
الجامعة الأردنية
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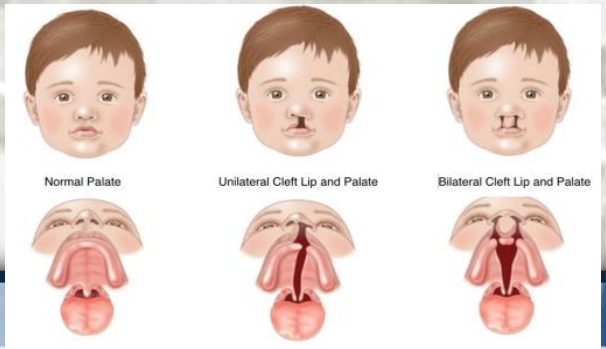
Introduction

- 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)

↓

1 week later

Normal Anatomy

We do our best to reconstruct these structures

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

musculature:-

- orbicularis oris
- levator labii superioris
- Nasal muscles

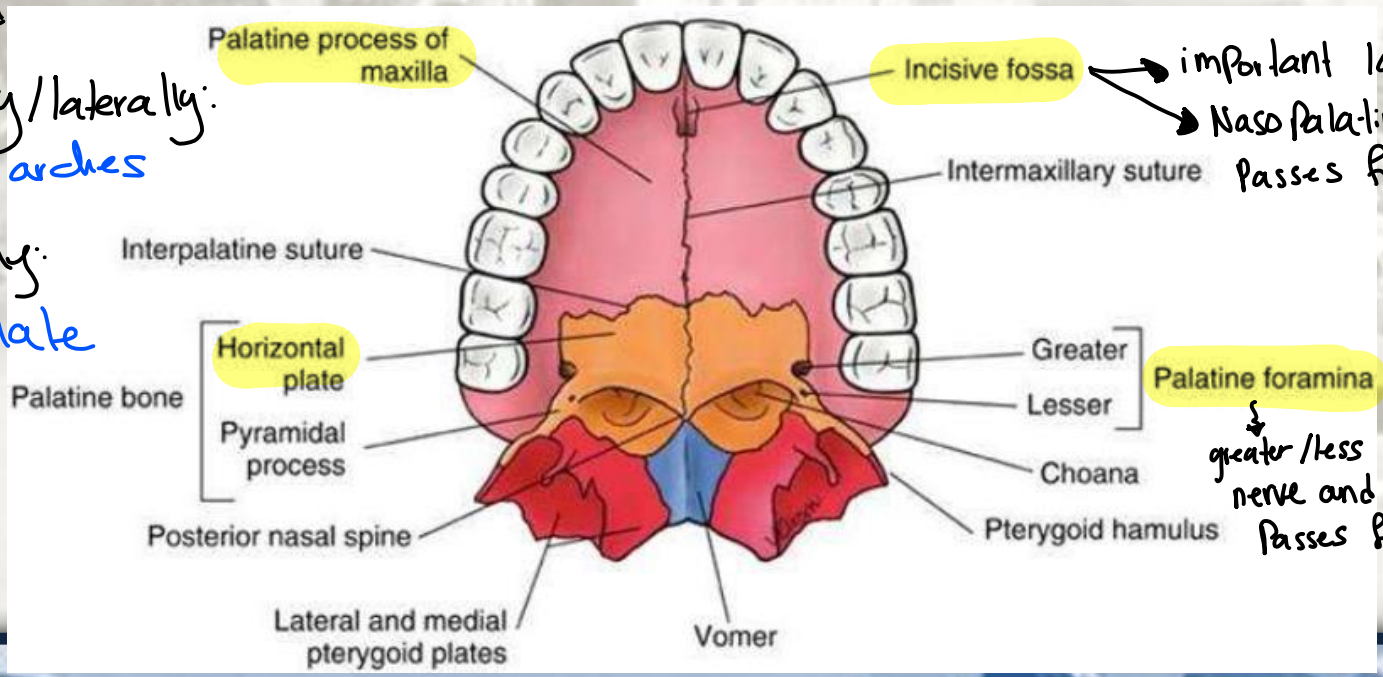
Normal Anatomy

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

Bounded

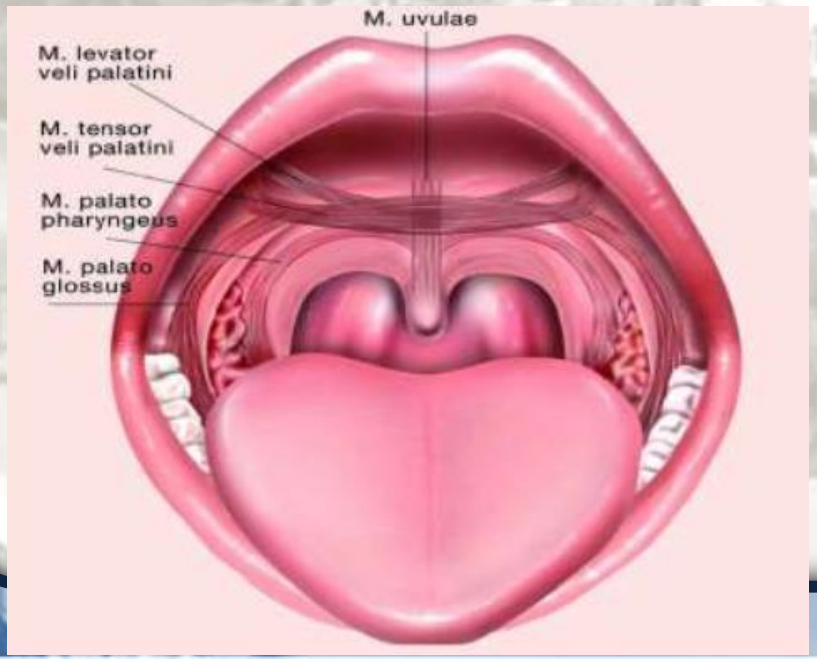
