

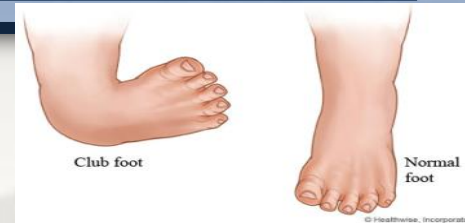


Introduction

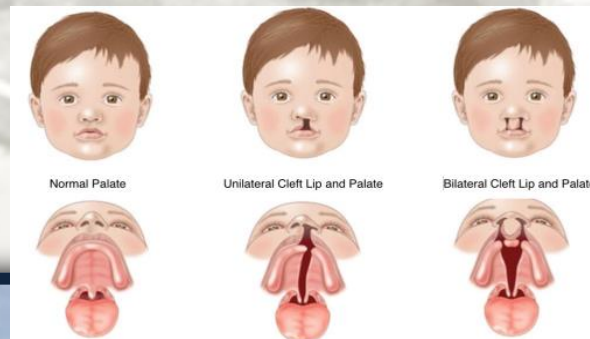


- 🛡️ Hippocrates (400 BC) and Galen (150 AD) mentioned cleft lip, but not cleft palate in their writings
- 🛡️ Cleft palate – Fanco (1556)
- 🛡️ Repair of cleft lip – as early as 255-206 BC in China
- 🛡️ The first successful closure of a soft palate defect was reported in 1764 by LeMonnier, a French dentist.

Introduction



- ❖ Facial clefting is the second most common congenital deformity (after clubfoot).
- ❖ Among the 15 types of orofacial clefting, cleft lip and palate is the most common one.
- ❖ 1 in 700 live births (1/1000 in the US)
- ❖ Associated problems include otological disease, speech and language problems, dental deformities, and psychosocial issues
- ❖ Best managed with a multidisciplinary approach (medical and surgical)



Incidence



- 🛡️ *A child is born with a cleft somewhere in the world every 2 minutes according to a WHO study published in 2001*
- 🛡️ The prevalence rate of cleft lip and palate in Jordan on 2001 was 1.39 per 1000 live births
 - Cleft palate craniofac. J 2004



🛡️ Ethnic groups(CL+/-P)

- Highest rate
 - Native American and Asians (2/1000 live births)
- Intermediate rate
 - European descendants (1/1000 live births)
- Lowest rate
 - African populations (1/2500 live births)

- 🛡️ • No difference between ethnic groups for cleft palate only (1/2000 live birth)

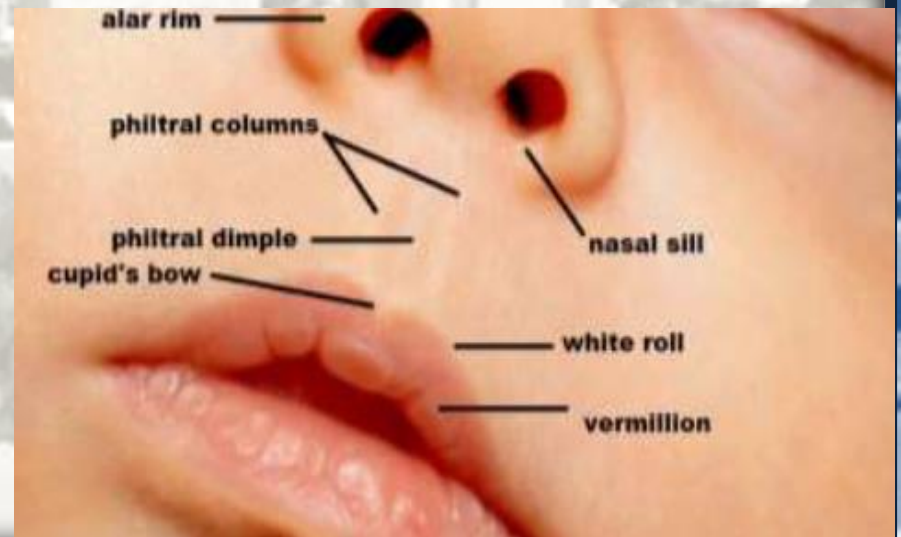
🛡️ Gender

- 2:1 – M:F ratio – cleft lip +/- palate
- 1:2 – M:F ratio – cleft palate only (late closure of palatine shelves)

Normal Anatomy



- Orbicularis oris
- Vermillion (wet/dry border)
- Cupid's bow
 - Along the upper vermilion cutaneous border (white roll), two midline elevations form the bow
- Philtrum
 - Philtral columns and dimple

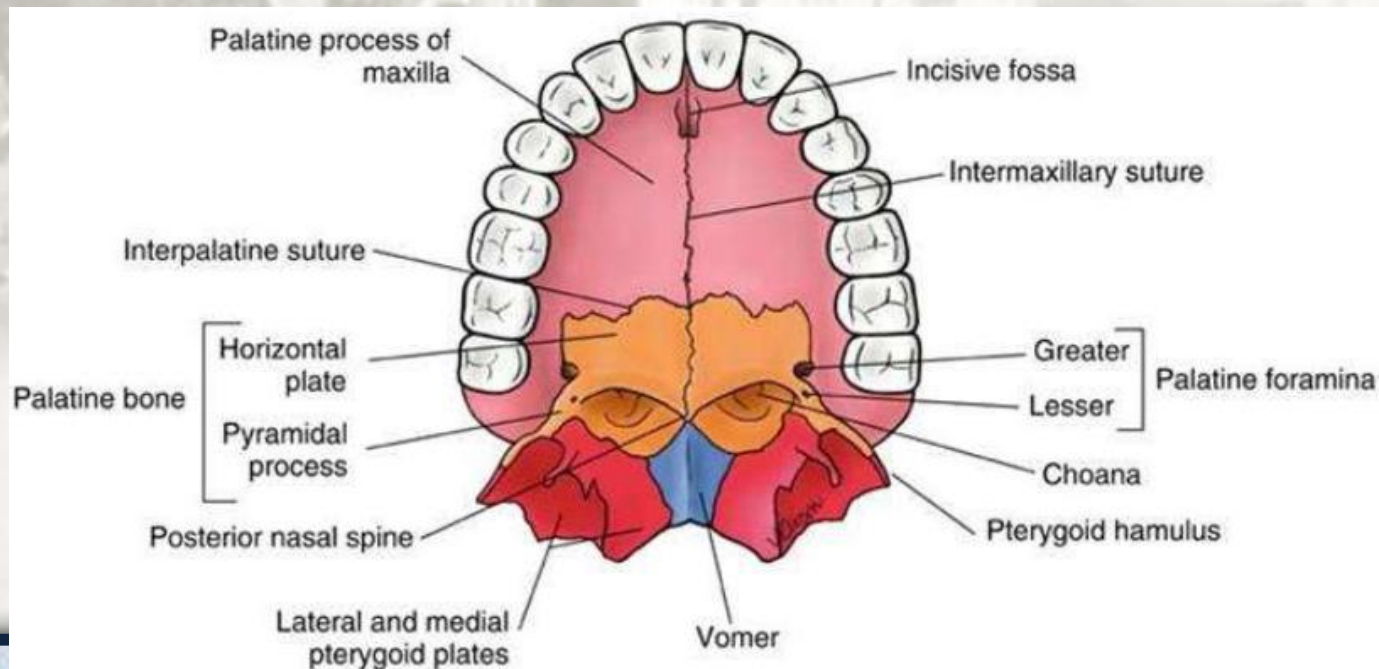


Normal Anatomy



Hard Palate

- Palatine processes of the maxilla
- Horizontal plates of the palatine bone
- Incisive foramen, greater and lesser palatine foramen

