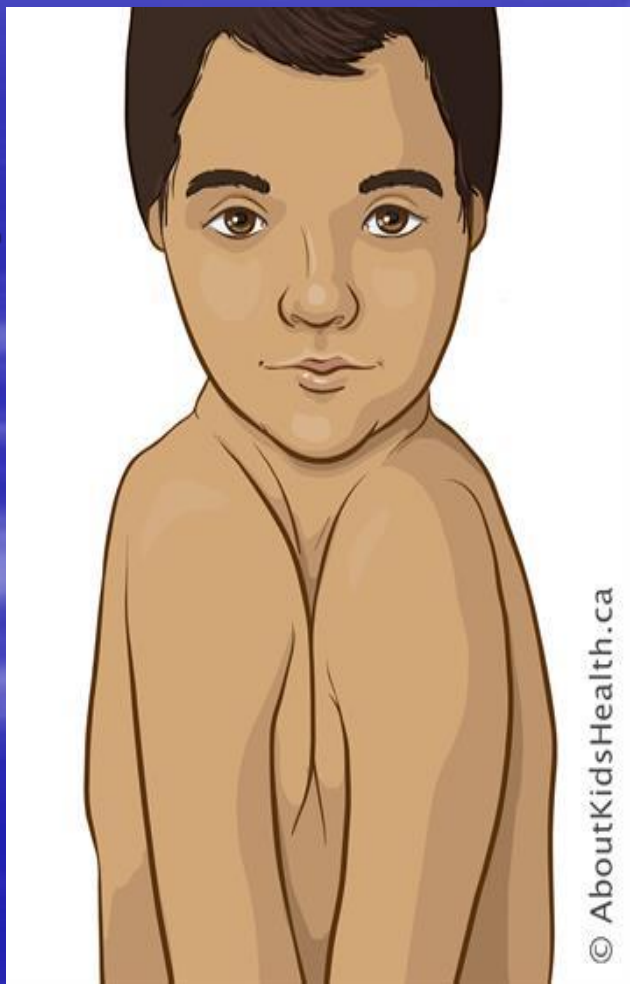
-Abnormal growth of facial bone, skull and clavicle with failure of tooth eruption.

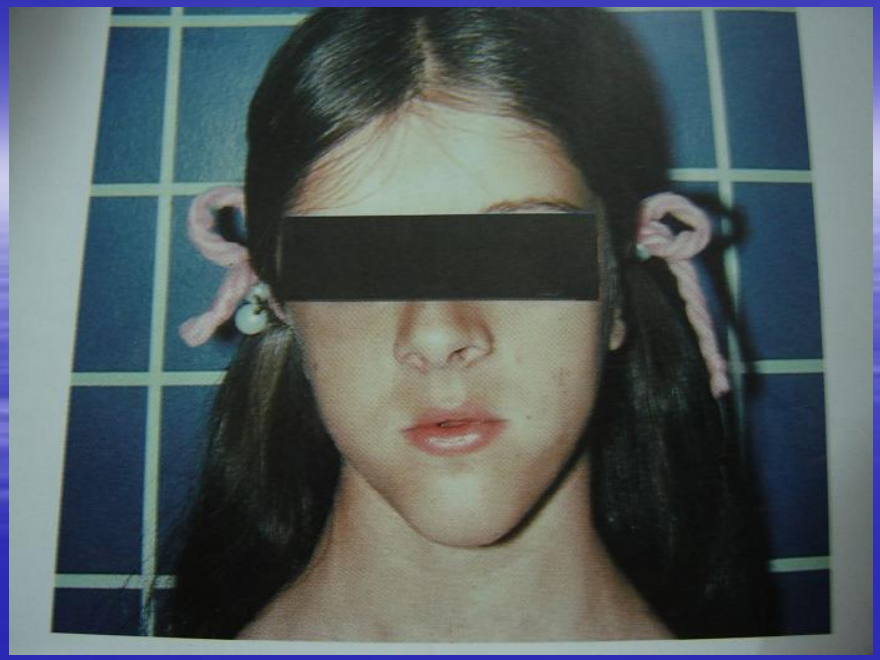
-Patient able to appose the shoulder near the midline of chest.

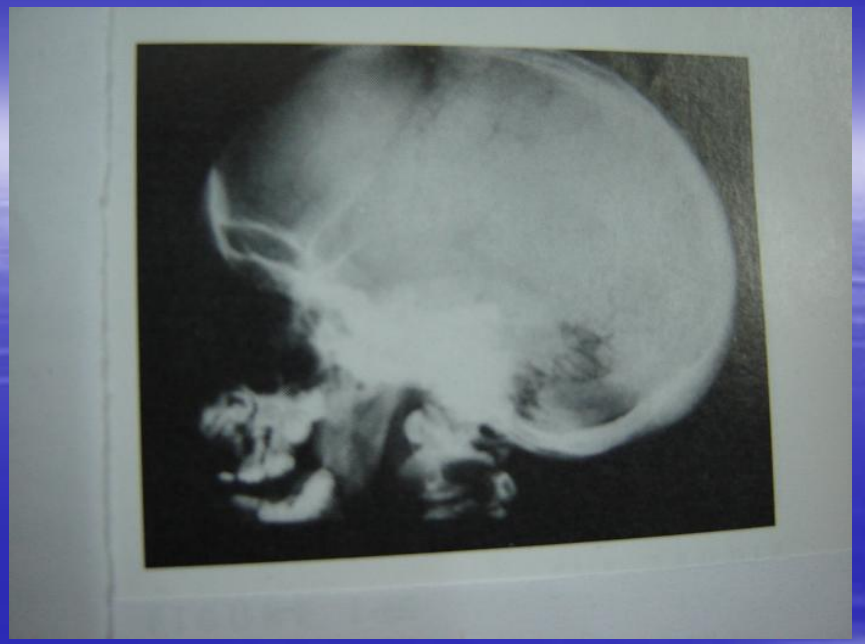
-Face with boosing frontal bone, depressed midface, & prominent chin.

