CLEFT LIP AND PALATE

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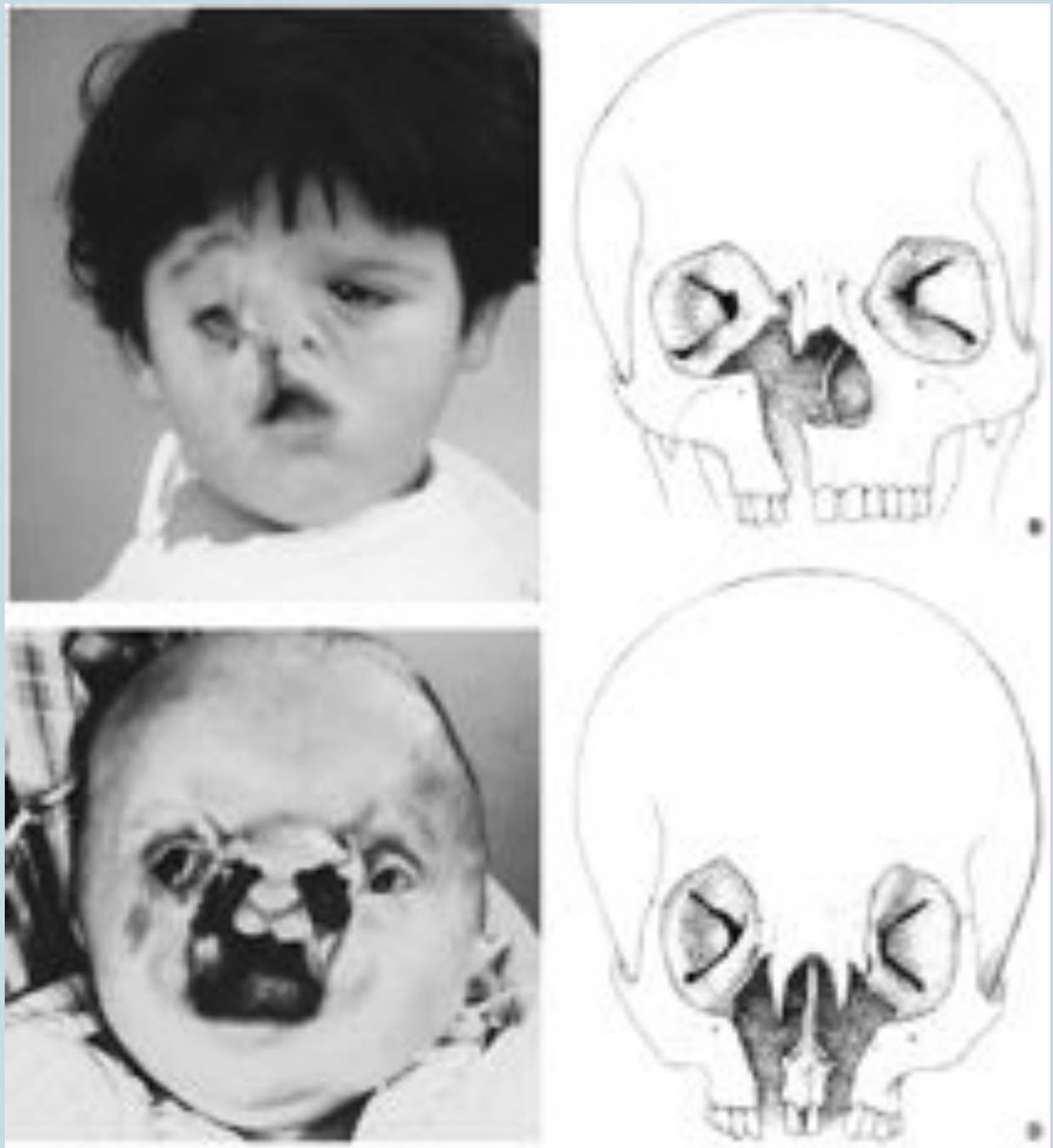
Three Types of Dysmorphogenesis

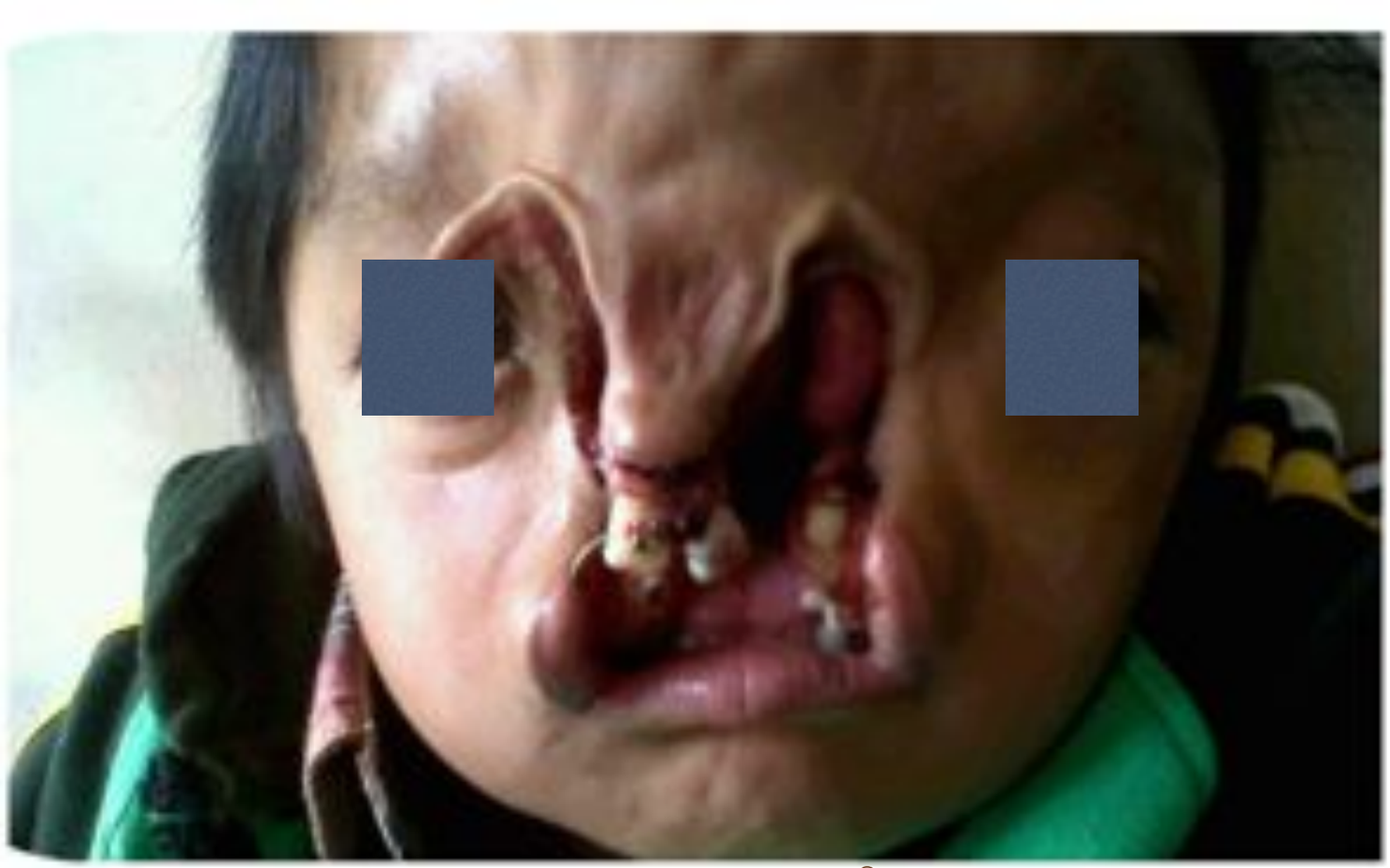
Types of Anomaly	Developmental Process	Craniofacial examples
Malformation	Abnormal development of tissue	Cleft lip and palate, microcephaly
Deformation	Unusual forces on normal tissue	Positional plagiocephaly, Robin sequence
Disruption	Breakdown of normal tissue	Hemifacial microsomia, rare facial clefts