Anteriorly/laterally:
Alveolar arches

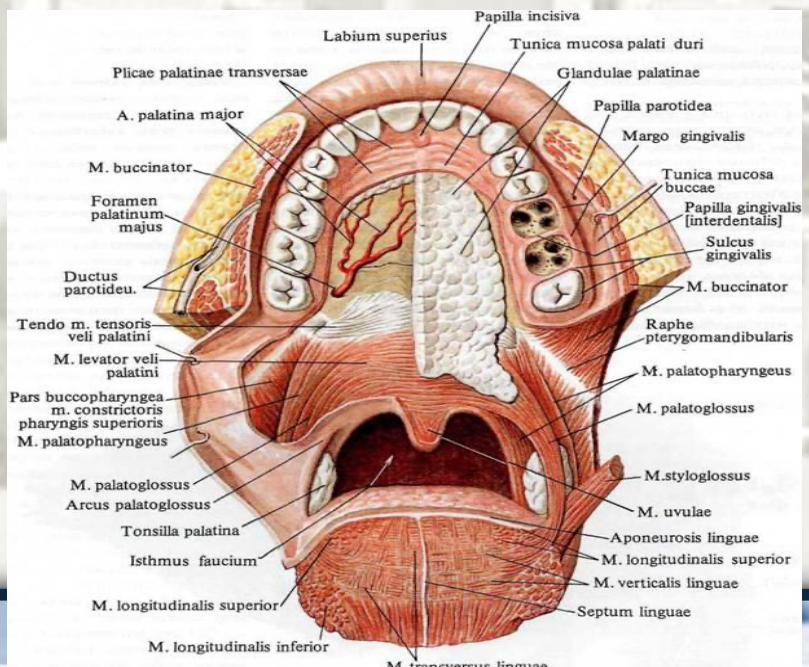
Posteriorly:
Soft Palate



Normal Anatomy

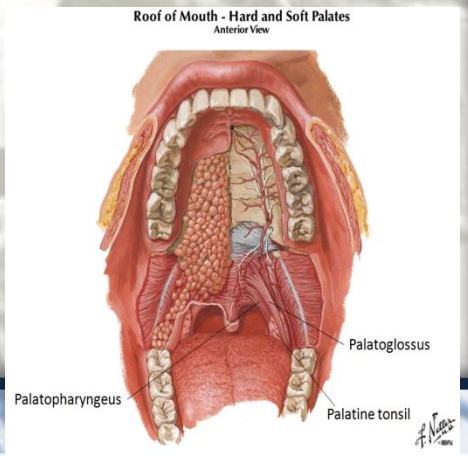
- Soft palate
  - Palatine aponeurosis – tendon of tensor veli palatini
    - connection between hard palate and muscle posteriorly
    - Area through which muscles are inserted into
  - Muscular portion consists of the tensor veli palatini (CN V), levator veli palatini, palatoglossus, palatopharyngeus, and musculus uvulae (CN X), Superior constrictor muscle