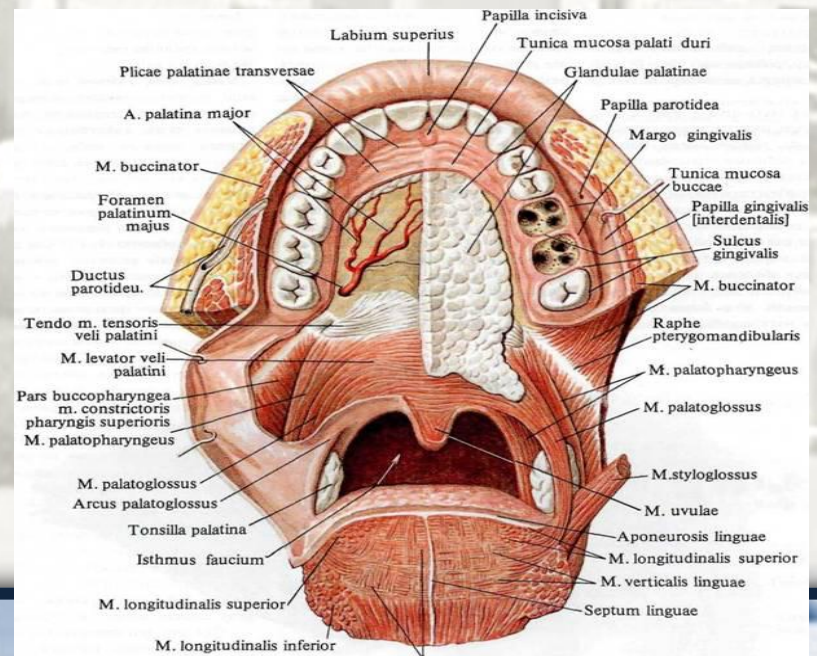
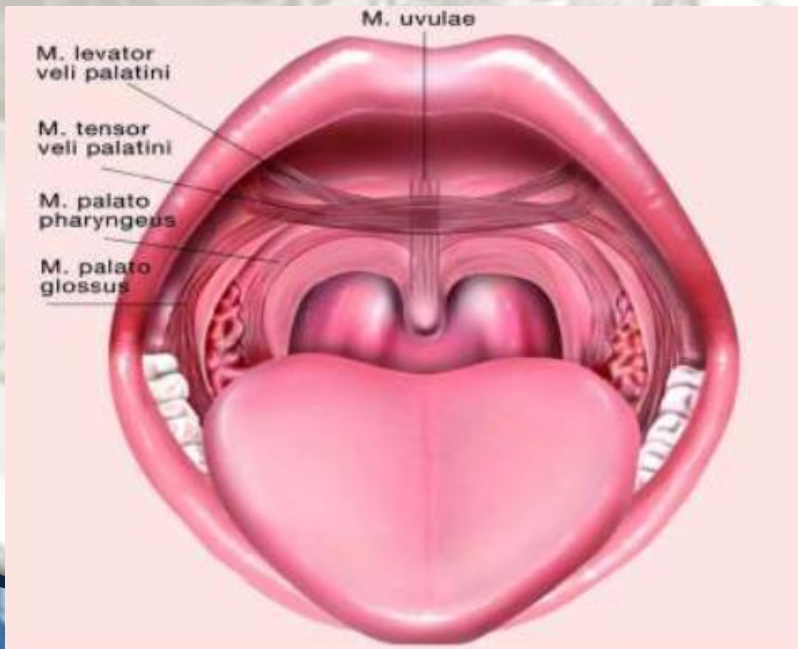


Normal Anatomy



Soft palate

- Palatine aponeurosis – tendon of tensor veli palatini
- Muscular portion consists of the tensor veli palatini (CN V), levator veli palatini, palatoglossus, palatopharyngeus, and musculus uvulae (CN X)



Normal Anatomy



🛡️ Vasculature

- Greater palatine, lesser palatine, and ascending palatine artery

🛡️ Nerve supply

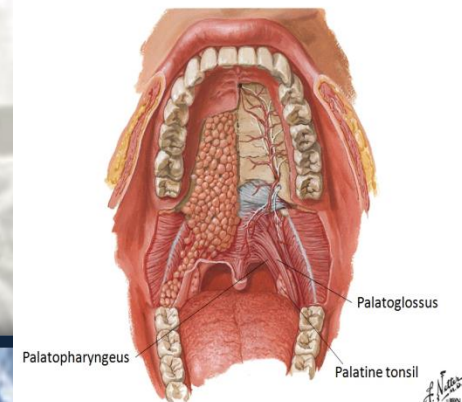
– Hard palate

- Greater palatine nerve supplies the gingivae, mucous membranes, and glands of most of the hard palate
- Nasopalatine nerve supplies the mucosa of the anterior portion of the hard palate

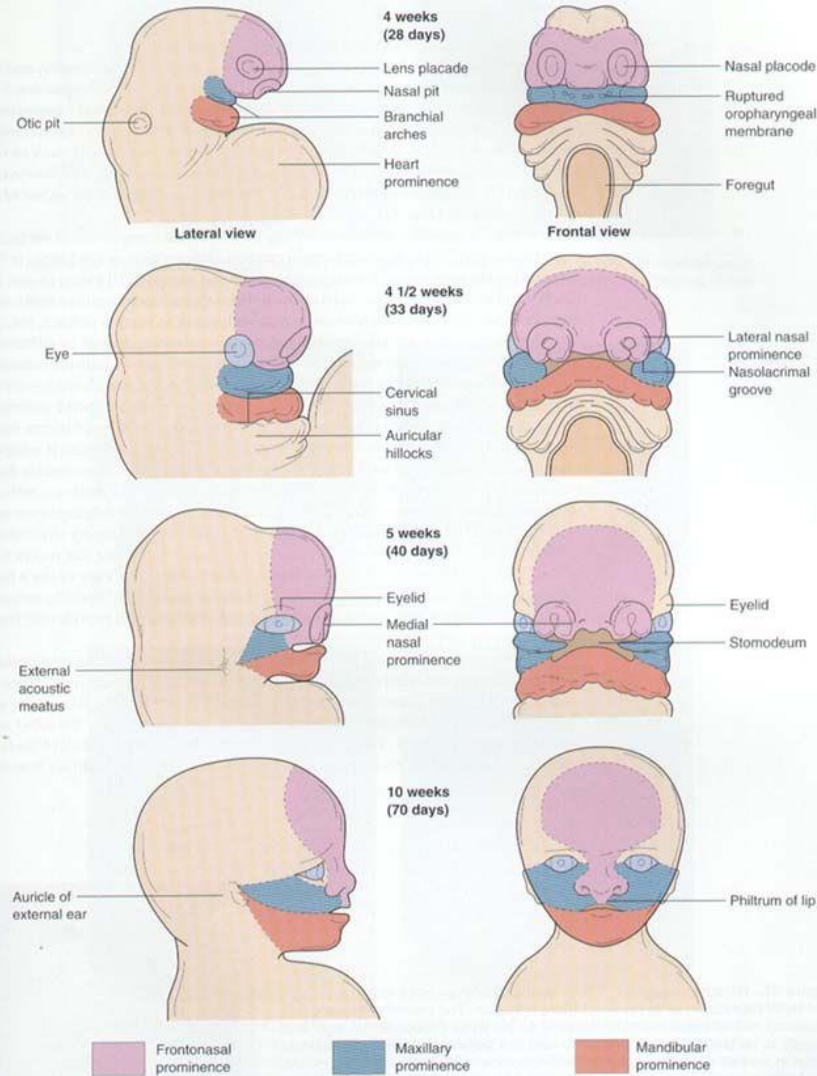
– Soft palate

- Sensory – lesser palatine nerves
- Motor – tensor veli palatini is supplied by CN V and the other muscles are supplied by CN X