Other classification ➜ Committee on Nomenclature and Classification of Craniofacial Anomaly of the American Cleft Palate Association in 1981

- I. Facial clefts/encephaloceles
- II. Atrophy/hypoplasia
- III. Neoplasia/hyperplasia
- IV. Craniosynostosis
- V. Unclassified

Facial Cleft





Encephalocele

Hypoplasia /atrophy

Romberg Disease

Fig. 3. A. Patient with 2 years and 8 month, without syndrome alterations; B. Patient with 5 years old with mild enophthalmic signs and facial atrophy in right side; C. With 11 years old, the exophthalmia is evident, 'coup de sabre' scar in parasinfisis region and mid right maxillary lip featuring the hemifacial atrophy.

Hemifacial Microsomia

Neurofibrome/ neurofibromatosis

Definition of CLP

- Ceft Lip and Palate is congenital anomaly, characterized by varying degrees of separation of the lip, alveolus and palate with or without nose distorsion

Introduction

- A TEAM APPROACH IS REQUIRED
  - Plastic/ general surgeon
  - Anesthetician
  - Pediatrician
  - Orthodontist
  - ENT
  - Psychiatrist
  - Speech therapist
  - Nurse coordinator
  - Social worker

Introduction

- Most common congenital malformation of H and N (1:1000 in US; 1:600 in UK)
- Second most common overall (behind club foot)
- cleft lip and palate at 46%, followed by isolated
- cleft palate at 33%, then isolated cleft lip at 21%

Epidemiology

- Syndromic CLAP
  - associated with more than 300 malformations
  - Pierre Robin Sequence; Treacher-Collins, Trisomies 13,18,21, Apert's, Stickler's, Waardenburg's, hemifacial microsomia
- Nonsyndromic CLAP
  - diagnosis of exclusion