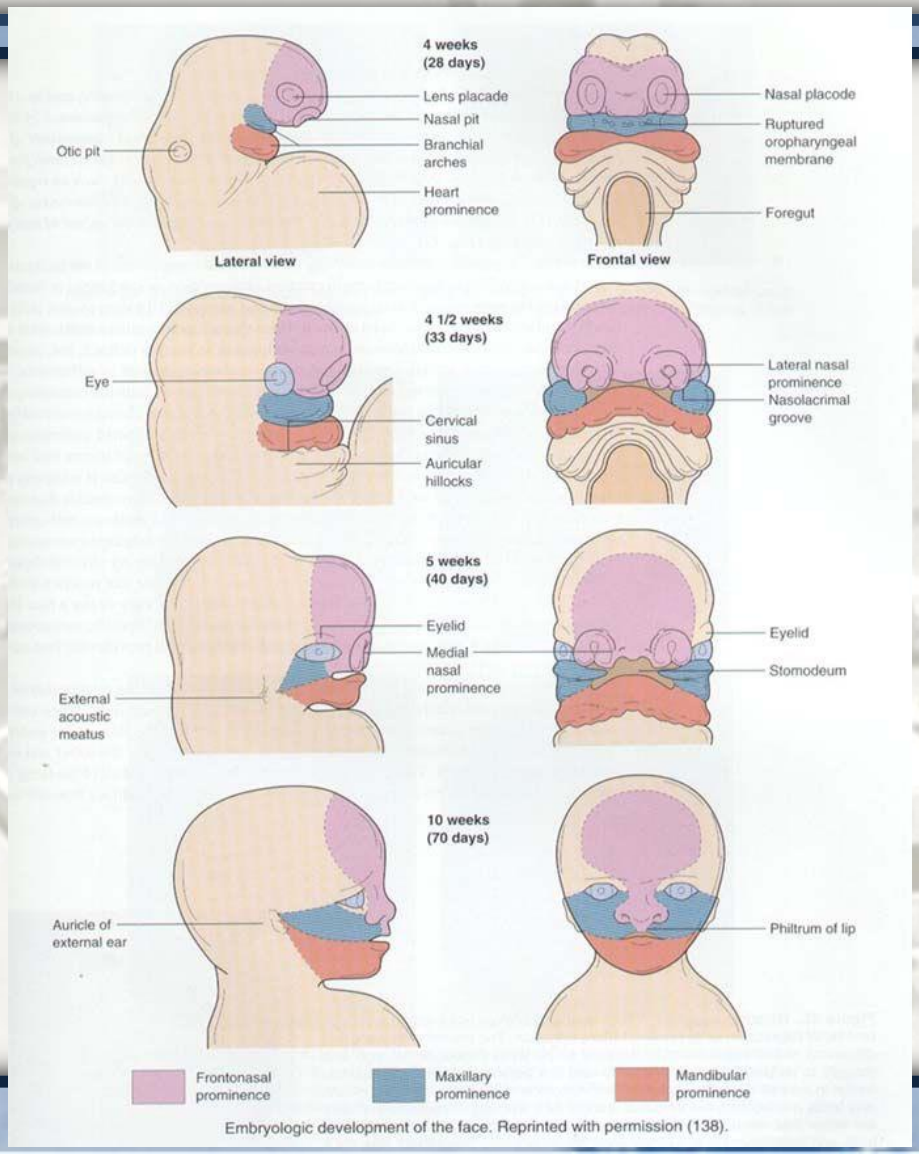


Normal Anatomy

- Vasculature → branch from maxillary artery → main blood supply
  - 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 (glossopharyngeal)
    - Motor – tensor veli palatini is supplied by CN V and the other muscles are supplied by CN X (vagus)