-Patient retain primary dentition into adulthood.

-Supernumerary teeth may seen radiographically.







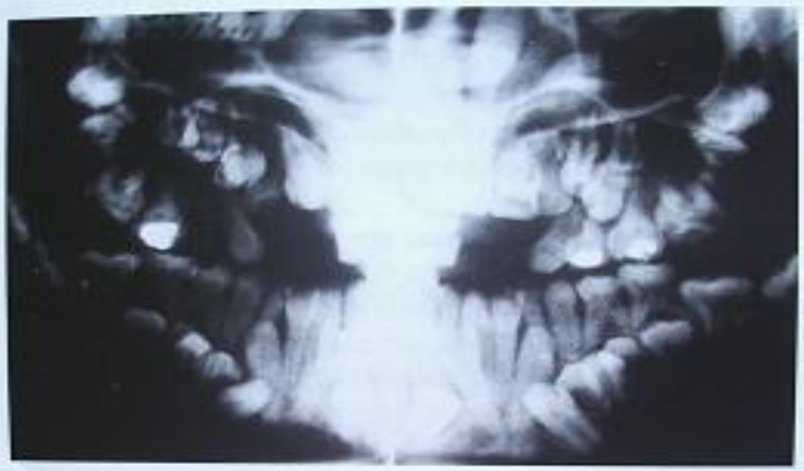
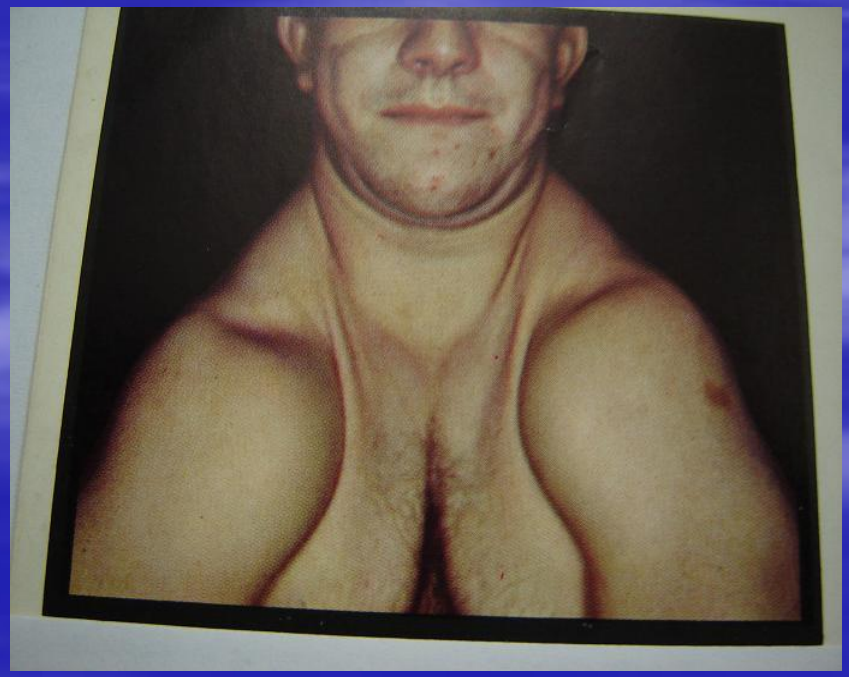