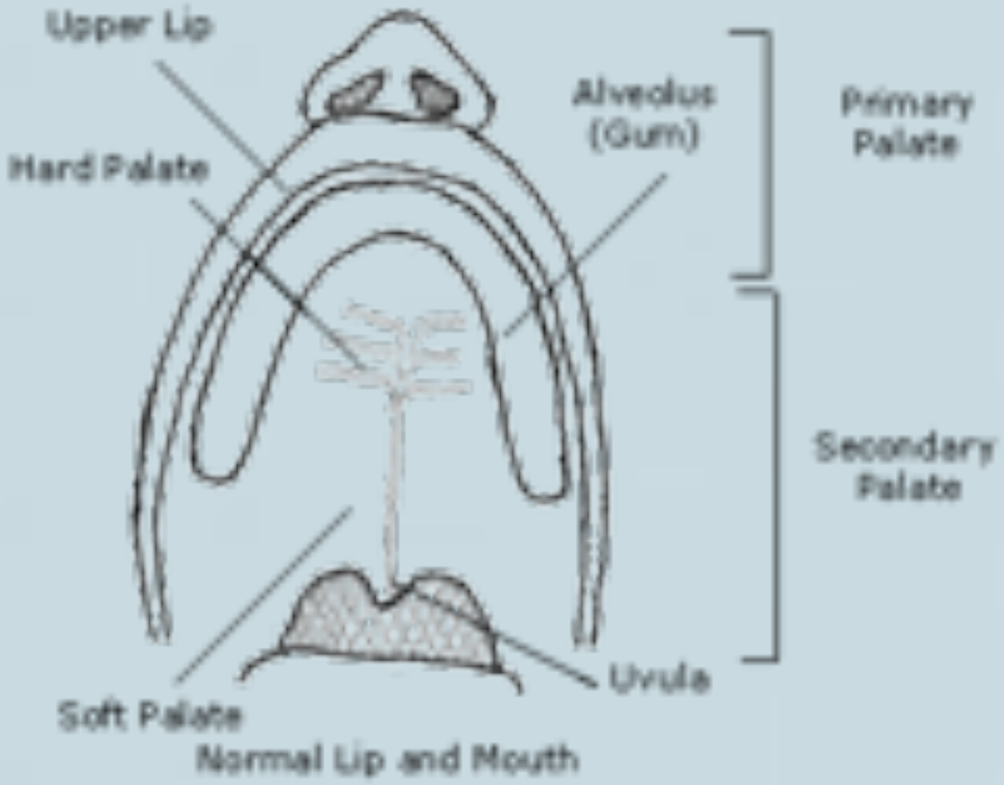
Apert Syndrome

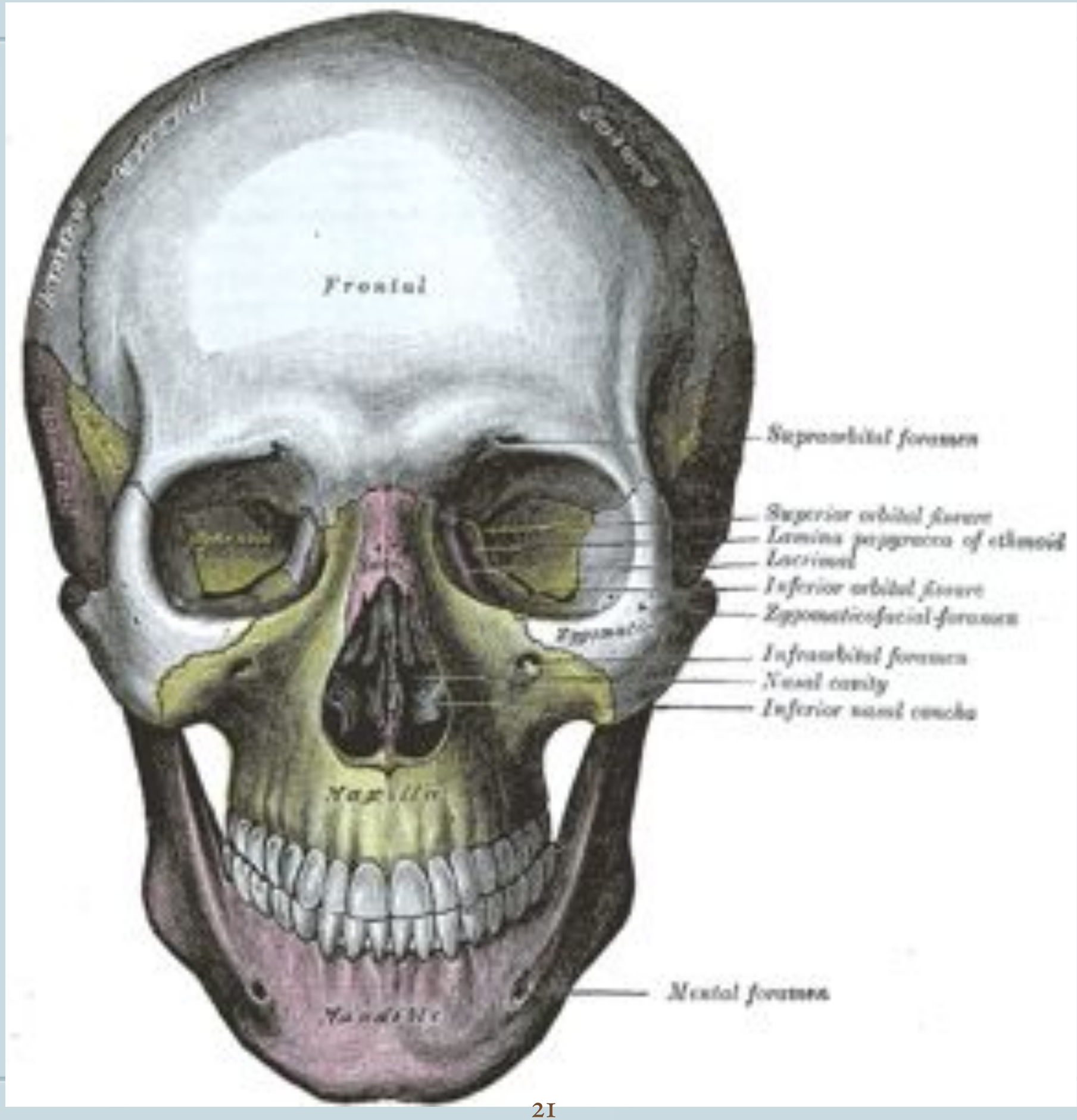
Embryology

- Primary versus secondary palate
  - divided by incisive foramen
  - primary palate develops 4-5 wks
  - secondary palate develops 8-9 wks
- Primary palate
  - mesodermal proliferation of frontonasal and maxillary processes
  - never a cleft in normal development

- Secondary palate
  - medial ingrowth of lateral maxillae with midline fusion
  - always a cleft in normal development
  - macroglossia, micrognathia may provide anatomical barriers to fusion

Anatomy- normal




Frontal
Supraorbital foramen
Superior orbital fissure
Lamina papyracea of ethmoid
Lacrimal
Inferior orbital fissure
Zygomaticofacial foramen
Infraorbital foramen
Nasal cavity
Inferior nasal concha
Mental foramen