Roof of Mouth - Hard and Soft Palates
Anterior View



EMBRYOLOGY

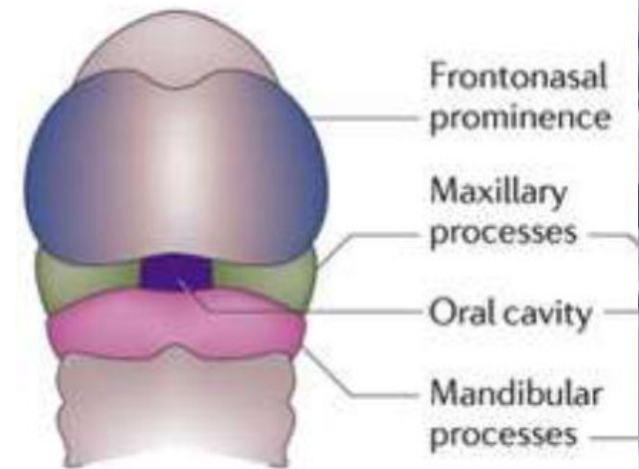


Embryologic development of the face. Reprinted with permission (138).

EMBRYOLOGY



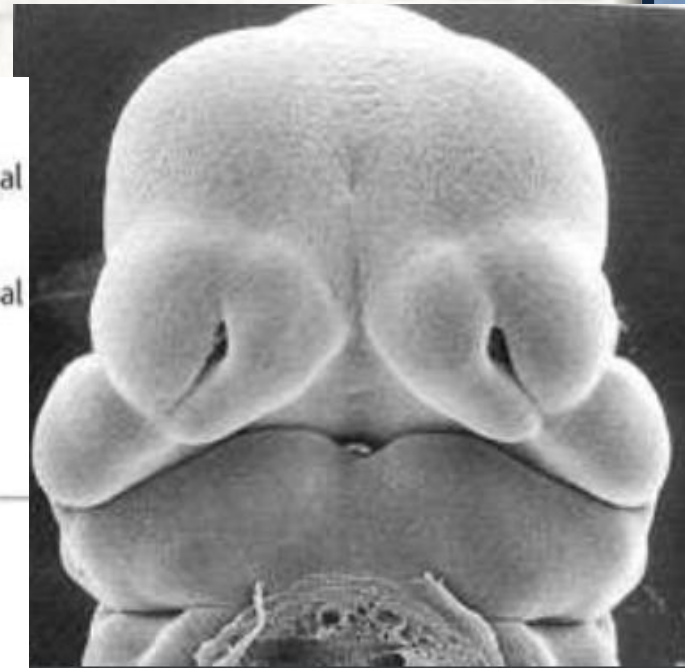
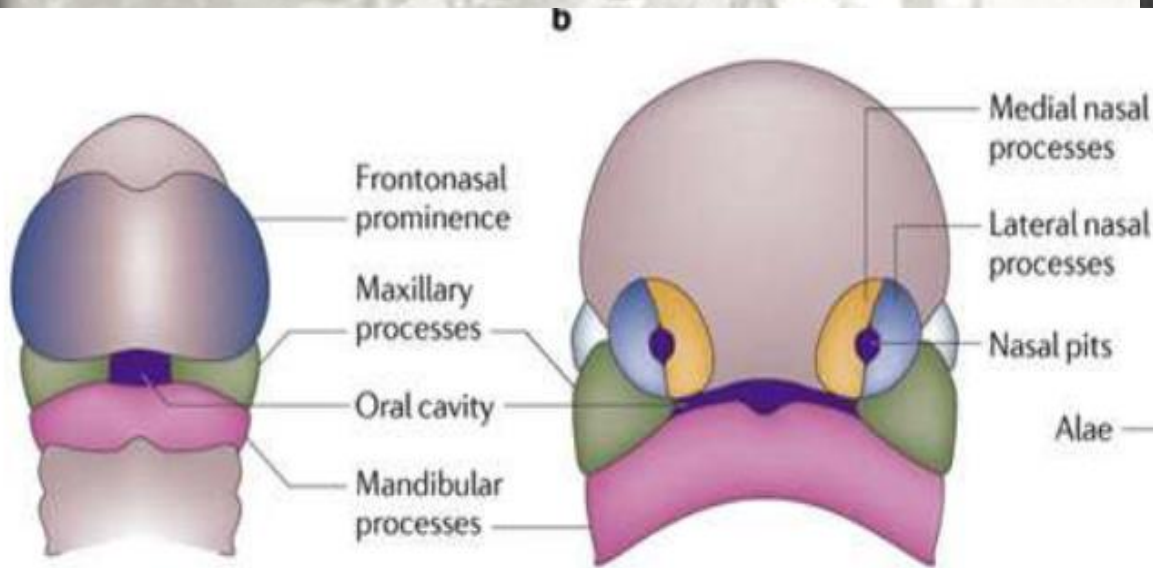
- 🛡️ Development of the lip and palate begins around the 4th week of embryological development
- 🛡️ Completed by the end of the 12th week
- 🛡️ By the end of the 4th week
 - 5 facial prominences have formed
 - frontonasal process
 - paired maxillary processes
 - paired mandibular processes



EMBRYOLOGY



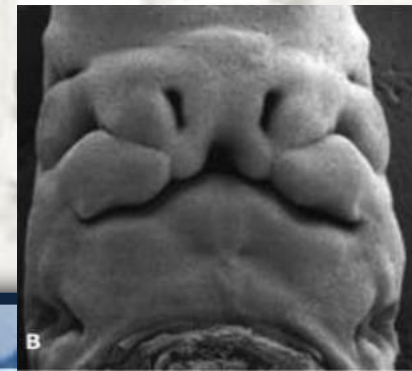
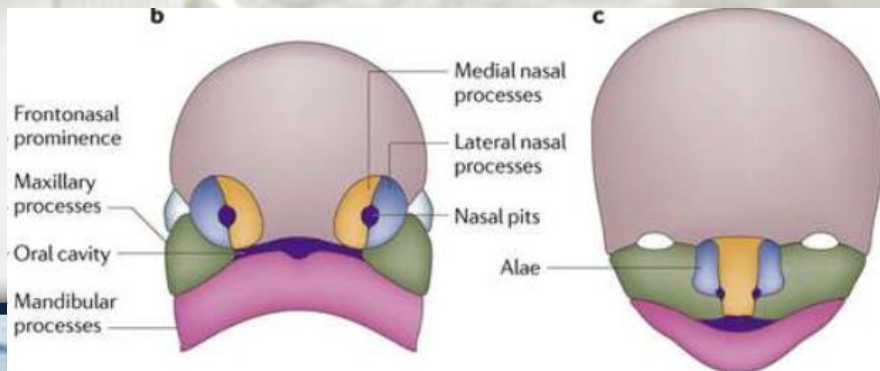
- During the 5th week
- Nasal placodes invaginate to form the nasal pits
- Lateral and medial nasal prominences