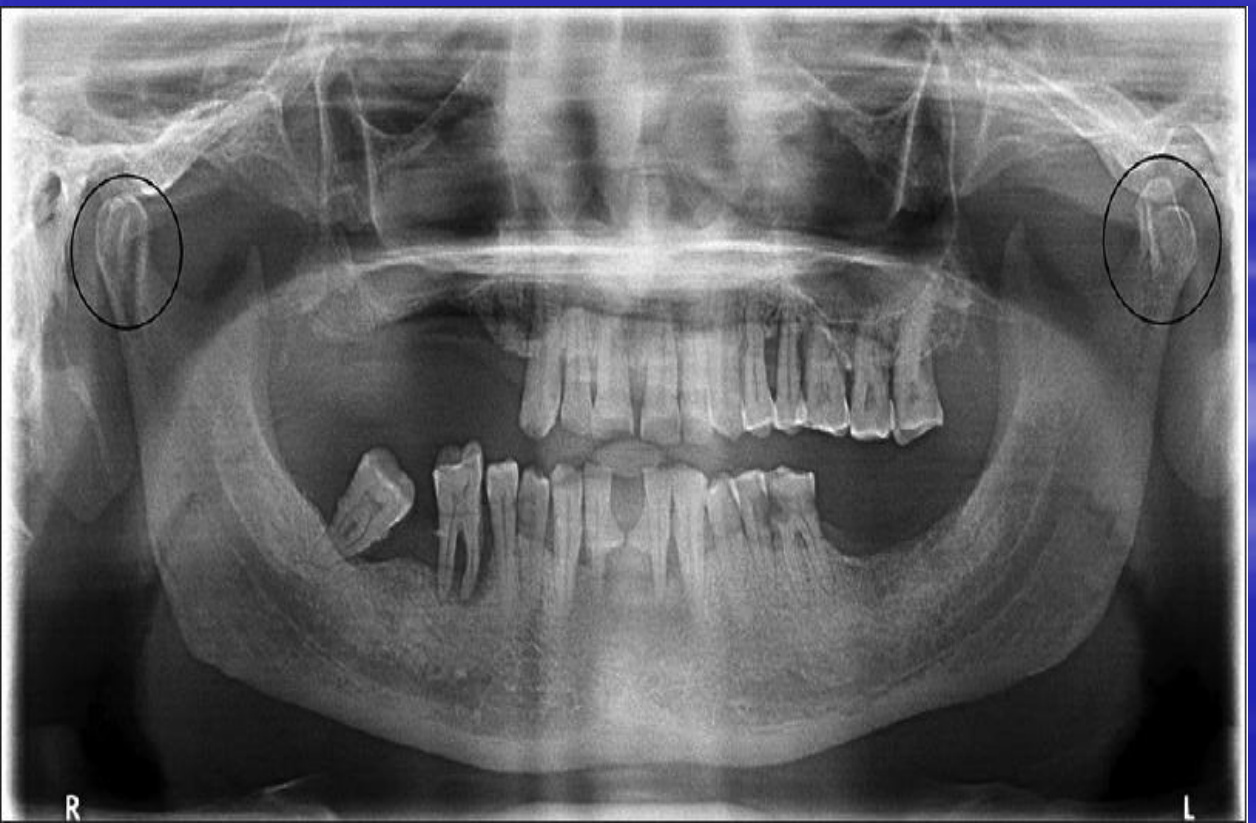
FIGURE 1-63

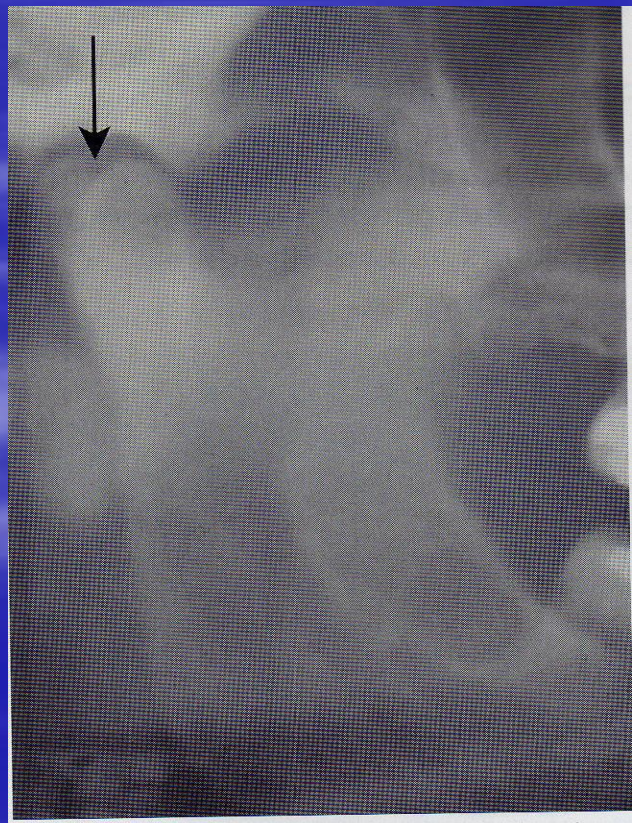
Cleidocranial dysplasia. Panoramic radiograph of dentition demonstrating the presence of a permanent dentition with associated supernumerary teeth embedded within the mandible and maxilla and deciduous dentition that has failed to exfoliate.