Topography of the lip

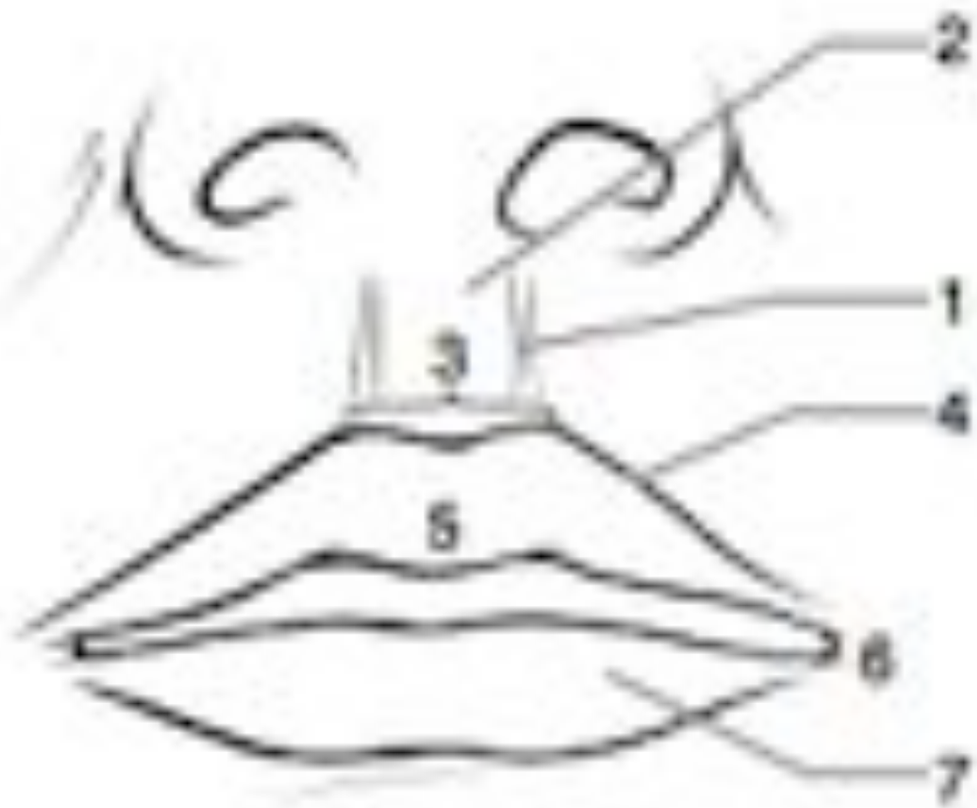
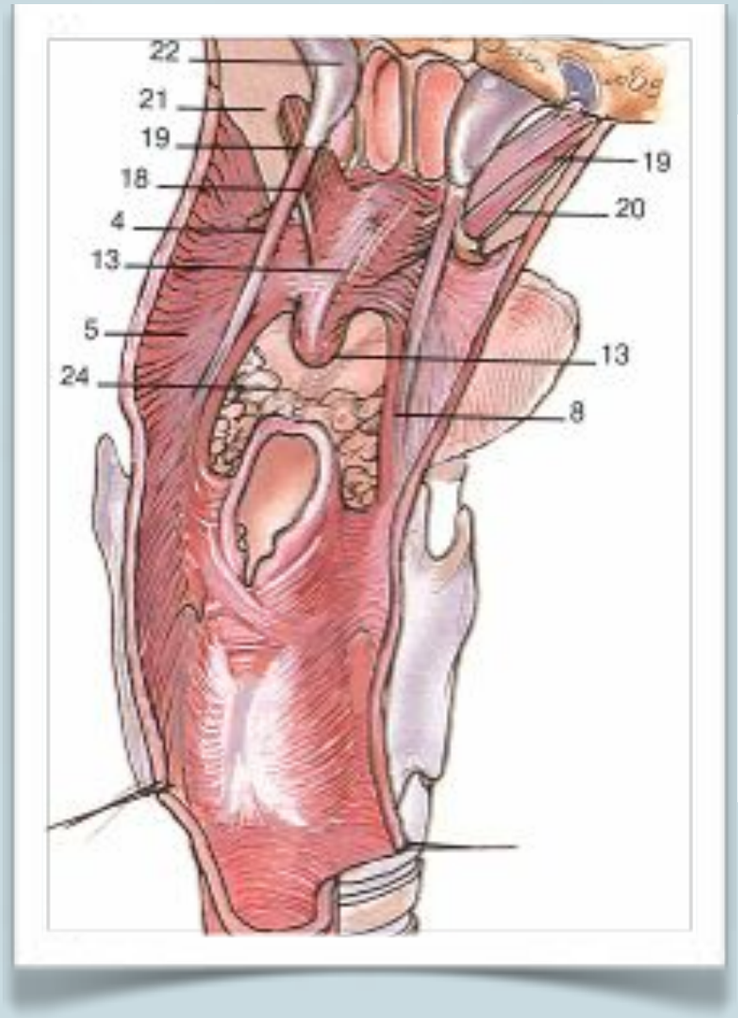
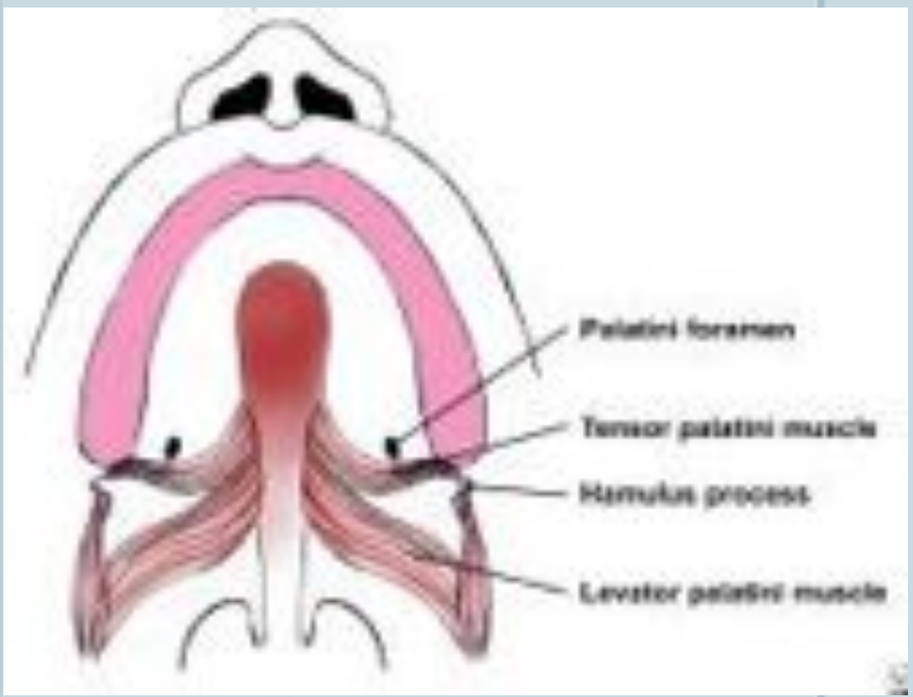


FIGURE 36.2. Topographic anatomy of the lips. 1, Philtral columns. 2, Philtral groove or dimple. 3, Cupid's bow. 4, White roll upper lip. 5, Tubercle. 6, Commissure. 7, Vermilion. (Redrawn after Zide BM. Deformities of the lips and cheeks. In: McCarthy JE, ed. *Plastic Surgery*. Philadelphia: Saunders; 1990-2009.)

Anatomy palatal muscle

- Muscles
  - Superior constrictor
  - primary sphincter
  - Tensor veli palatini
  - tenses palate
  - Levator Veli palatini
  - elevates palate
  - dilates ET
  - Salpingopharyngeus, palatopharyngeous, palatoglossus: minor contribution