EMBRYOLOGY



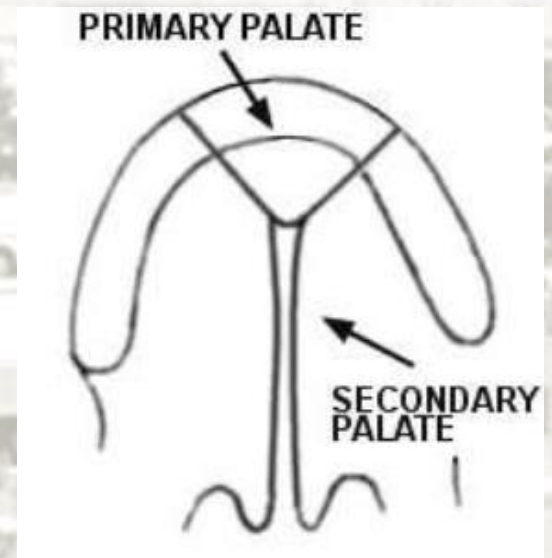
- By the end of the 6th week
- Paired maxillary processes have grown medially and pushed the paired medial nasal prominences together
- Fusion of the paired medial nasal prominences form:
 - Philtrum
 - Middle upper lip
 - Nasal tip Columella
- Fusion of the paired maxillary prominences with the paired medial nasal prominences forms the complete upper lip (maxillary prominences form lateral lip)
- The lateral nasal prominences form the bilateral nasal ala



Palate Formation



- Palate formation begins at the end of the 5th week of development and is completed by the 12th week
- The completed palate is formed by the primary palate and the secondary palate which are separated by the incisive foramen



Primary Palate



- Medial nasal prominences fuse to form **intermaxillary segment** → primary palate
- Consists of maxillary alveolar arch with 4 incisors and the hard palate anterior to the incisive foramen
- Primary palate forms before the secondary palate begins formation

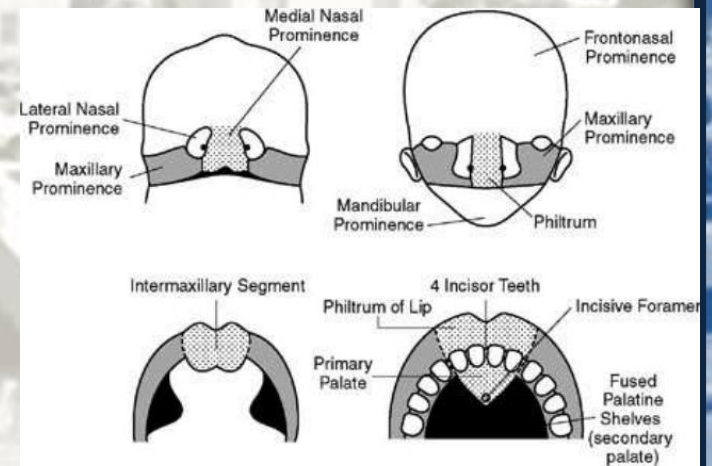
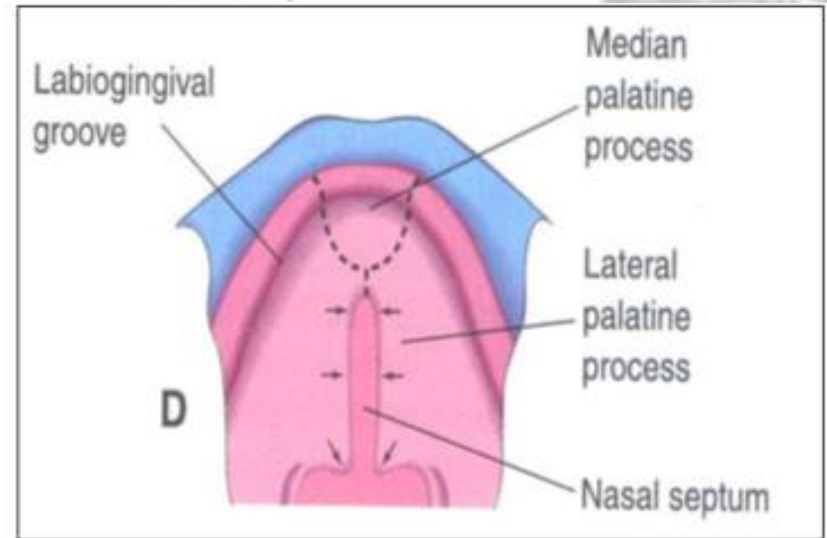
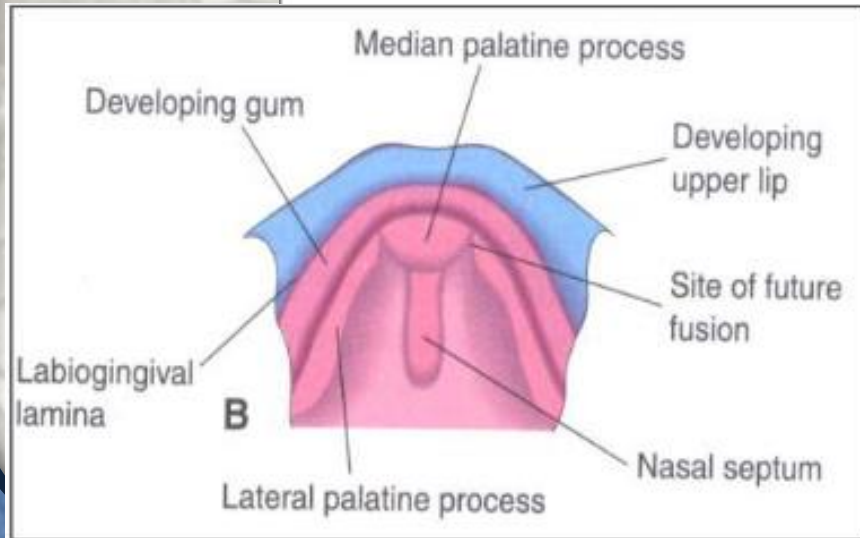
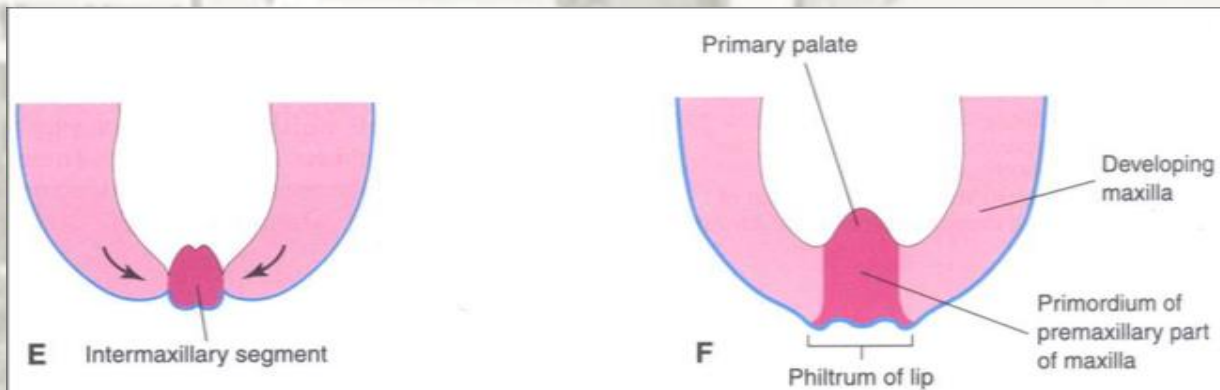