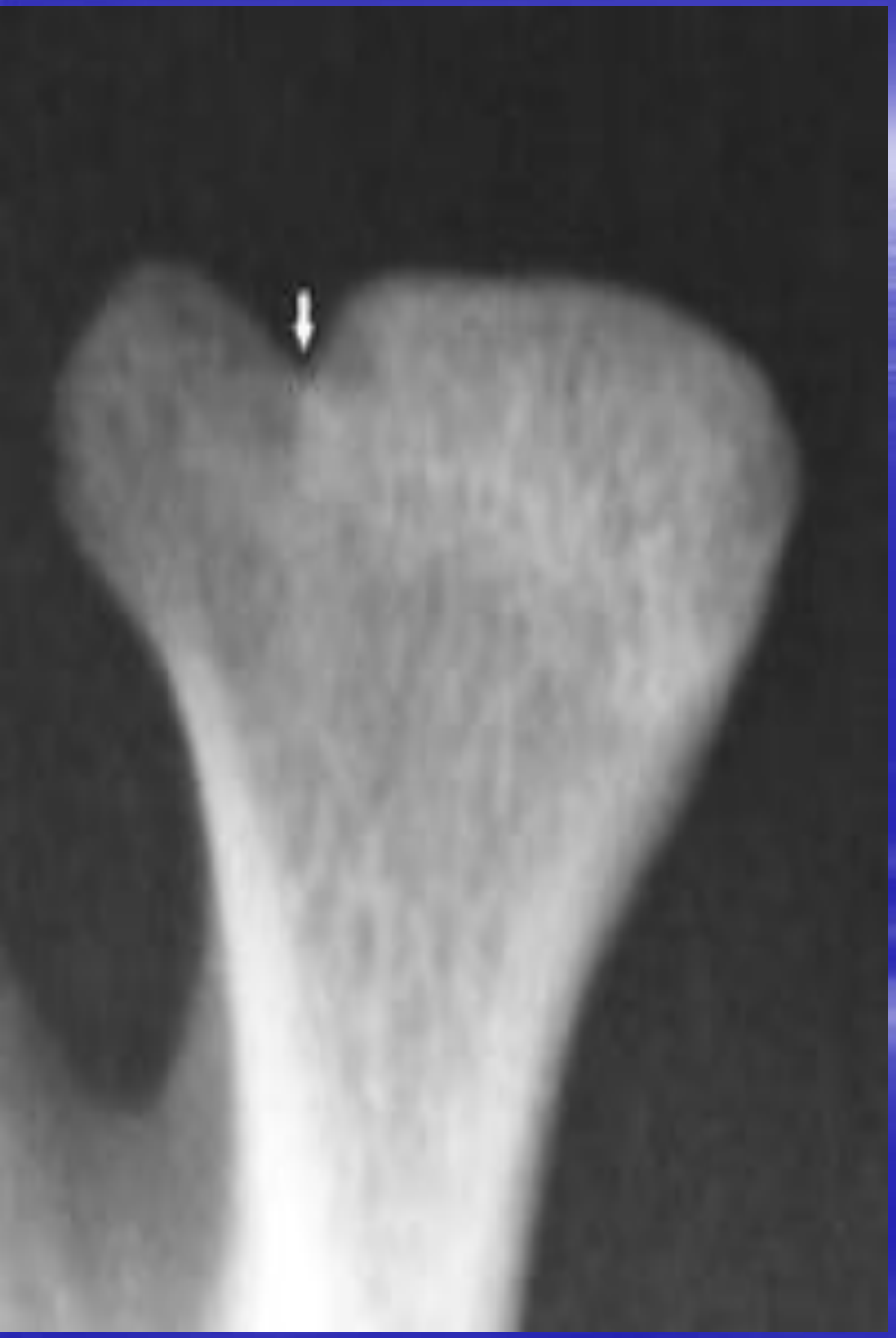


G- Bifid condyl

- Double-headed mandibular condyl of uncertain cause.
- Antero-posterior bifid condyl may be traumatic in origin during childhood.
- Medio-lateral one may result from abnormal muscle attachment.







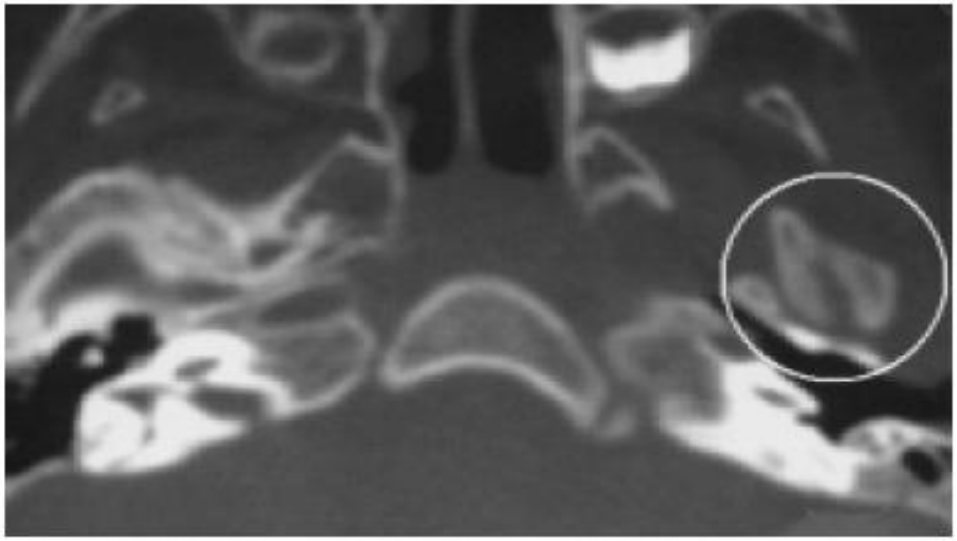
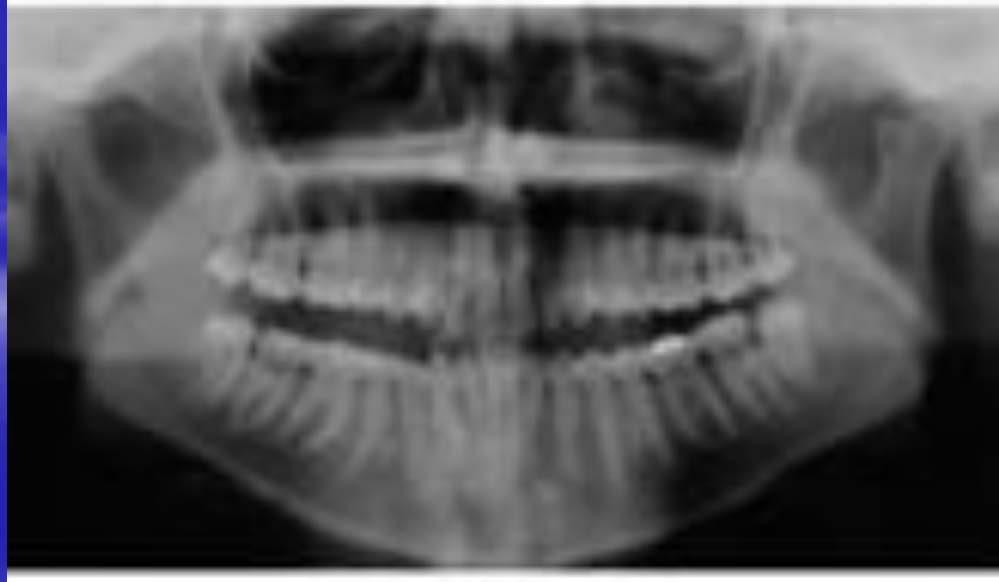