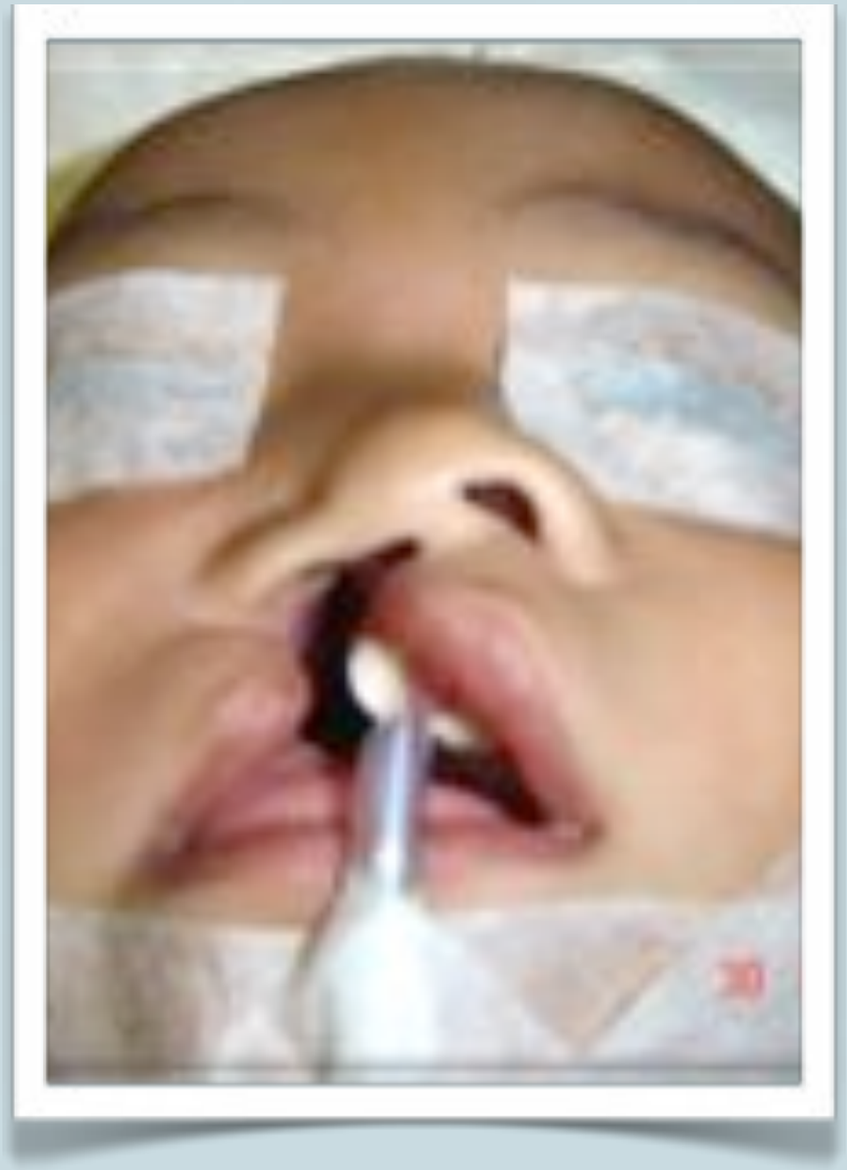


Cleft anatomy

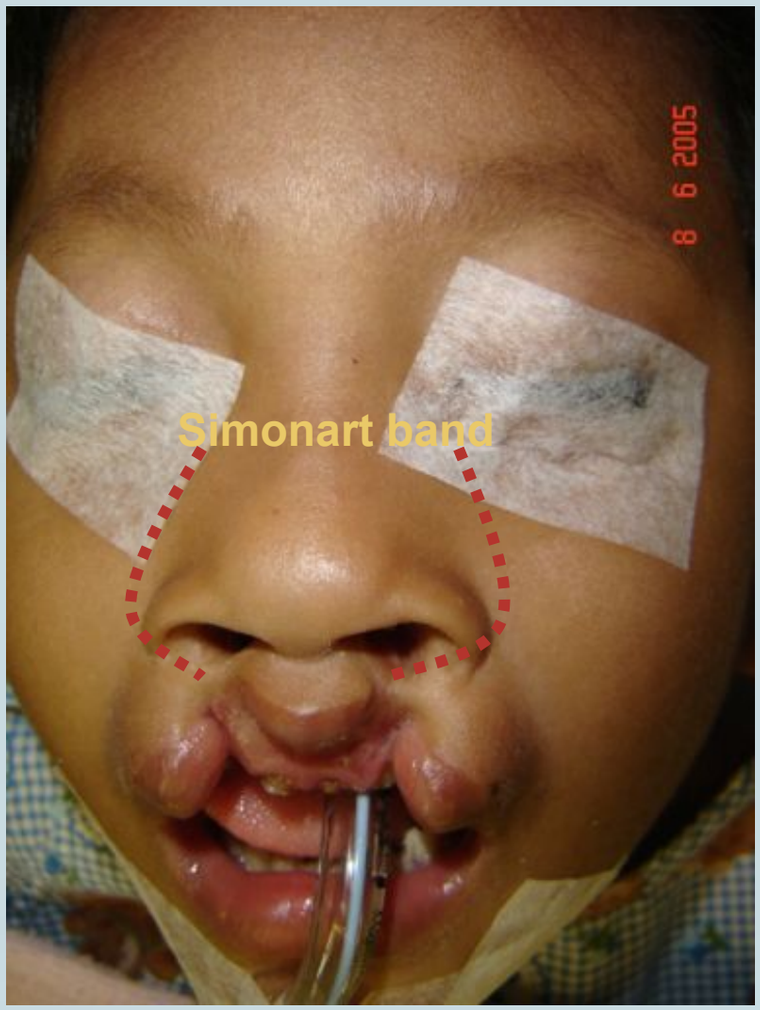
- lack of mesodermal proliferation
- cleft of orbicularis
- medial portion to columella
- lateral portion to nasal ala
- cleft of alveolus

The nose

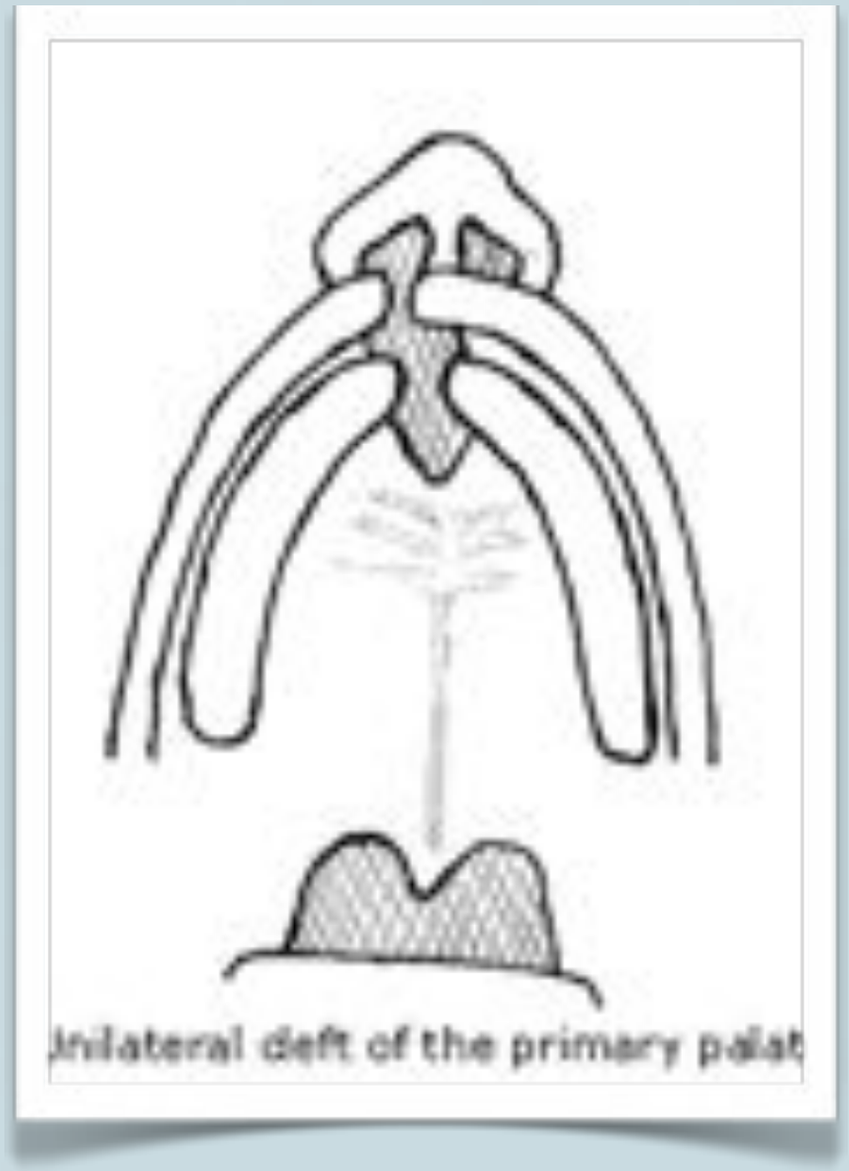
- flattened
- rotated downward
- Short columella
- Bifid tip