Figure III-6-9. Palate and Face Development

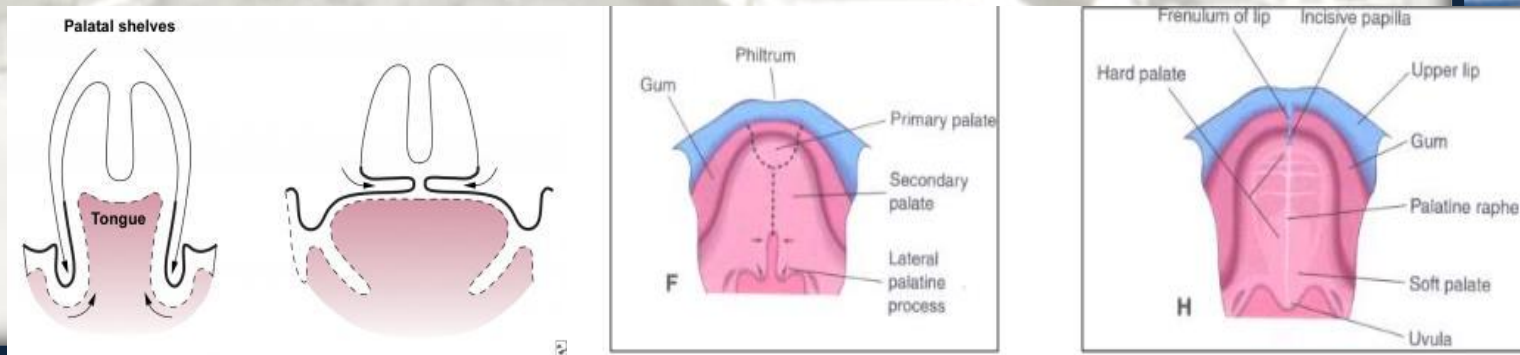
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Secondary Palate



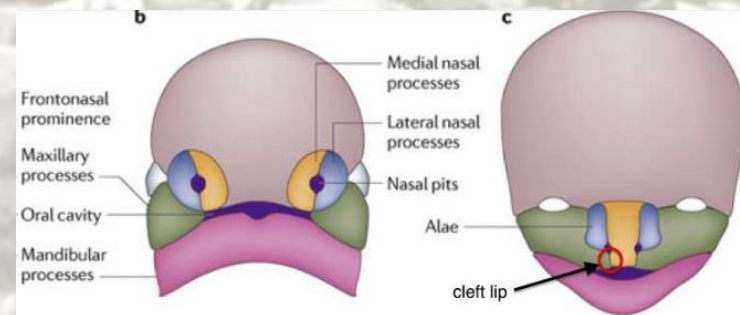
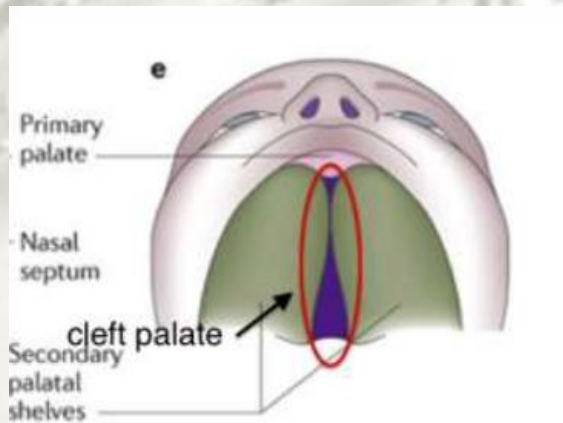
- During the 6th week
 - Shelf-like outgrowths from the bilateral maxillary processes, grow vertically down on both sides of the tongue
- During the 7th week
 - The tongue moves inferiorly and the palatal shelves migrate to a horizontal position above the tongue
- Palatal fusion occurs in an anterior to posterior direction and completes with uvular fusion (1 week later in females)