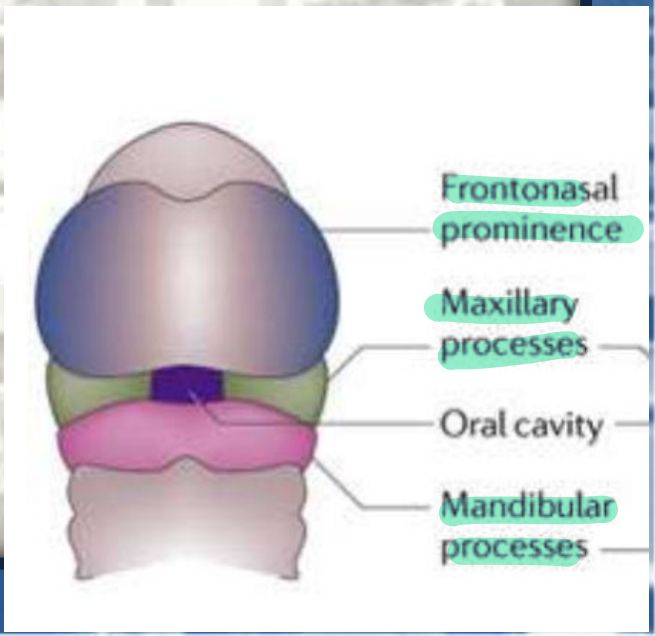
EMBRYOLOGY



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

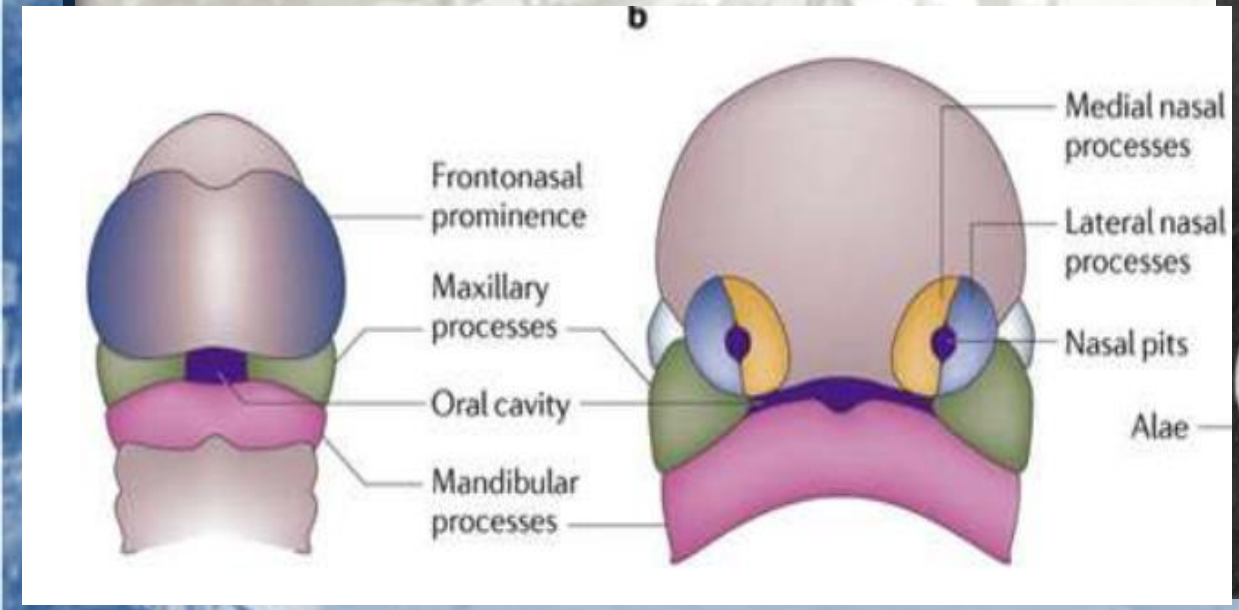
EMBRYOLOGY

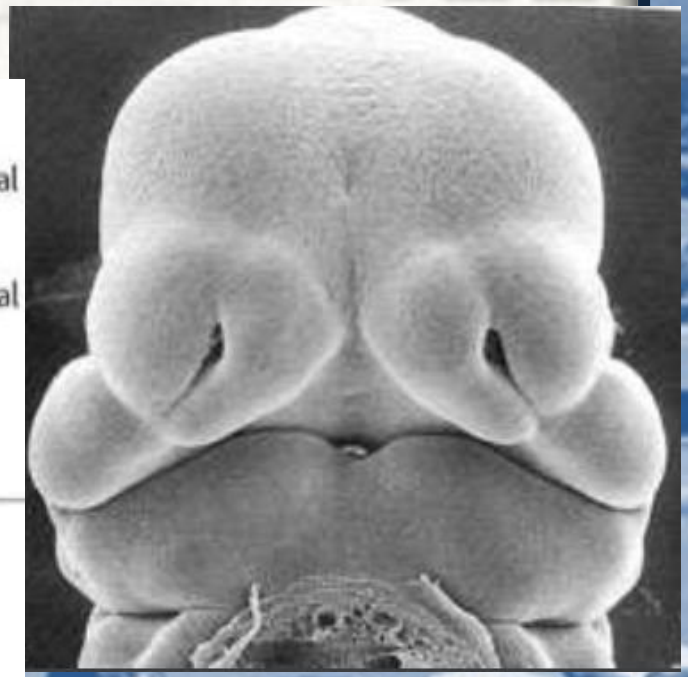
- Development of the lip and palate begins around the 4th week of embryological development
- Completed by the end of the 12th week
- By the end of the 4th week
  - 5 facial prominences have formed
    - frontonasal process
    - paired maxillary processes
    - paired mandibular processes



EMBRYOLOGY

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



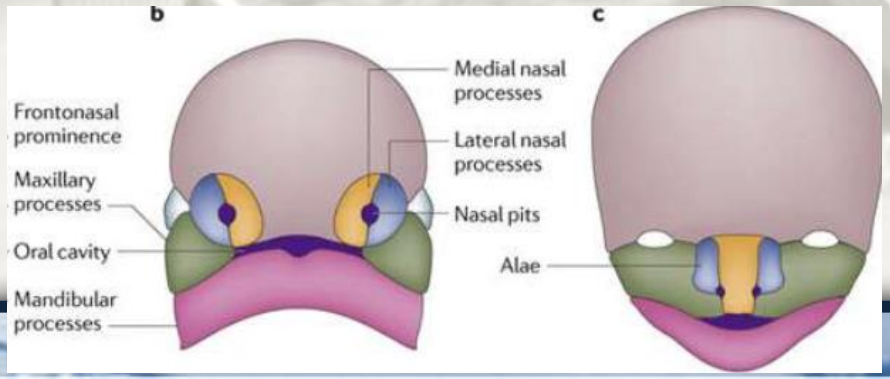


EMBRYOLOGY

- **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

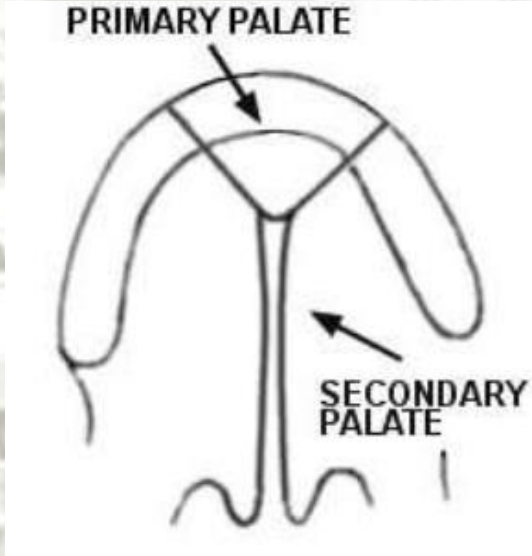
nasal: medial lip
maxillary: lateral lip