Figure 3. 2D-CT axial view (bone window) demonstrates the bifid mandibular condyle in the left side with mediolateral heads. (white circle).



H-Mandibular dysostosis (Treacher-Collins Syndrom)

- Autosomal dominant disorder characterized by:-
- -Hypoplastic zygoma, resulting in narrow face with depressed cheek &downward slanting of palpebral fissures.
- -Underdeveloped mandible with retruded chin &cleft palate may be seen.

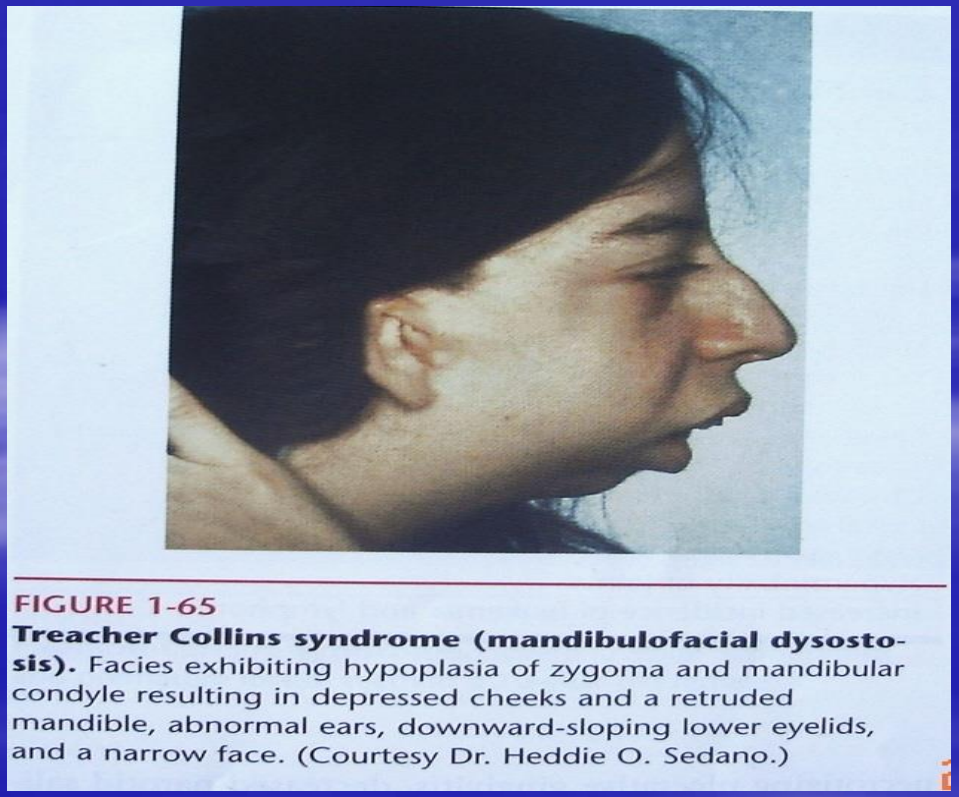
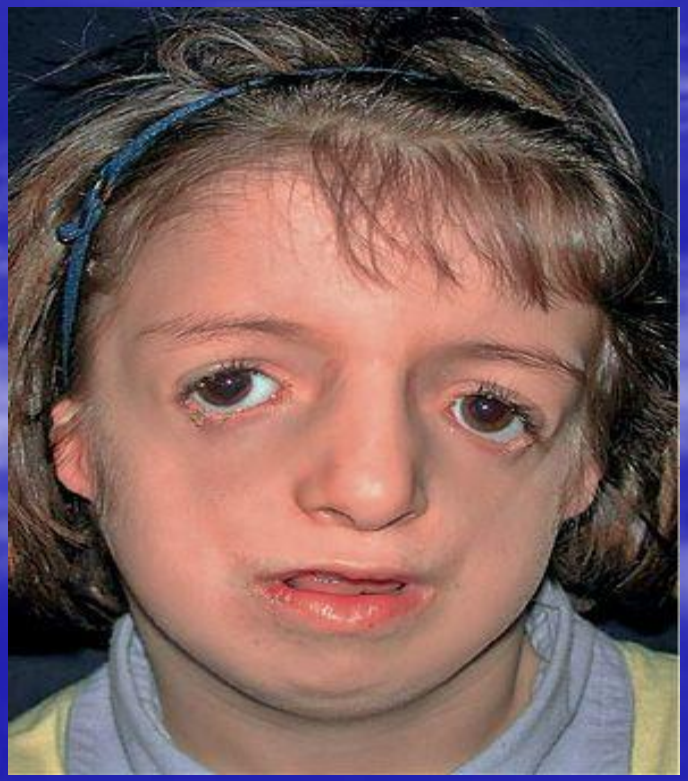