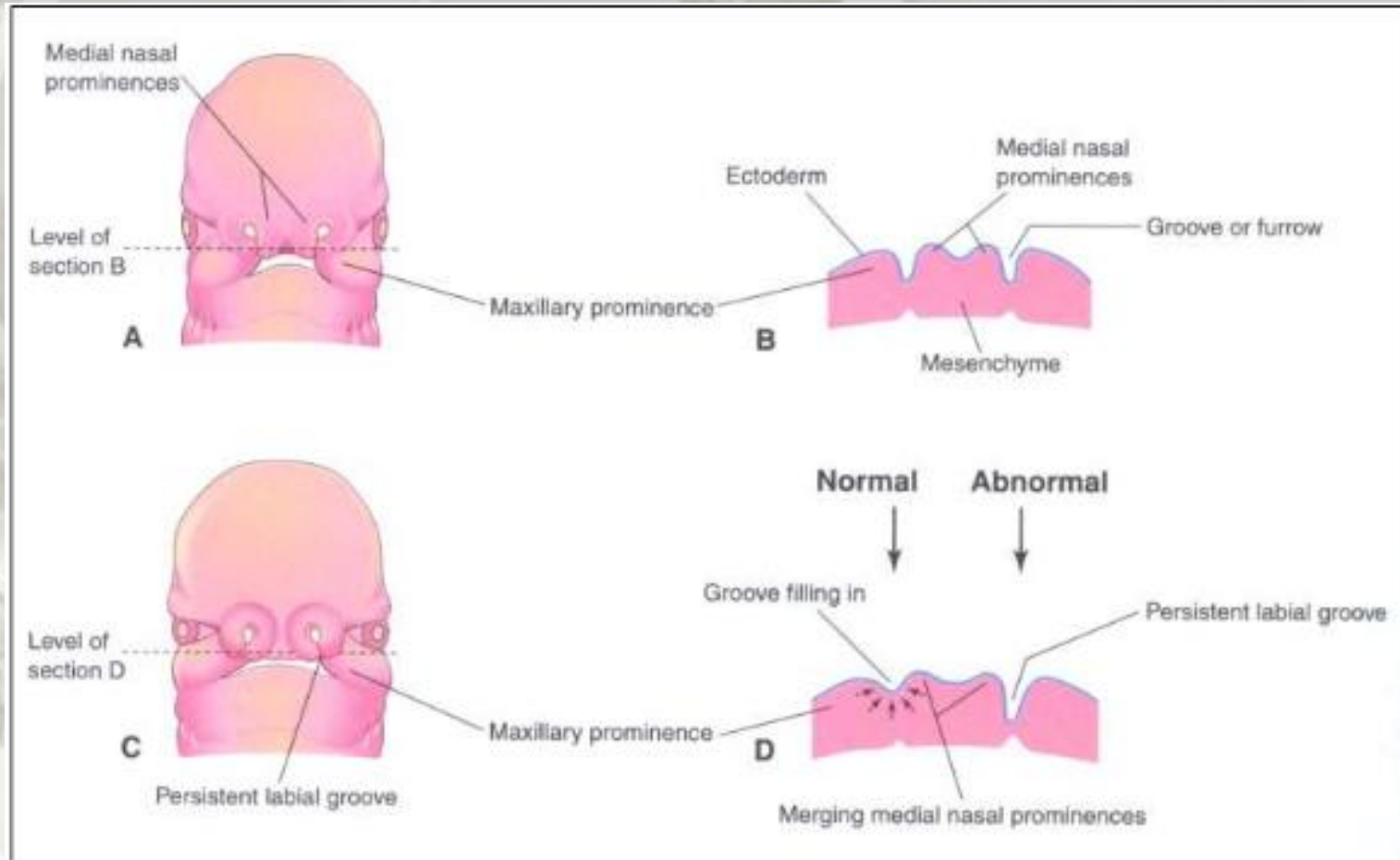
Formation of clefts



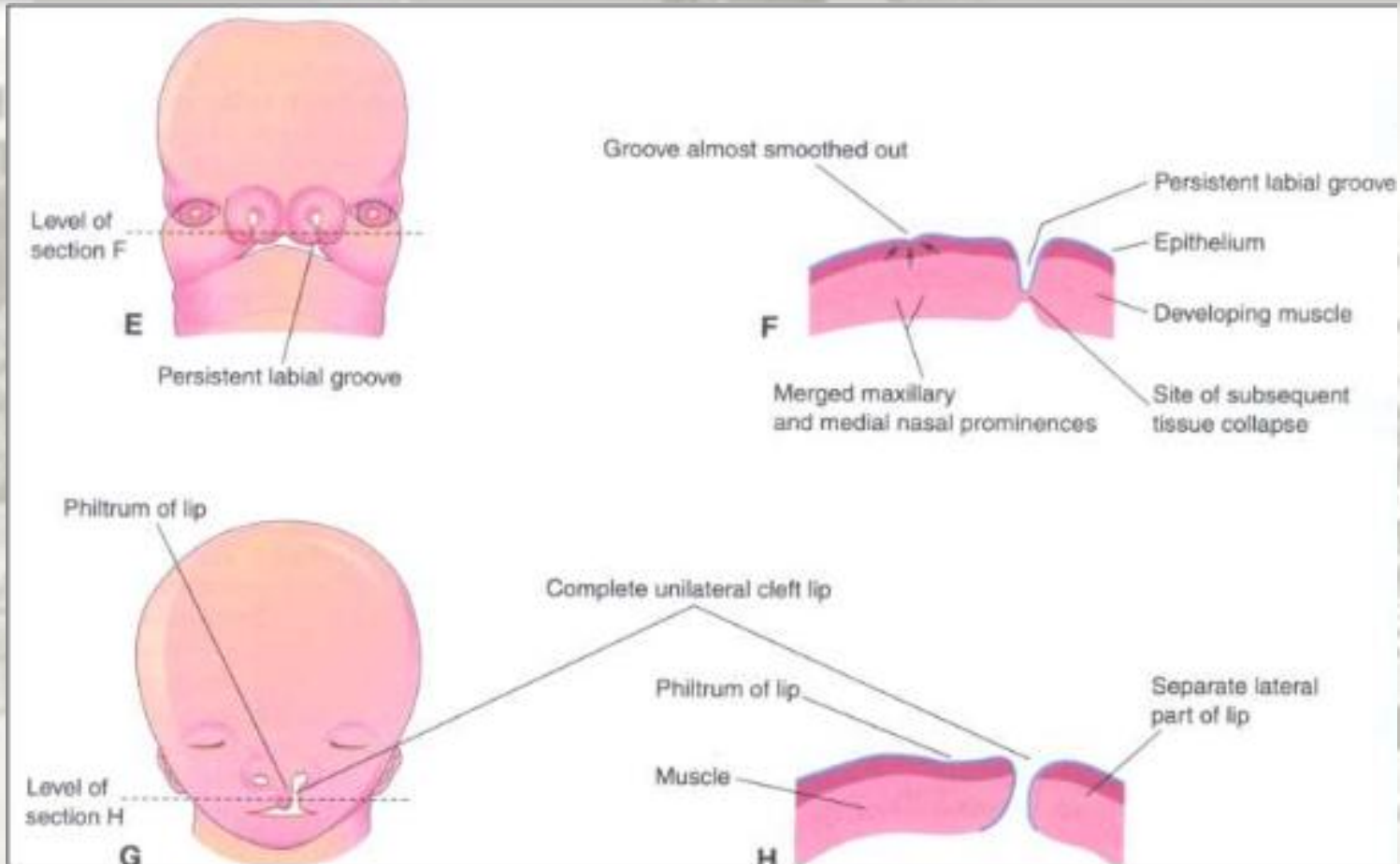
- ❖ Cleft lip – failure of proliferation of mesodermal cells in midline
- ❖ Failure of fusion of maxillary and medial nasal processes – anterior to incisive foramen
- ❖ Failure of fusion of palatine shelves – posterior to incisive foramen



Formation of clefts



Formation of clefts



ETIOLOGY



ETIOLOGY



Ancient Folklore explanations

- Aztecs – eclipses occurred because a bite had been taken out of the moon
- Prevented with an obsidian knife above the pregnant abdomen
- Modern Mexico prevented with keys and safety pins





🛡️ Early Chinese

- Eating rabbit “hare lip”
- Bad karma or wrong doings

🛡️ Philippines

- Force to the fetal face

🛡️ Familial or “In the blood”





Etiology

Multifactorial

Familial



- 🛡️ 2 unaffected parents with 1 child affected
 - Risk for future children:
 - 4.4% for CL+/- palate
 - 2.5% for CP only
- 🛡️ 1 parent affected
 - Risk for future children
 - 3.2% for CL+/- palate
 - 6.8% for CP only
- 🛡️ 1 parent affected with 1 child affected
 - Risk for future children
 - 15.8% for CL+/- palate
 - 14.9% for CP only

Etiology



- Majority of orofacial clefts are nonsyndromic
 - 70% of CL +/- palate
 - 50% of CP
- Nonsyndromic clefts
 - multifactorial
 - Clusters in families but not mendelian
 - Palate development complex process with several proteins, growth factors, and transcription factors involved
 - IRF-6, TGF -B2, TGF-alpha
 - Any disturbance in the process can result in clefting



Syndromic clefts:

- Associated with over 300 syndromes
 - Van der Woude syndrome – the most common
 - Autosomal dominant
 - Lower lip pits



Etiology



Environmental factors








- Maternal smoking or tobacco exposure
- Viral infections
- Poor nutrition
- Certain Medicinal drugs
- Teratogens like:
 - Rubella virus, Cortisone/ steroids, Mercaptopurine, Methotrexate, Valium, Dilantin



» Peter Mosby et al. Cleft Lip and Palate. Lancet 2009; 374: 1773–85

PREDISPOSING FACTORS



-  Advanced maternal age
-  Diabetes
-  Toxemia
-  Reduced blood supply
-  Folic acid deficiency
-  Racial – mongoloids
-  Radiations

» Peter Mosby et al. Cleft Lip and Palate. Lancet 2009; 374: 1773–85

Classifications



Clefts

- Unilateral or Bilateral
- Complete or incomplete

Veau classification

- Class I – incomplete cleft involving only the soft palate
- Class II – cleft involving the hard and soft palate
- Class III – complete unilateral cleft involving the lip and palate
- Class IV – complete bilateral cleft

Modified versions

Unilateral cleft lip

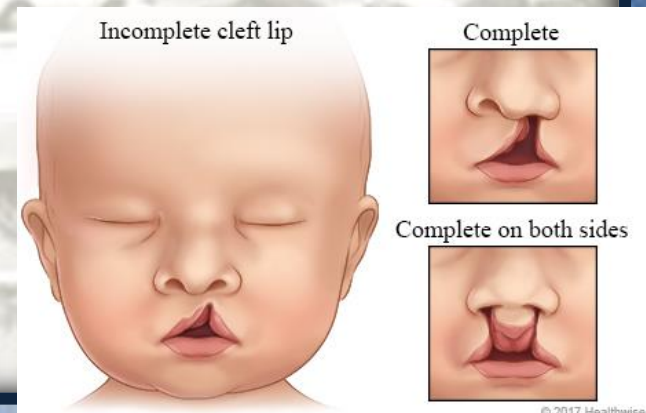


🛡️ Incomplete

- Muscle fibers of the orbicularis oris are often intact but hypoplastic
- Varying degrees of clefting

🛡️ Complete

- Orbicularis oris inserts at the columella medially and ala laterally on the cleft side
- Columella is displaced to the normal side
- Nasal ala on the side of the cleft is displaced laterally, inferiorly, and posteriorly
- Nasal tip is deflected towards the non cleft side
- Alveolus may or may not be involved



Bilateral Cleft Lip



- Orbicularis oris attaches at the lateral cleft margins bilaterally at the nasal ala
- Premaxilla protrusion
 - Symmetrical nasal deformities
 - Laterally displaced ala
 - widely flared
 - Extremely short columella