Bilateral incomplete cleft

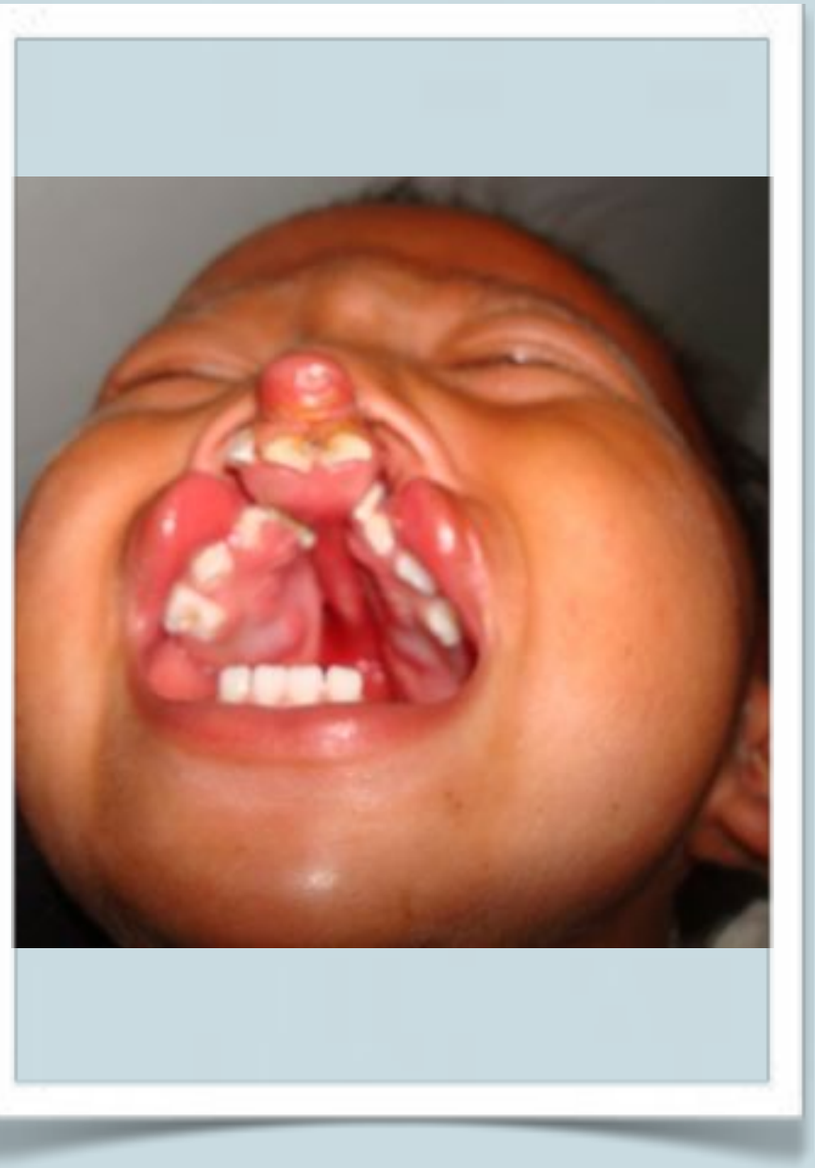


- Clefts of the primary hard palate/ alveolus
- cleft alveolus always associated with cleft lip
- cleft lip not necessarily associated with cleft alveolus
- By definition there is opening into nose



- Clefts of secondary palate
- Failure of medial growth maxillae
  - fusion at incisive foramen
  - macroglossia
- Submucous vs. complete
- Vomer

- Bilateral Cleft Lip/Alveolus/nose
  - duplication of unilateral defect
  - premaxilla
  - orbicularis to alar cartilages bilaterally
  - bifid tip
  - extremely short columella
  - Vomer