FIGURE 1-65
Treacher Collins syndrome (mandibulofacial dysostosis). Facies exhibiting hypoplasia of zygoma and mandibular condyle resulting in depressed cheeks and a retruded mandible, abnormal ears, downward-sloping lower eyelids, and a narrow face. (Courtesy Dr. Heddie O. Sedano.)



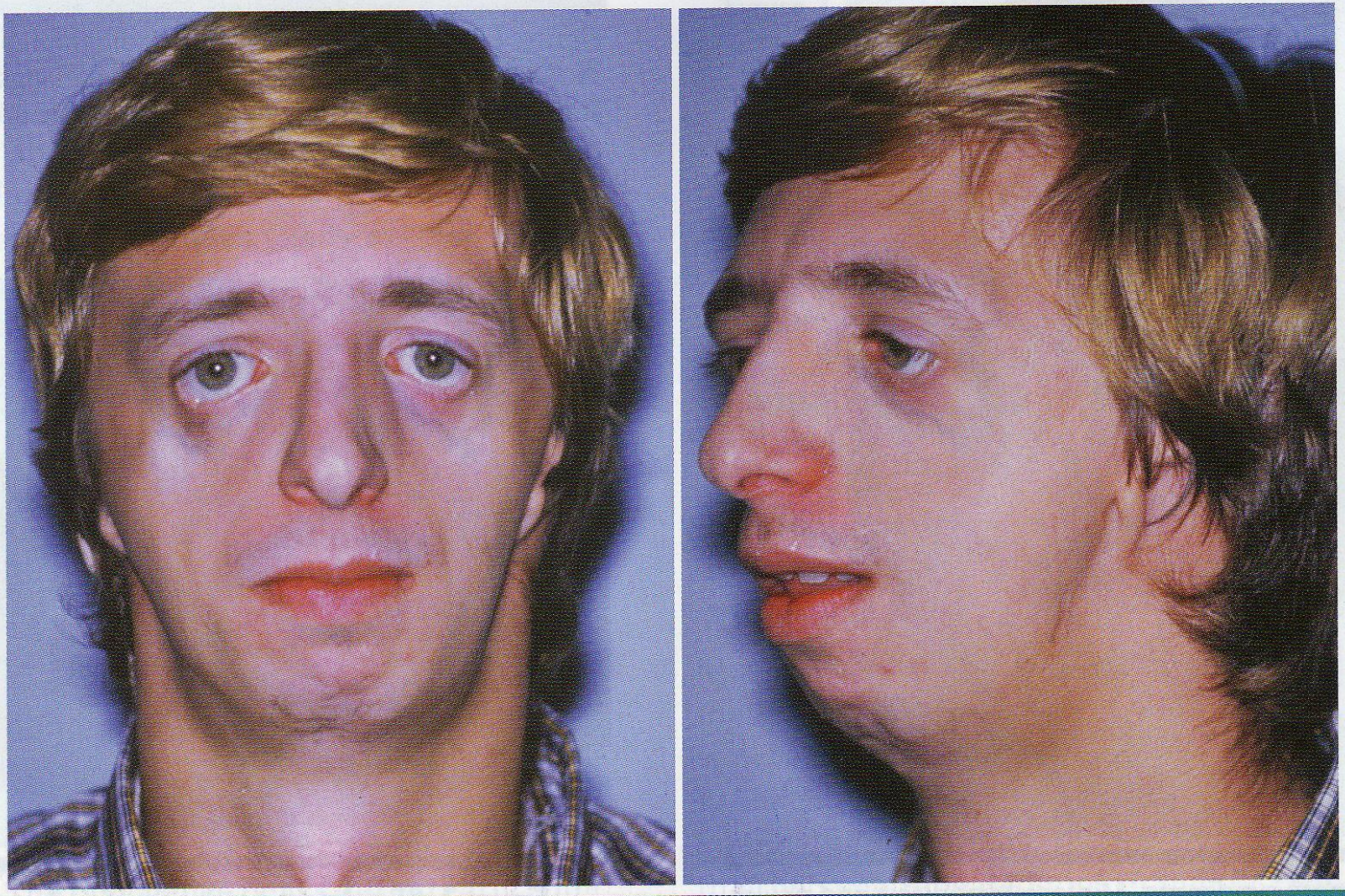
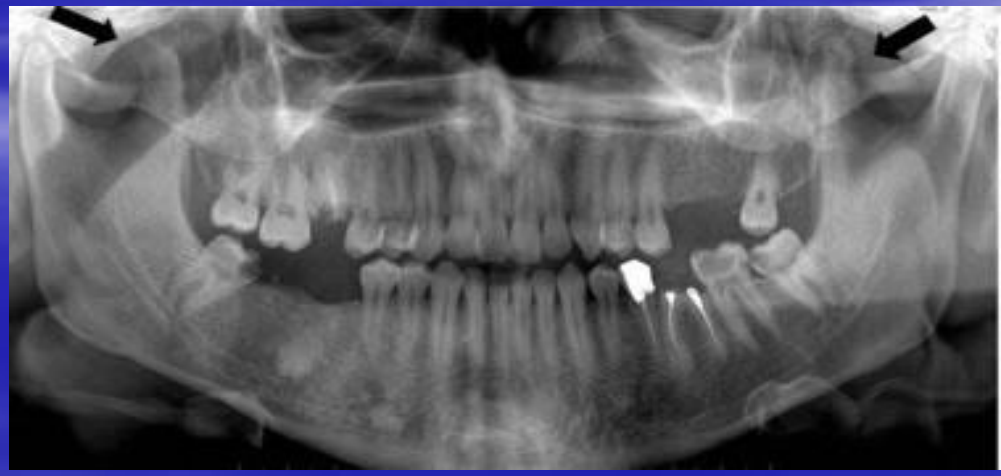