Unilateral cleft lip



Bilateral cleft lip

BILATERAL CLEFT LIP SPECTRUM

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BILATERAL INCOMPLETE CLEFT LIP



MICROFORM RIGHT & COMPLETE LEFT CLEFT LIP



INCOMPLETE RIGHT & COMPLETE LEFT CLEFT LIP & ALVEOLUS



BILATERAL COMPLETE CLEFT LIP & PALATE



Cleft Palate



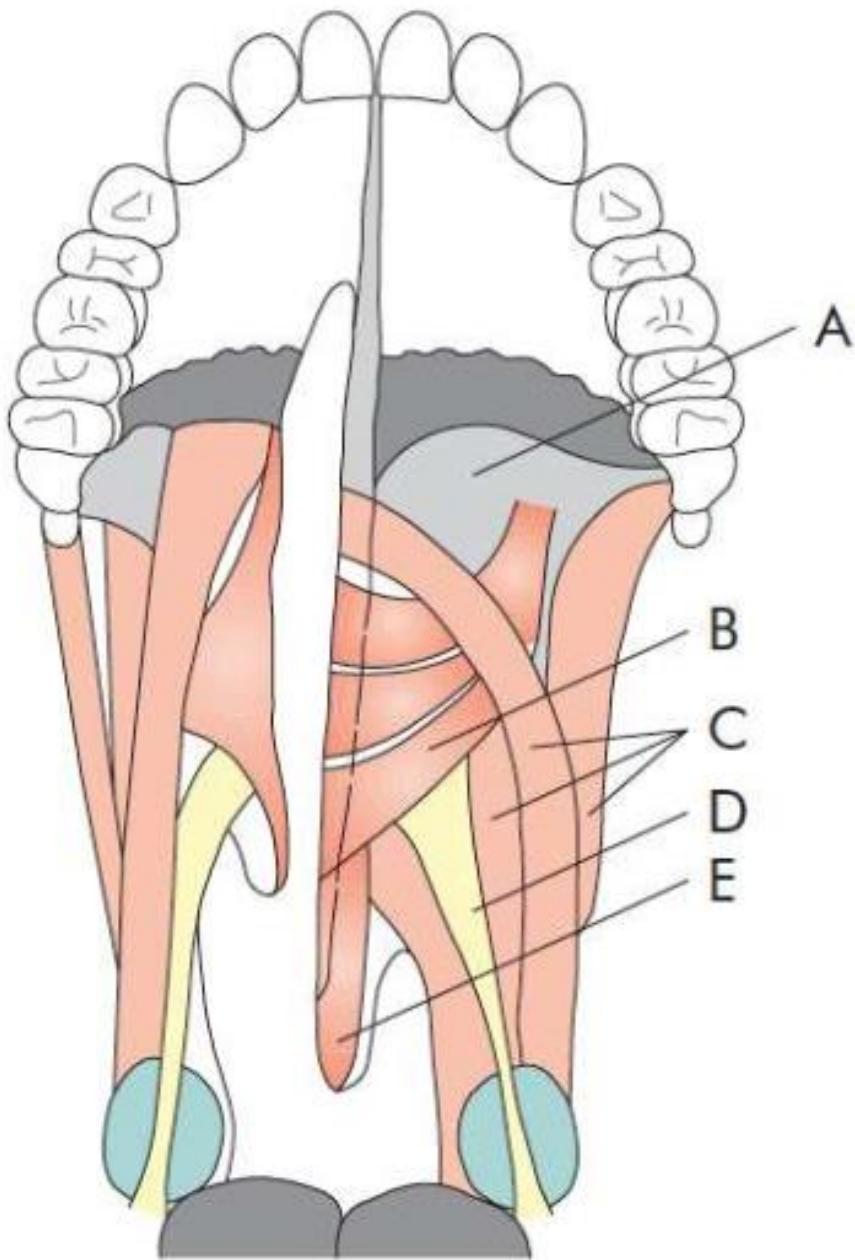
Primary palate
– Clefts anterior to the incisive foramen

Secondary palate

- Posterior to the incisive foramen
- Develops due to failure of the palatal shelves to fuse
- Abnormal insertion of the muscles
- Vomer attachment is variable



FIGURE 1- Different extents of isolated cleft palate: A) cleft lip, B) soft palate, C) incomplete hard palate and D) complete hard palate



Cleft Lip/Palate Management



🛡️ Multidisciplinary approach

🛡️ Cleft care team

- Plastic surgery
- Audiology
- Speech pathology
- Otolaryngology
- Orthodontist
- Oral maxillofacial surgery
- Psychologist
- Geneticist
- Pediatrician

Management



- 🛡️ Birth
 - Airway concerns
 - Feeding problems

- 🛡️ Otological disease

- 🛡️ Speech and language problems

- 🛡️ Surgical Repair

Airway Management



- 🛡️ Isolated cleft palate rarely results in airway compromise
- 🛡️ Airway issues are usually associated with coexisting structural abnormalities
- 🛡️ Pierre Robin sequence – most documented
 - Micrognathia, glossoptosis, and cleft palate
 - Associated with several syndromes (stickler, velocardiofacial syndrome, etc.)
 - Management – prone positioning (severe cases sometimes require tracheostomy)
 - Mandibular distraction



Feeding Difficulties



🛡️ Critical aspect in management

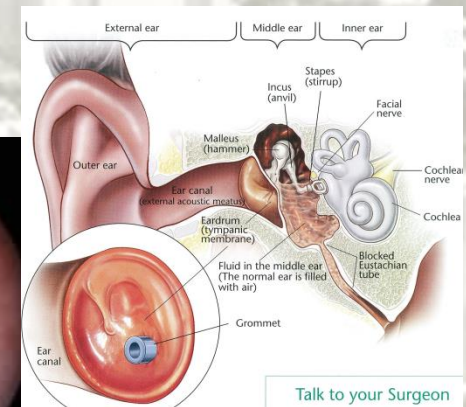
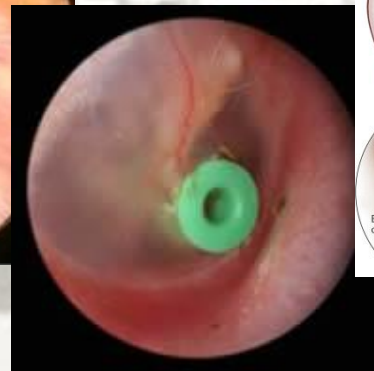
- Nutrition and feeding
- Cleft palate limits the ability to suck due to the common cavity
- Cleft lip alone
- Special bottles
- Premaxillary orthopedics



Otological Manifestations



- Abnormal insertion of tensor veli palatini
- Persistent OME has been estimated to be between 80-95% in children with cleft palate
 - The majority of them will need 1-2 sets of myringotomy tubes (grommet tubes)