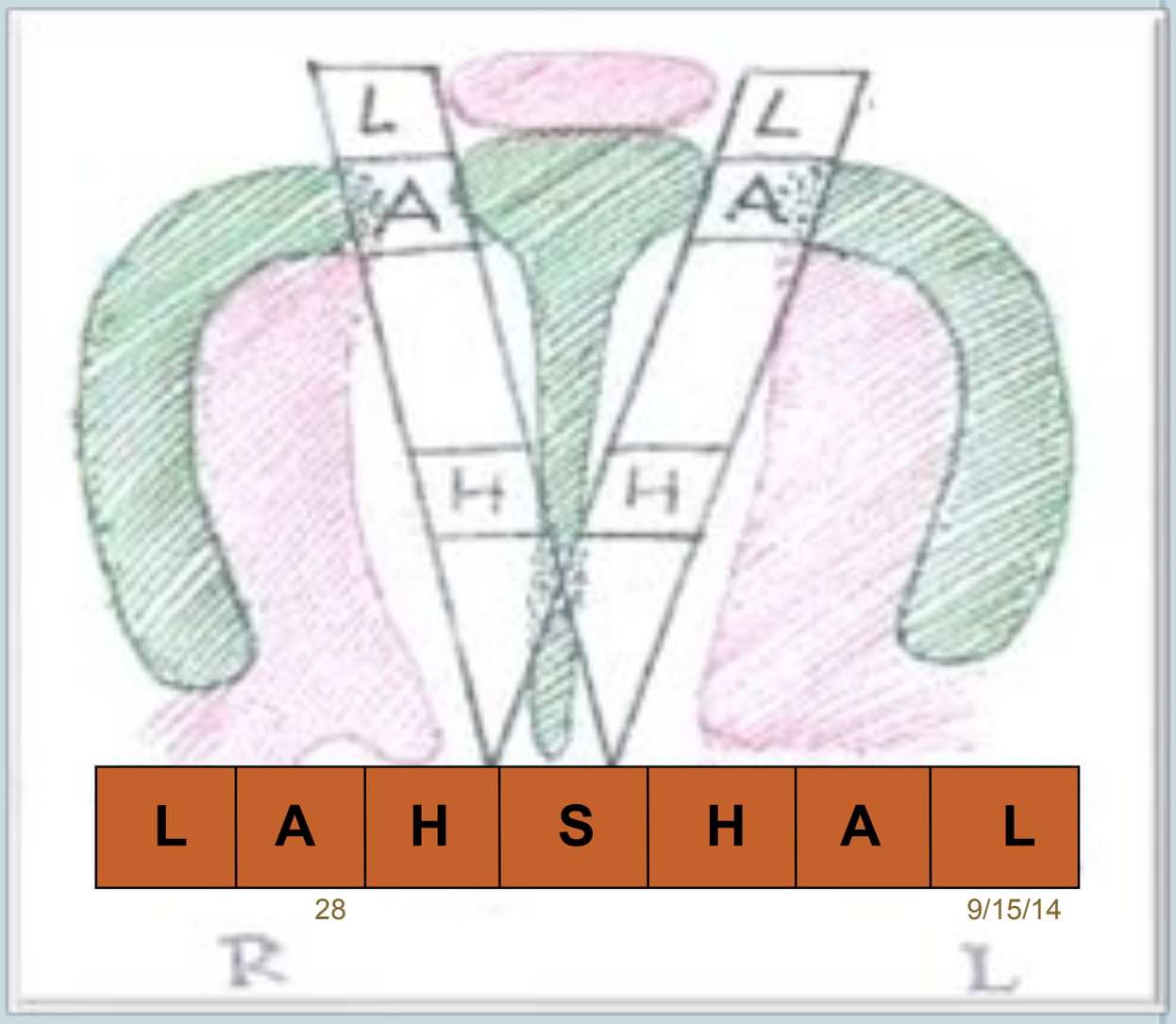
Classification

- Veau Classification - 1931
  - Veau Class I: isolated soft palate cleft
  - Veau Class II: isolated hard and soft palate
  - Veau Class III: unilateral CLAP
  - Veau Class IV: bilateral CLAP
- Iowa Classification - a variation of Veau Classification

- Complete Clefts
  - absence of any connection with extension into nose
  - vomer exposed
- Incomplete Clefts
  - midline attachment (may be only mucosal)
  - ex: submucous cleft (midline diasthasis, hard palatal notch, bifid uvula)

- Otto Kriens

•Complete : L
•Incomplete : l
•Microform :(l)



Simonart band

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Multi disciplinary approach

- These are not merely surgical problems

- Requires team approach throughout life
  - neonatal period
  - toddler
  - grade school
  - adolescence
  - young adulthood

SURGICAL TREATMENT OF A CLEFT LIP AND PALATE BY AGE

Age	Treatment	Cleft team members
Prenatal	Prenatal imaging, diagnosis, and counseling	Multidisciplinary
Newborn[a]	Feeding assessment, medical assessment, genetic counseling, treatment information	Multidisciplinary
0–3 months	Presurgical orthopedics	Orthodontist, plastic surgeon
3 months (or after presurgical orthopedics)[b]	Primary cleft lip repair and tip rhinoplasty ± gingivoperiosteoplasty	Plastic surgeon
12 months (delayed if airway or medical concerns)[c]	Primary cleft palate repair with intravelar veloplasty ± bilateral myringotomy and tubes	Plastic surgeon, otolaryngologist
Diagnosis of velopharyngeal insufficiency (3–4 years)	Secondary palate lengthening or pharyngoplasty, speech clearance	Speech pathologist, plastic surgeon, otolaryngologist, orthodontist
School-age years	Treatment of secondary lip and nasal deformities	Plastic surgeon
7–9 years (mixed dentition)[d]	Secondary alveolar bone graft	Orthodontist, plastic surgeon, oral surgeon
Postalveolar graft[e]	Presurgical orthodontics	Orthodontist
Puberty	Definitive open rhinoplasty	Plastic surgeon
Skeletal maturity	LeFort I ± mandible orthognathic surgery	Plastic surgeon, oral surgeon

Neonatal period

Surgical Repair

- Cleft Lip
  - In US - “the rule of tens” - 10 wks, 10 lbs, Hgb 10
  - Lip adhesion vs baby plates
- Cleft Palate
  - Varies from 6-18 months - most around 10 mo
  - Early repair may lead to midface retrusion
  - Early repair improves speech

Toddler - years

- Priority: Speech
  - "Cleft errors of speech" in 30%
  - primary defects - due to VPI (hypernasality)
  - consonants are most difficult sounds (plosives)
  - secondary defects - due to attempted correction
  - glottic stops, nasal grimace
- Velopharyngeal insufficiency
  - diagnosed by fiberoptic laryngoscopy or nasal endoscopy
  - surgical repair after failed speech therapy - usually around age 4

The School Grade Year

- Three primary issues
- **Orthodontics**
  - poor occlusion
  - congenitally absent teeth
- **alveolar bone grafting**
  - fills alveolar defect - around age 12
- **psychological growth**
  - considered standard of care