↳ lateral side of lower end of nose



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

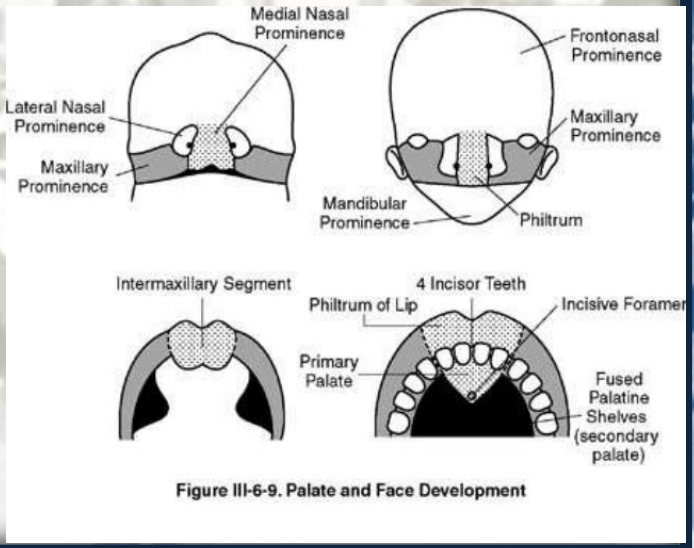
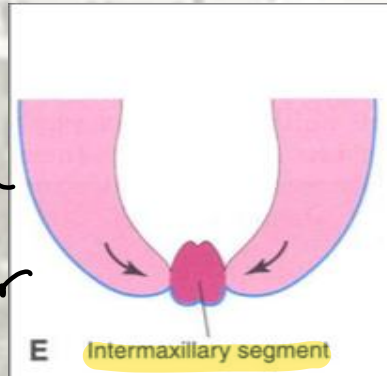


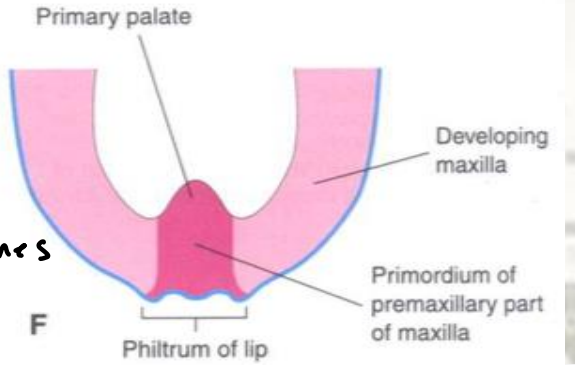
Figure III-6-9. Palate and Face Development