Speech Development

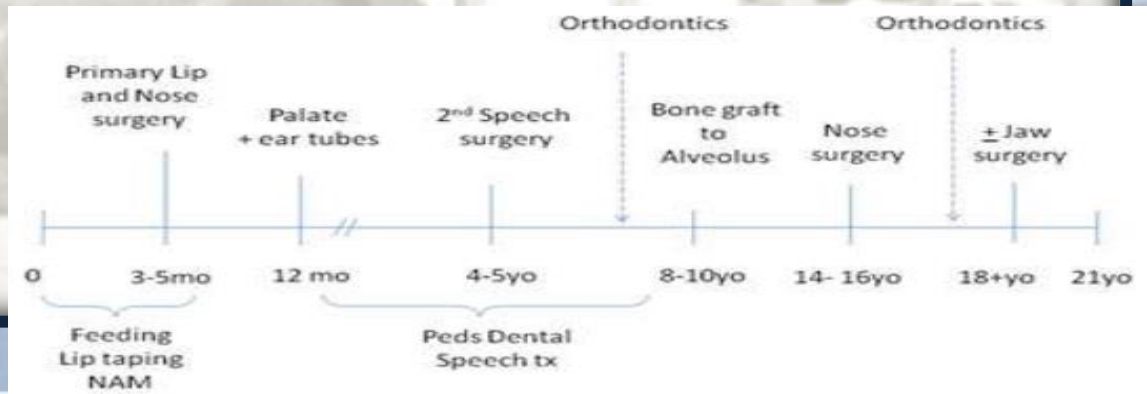


- Unrepaired cleft palate – speech abnormality
- Primary goal of palate repair is to restore function of the velopharyngeal valve
 - normal speech
- 10-20% will manifest VPI following surgical closure of the palate
- Hypernasality and articulation errors (glottal stops and pharyngeal fricatives)
- Speech pathologist – important role
- Managed surgically (pharyngoplasty or pharyngeal flap) or with dental prosthesis

Surgical Correction



- Age 1-3 months – Lip taping and nasoalveolar molding
- Age 3 months - Repair of cleft lip (and placement of ventilation tubes)
- Age 9-12 months - Repair of cleft palate
- Age 1-7 years - Orthodontic treatment
- Age 7-8 years - Alveolar bone graft
- 18 years old or skeletal maturity– Midface advancement and continued orthodontic treatment



Presurgical



- 🛡️ Wide cleft lip or premaxilla protrusion
 - Advantageous to narrow the cleft and mold the premaxilla before proceeding with surgery
- 🛡️ Taping
 - Effective in reducing the width of the cleft in a
- 🛡️ nonsurgical manner
 - Strip of hypoallergenic tape is placed with tension across the cleft and secured to the patient's cheek
 - Molds bony tissues by applying pressure to protruding portions of the maxilla
 - Must be worn 24 hours per day

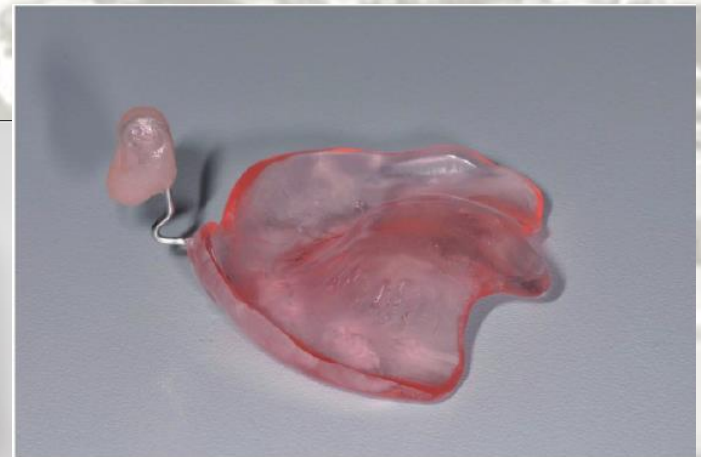
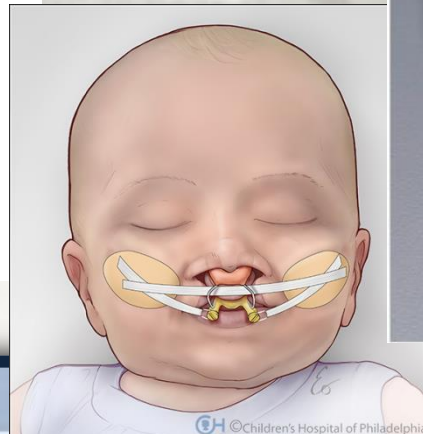
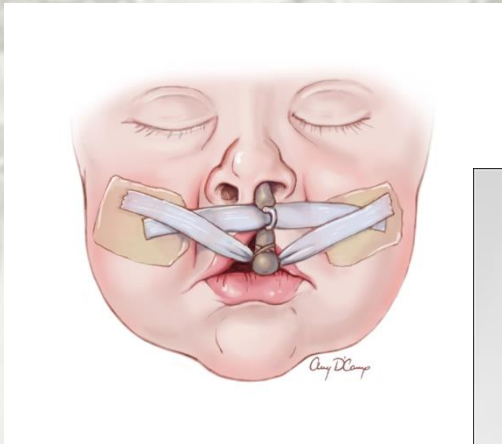


Presurgical



🛡️ Nasoalveolar molding devices

- Custom made devices which utilize wiring and nasal stenting to mold the nasal cartilage, premaxilla, and alveolar ridge
- Nasal stenting can be elongated and adjusted to lengthen the columella and mold the nasal cartilage
- Takes advantage of the malleability of nasal cartilage



Presurgical



🛡️ Lip adhesion

- Surgically convert a complete cleft to an incomplete cleft
- Performed at 2-4 weeks with definitive repair at 5-6 months

🛡️ Indications

- Wide unilateral cleft where conventional repair might produce excessive tension
- Bilateral cleft – premaxilla protrusion

🛡️ Disadvantages

- scar tissue



Cleft Lip Repair



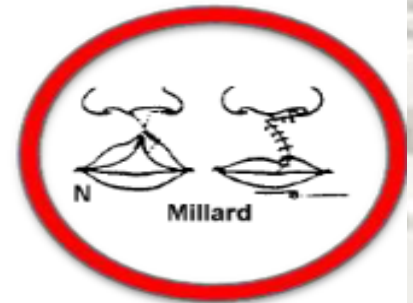
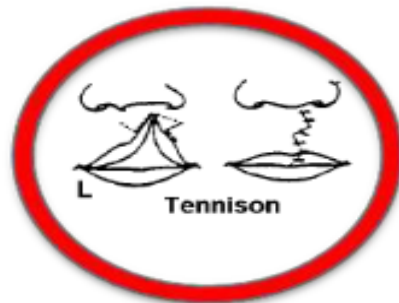
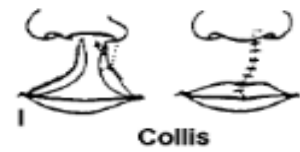
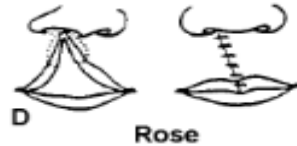
- ♣ Typically performed at 3 months of age
 - “Rule of Tens”
 - 10 weeks old, 10 lbs, and hemoglobin of 10
 - Wide clefts or clefts with premaxilla protrusion that require lip adhesions will have definitive lip repair at 5-6 months of age

Cleft Lip Repair



- ♣ Milliard rotation-advancement technique
 - Introduced in 1957
 - Most widely used procedure for unilateral cleft lip repair

Cleft Lip Repair



Bilateral Cleft Lip Repair



🛡️ Technically challenging

🛡️ Goals

- Symmetry
- Orbicularis oris closure
- Proper philtral size and shape
- Tubercle formation
- Positioning of alar cartilages to construct the nasal tip and columella

Cleft Palate Repair



Primary goals

- Separate the nasal cavity from the oral cavity
- Creation of velopharyngeal valve for swallowing and speech
- Preservation of midface growth

Timing

- Controversial
 - Speech outcomes improved with early closure
 - Midface growth maybe hindered by early closure (2 stage palate repair)
- Most repaired between 8-12 months of age to minimize speech abnormalities



Cleft Palate Repair



- Surgical techniques
 - Bardach two flap palatoplasty
 - Furlow double opposing z-plasty

Conclusion



- Common head and neck congenital malformations
- Multidisciplinary approach – Medical and surgical



• Teamwork is the key !

Thank you