Figure 1-87 • Mandibulofacial dysostosis. Patient exhibits a hypoplastic mandible, downward-slanting palpebral fissures, and ear deformities. (Courtesy of Dr. Tom Brock.)

- **I-Coronoid hyperplasia**
- Rare developmental anomalies ,result in limitation of mandibular movement.
- The condition may be:-

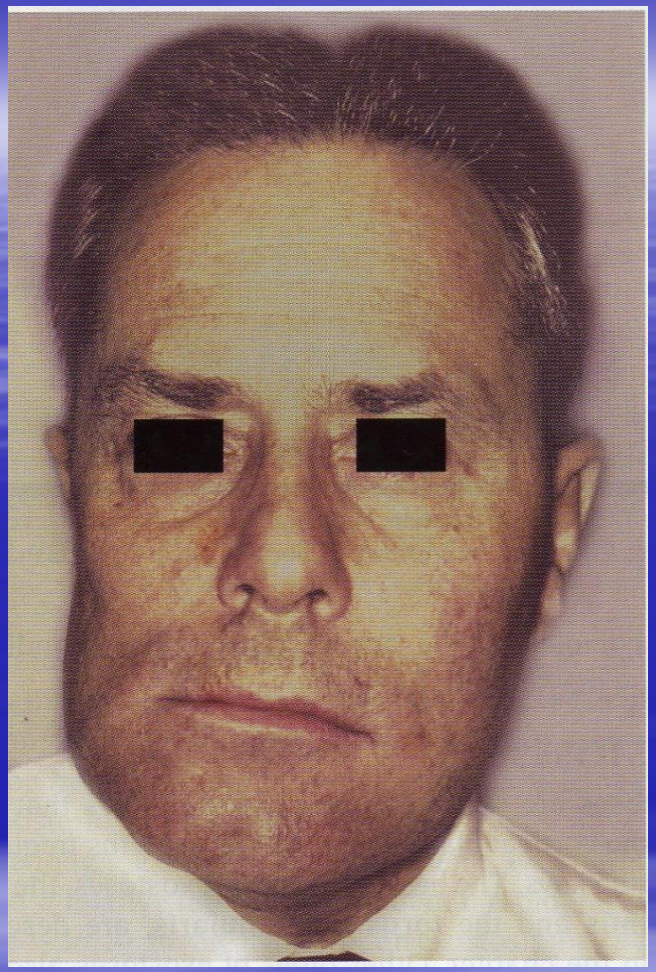
  - Unilateral, result from osteoma & osteosarcoma .