EMBRYOLOGY

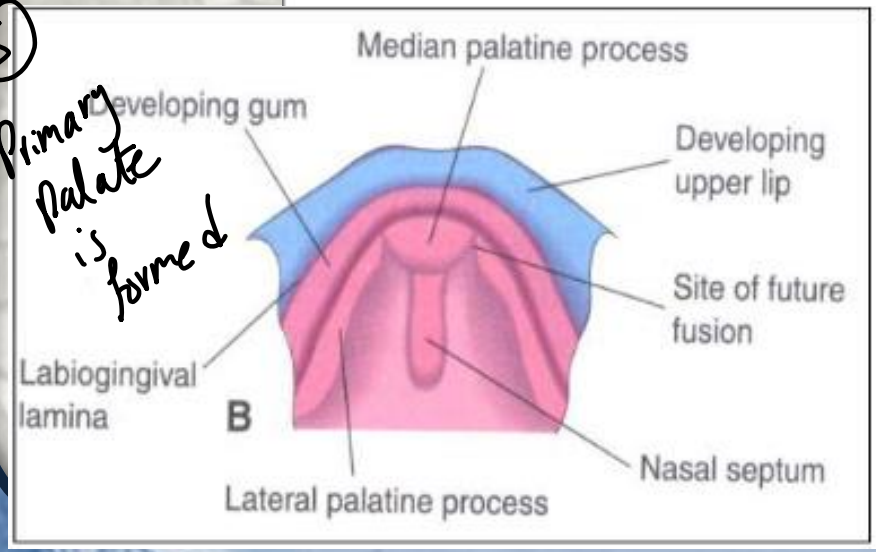
① medial nasal Prominence fuse together



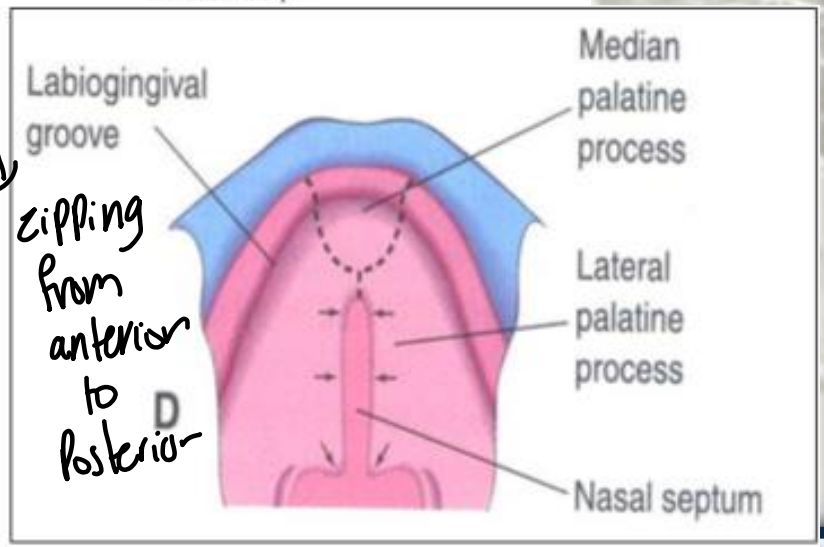
② then it fuses with maxillary forming: alveolar arches



③ Primary palate is formed

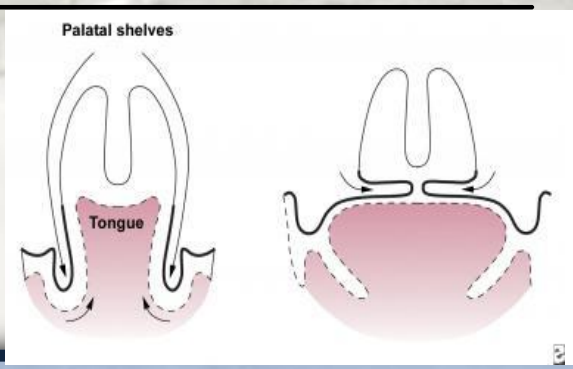


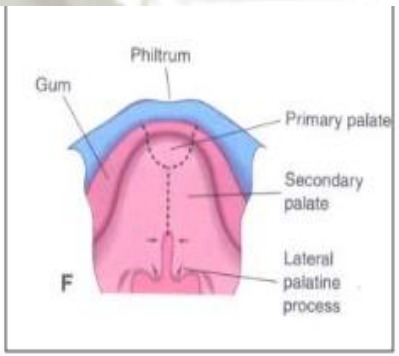
④ zipping from anterior to Posterior

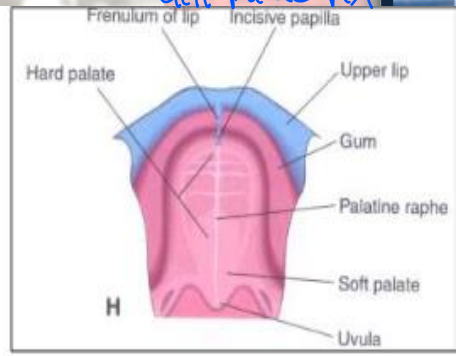


Secondary Palate

- 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)→ explain why cleft palate more