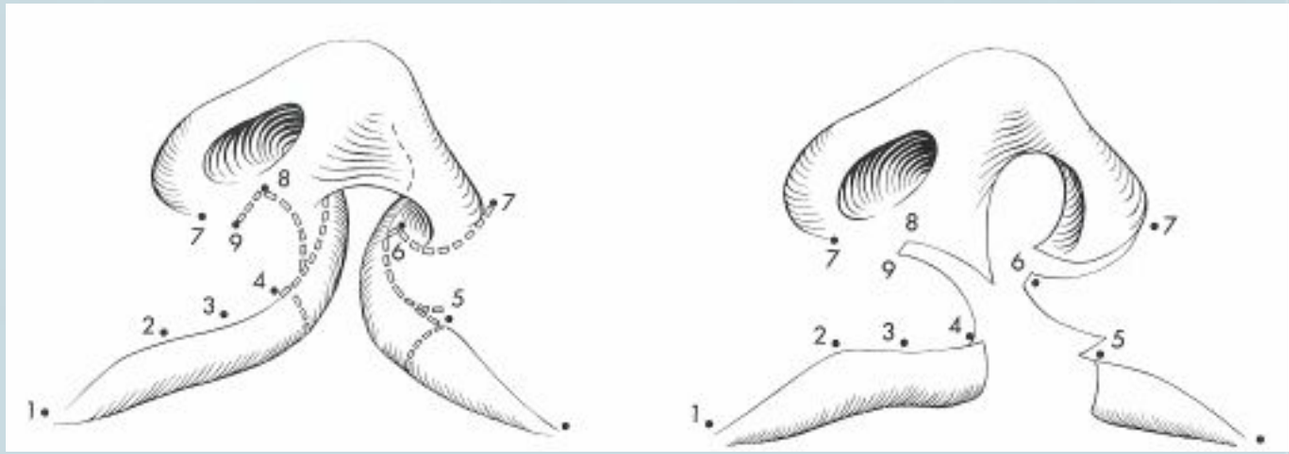
Teenage years

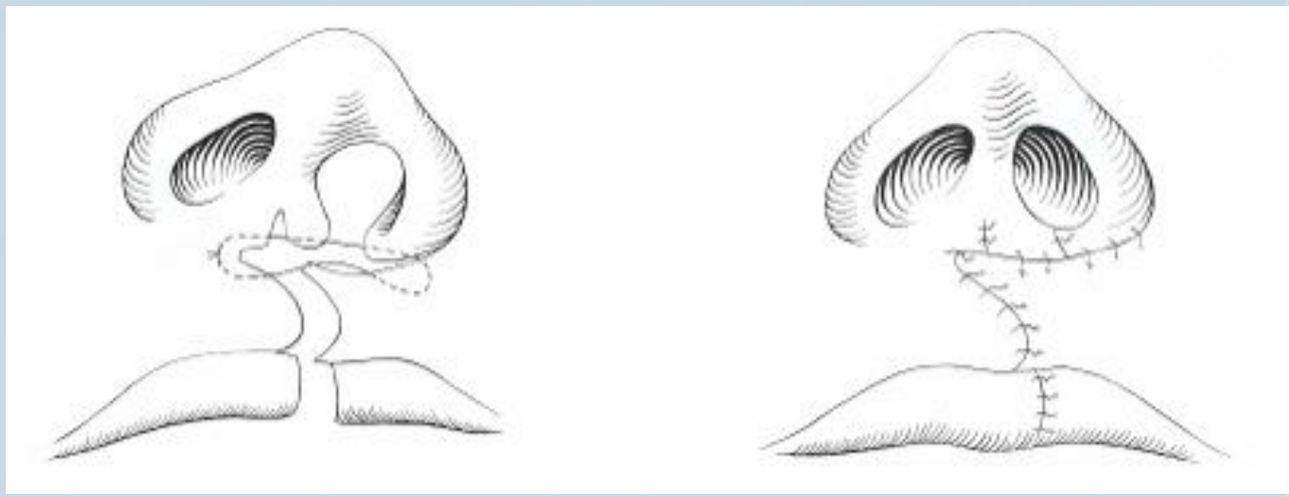
- •Midface retrusion

•surgical correction Le Fort I (mid facial advancement) osteotomies around age 18
•Rhinoplasty
•usually last procedure performed, around age 20

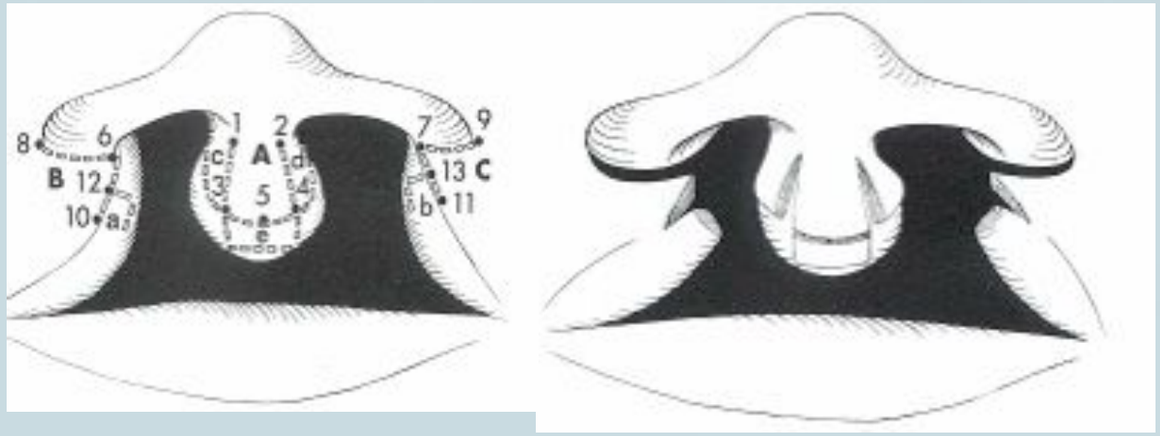
Surgical technique

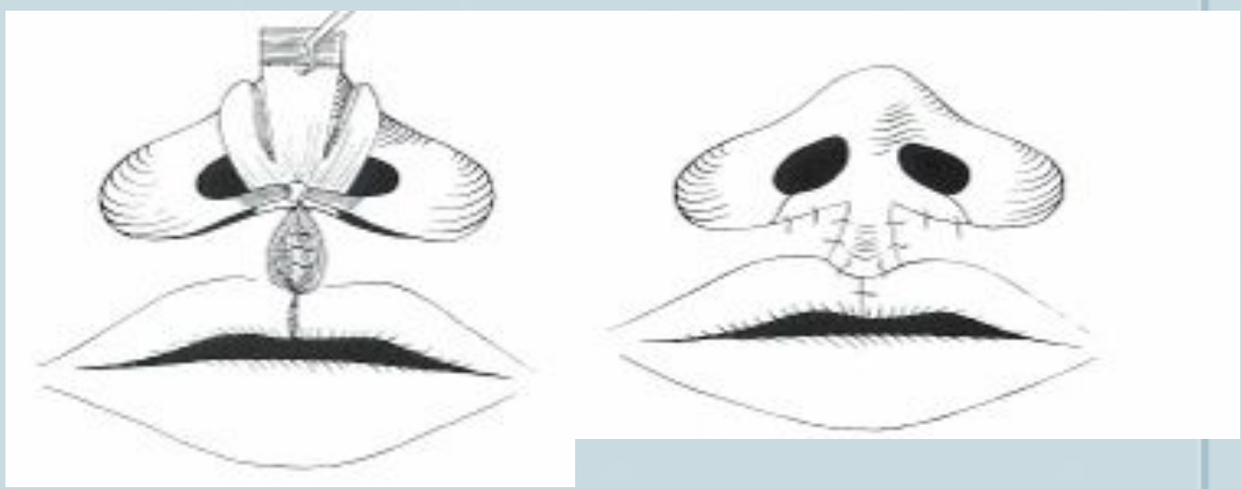
- Cleft Lip Repair
- unilateral
  - rotation-advancement flap developed by Millard
- complications
  - dehiscence
  - infection
  - thin white roll
  - excess tension





- Cleft Lip Repair
- bilateral
  - bilateral rotation advancement / straight line incision with attachment to premaxilla mucosa
- complications
  - dehiscence
  - thin white roll





Controversies- timing of repair

- Early repair
- Advantage: improved speech
  - Rohrich, et. al; retrospective study. The earlier the repair, the better speech.
- Disadvantage: worsening midface retrusion
  - Rohrich, et. al; people with unrepaired palates have less midface retrusion

Conclusion and future direction

- Multi disciplinary approach
- Not merely a “surgical problems”





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