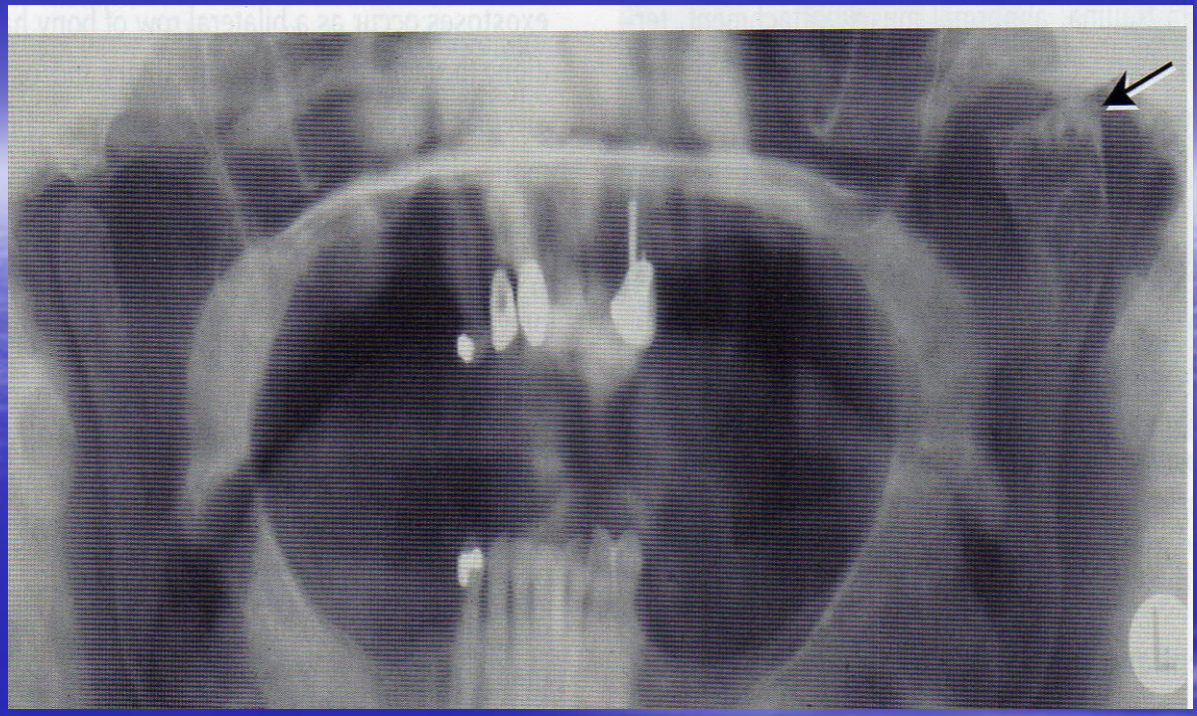
  -Bilateral, result from endocrine influence during puberty.

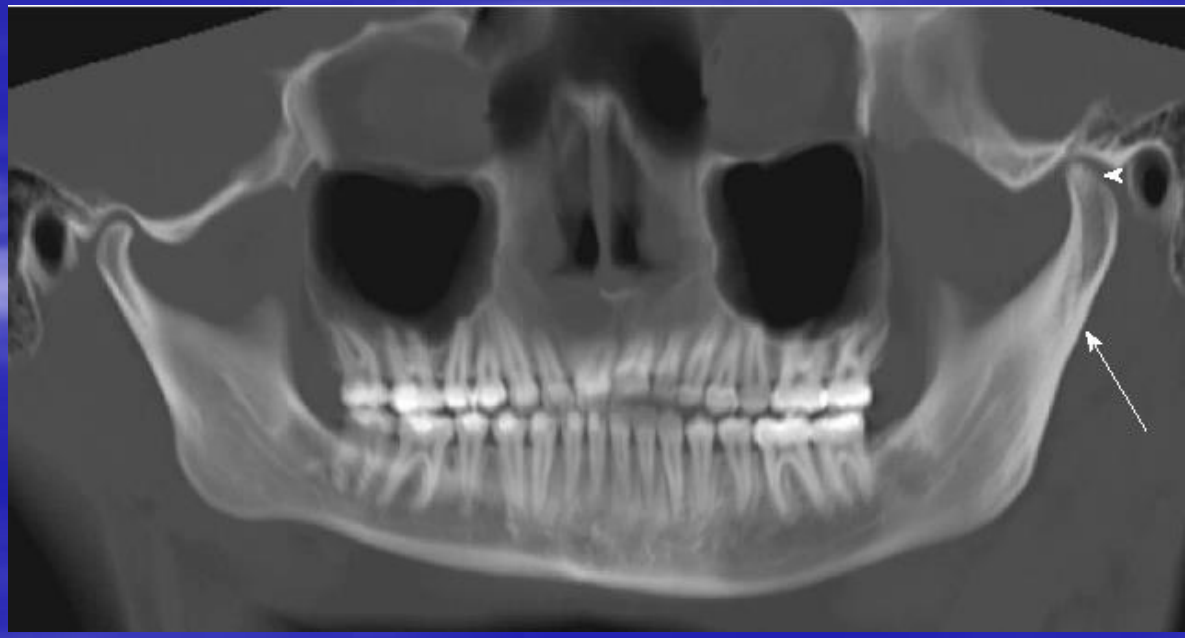


J-Condylar hyperplasia

- Excessive growth of one condyl .
- Unknown cause
- Local circulatory problems → such as endocrine disturbances & truma may be a possible etiologic factor.







K- Condylar hypoplasia

- ***Congenital*** :- Associated with **"Mandibular dysostosis"** .
- ***Acquired*** :- Result from disturbances of growth center of the developing condyl, secondary to truma, radiation , or rheumatoid arithritis.