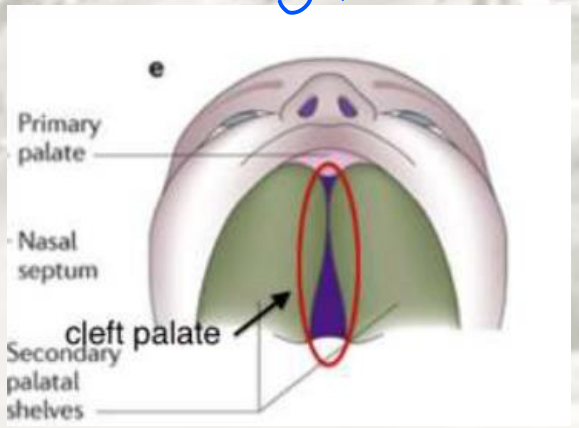


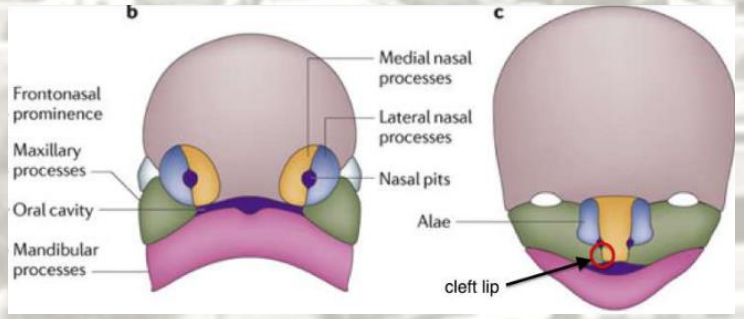




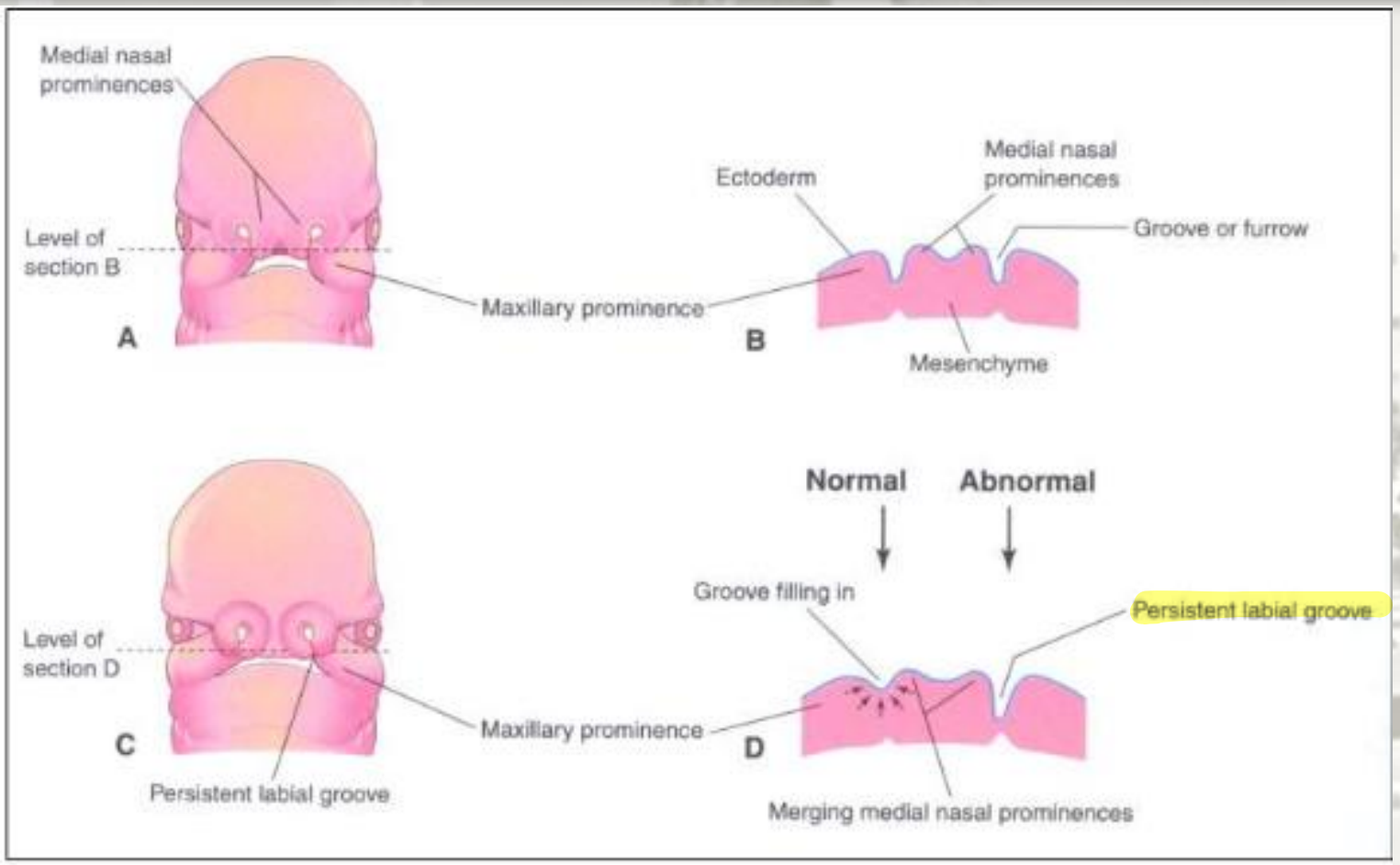
Formation of clefts

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

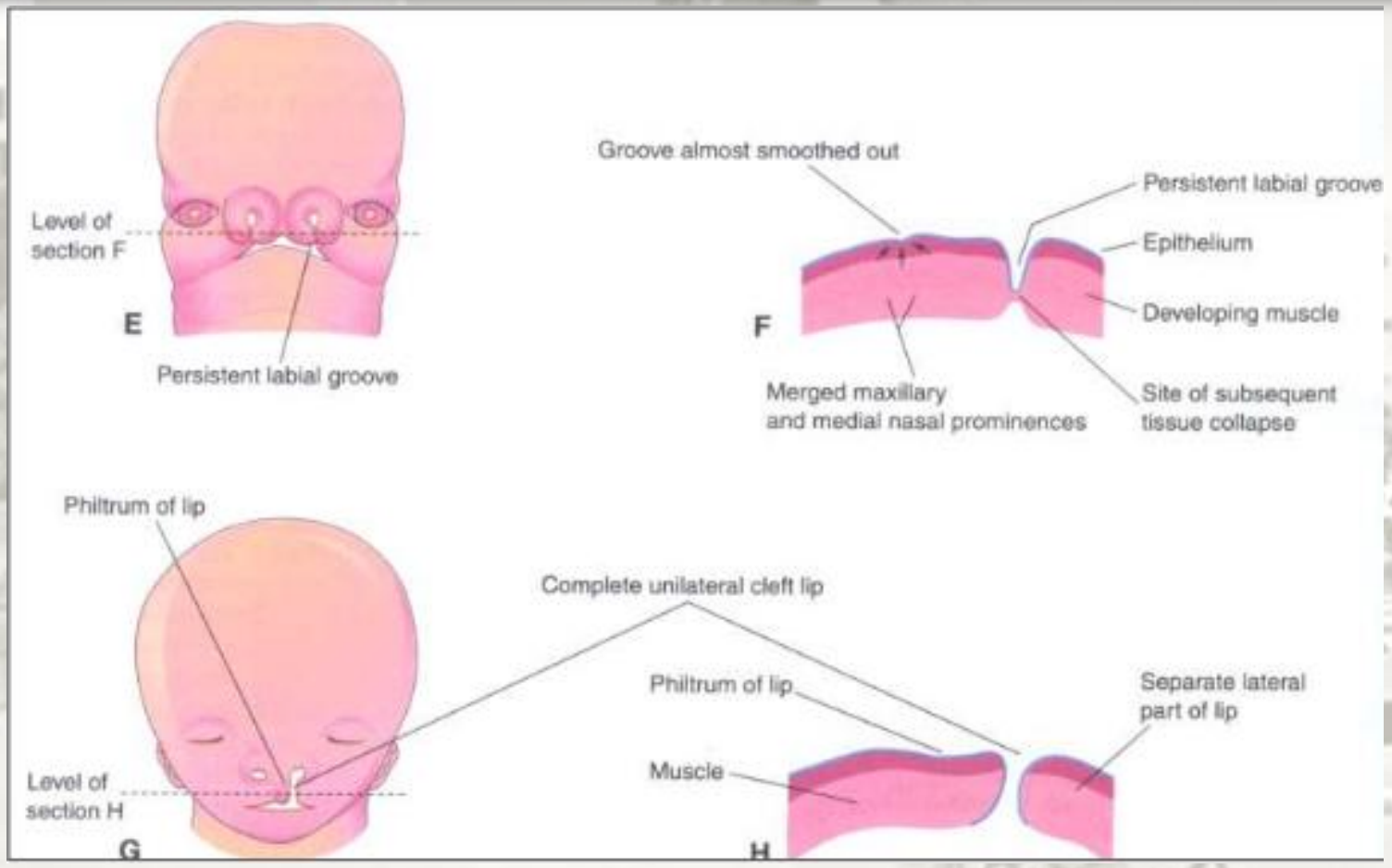




Formation of clefts



Formation of clefts



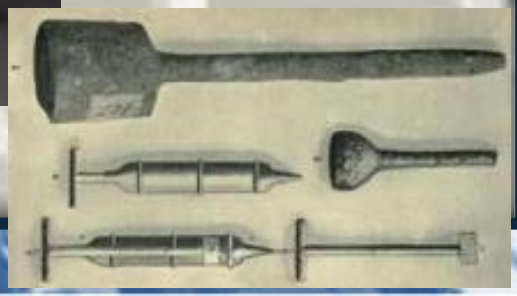
ETIOLOGY


CAUTION
RADIATION
HAZARD


Genetic factors is a
major contributing
cleft lip and cleft

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

↑ affected childs
↓
↑ % of having another child affected
↓
There is some source of familial cause but not exactly follow mendalian inheritance

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: less common
  - Associated with over 300 syndromes
    - Van der Woude syndrome – the most common
      - Autosomal dominant
      - Lower lip pits
      - abnormal genitalia
      - syndactely
      - Popliteal Pterygium syndrome

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

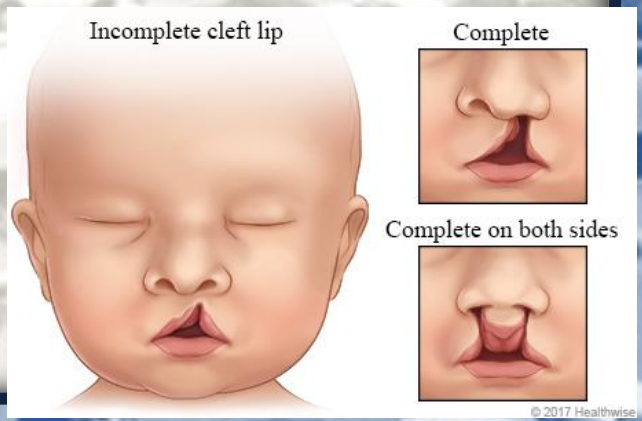
Unilateral cleft lip

- 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

- microform

A scar like , vermilion nutching , lip shortenning بتفكر المريض عامل عملية ، الخط زي

Cleft lip and palate



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

BILATERAL INCOMPLETE CLEFT LIP

MICROFORM RIGHT & COMPLETE LEFT CLEFT LIP

INCOMPLETE RIGHT & COMPLETE LEFT CLEFT LIP & ALVEOLUS

BILATERAL COMPLETE CLEFT LIP & PALATE

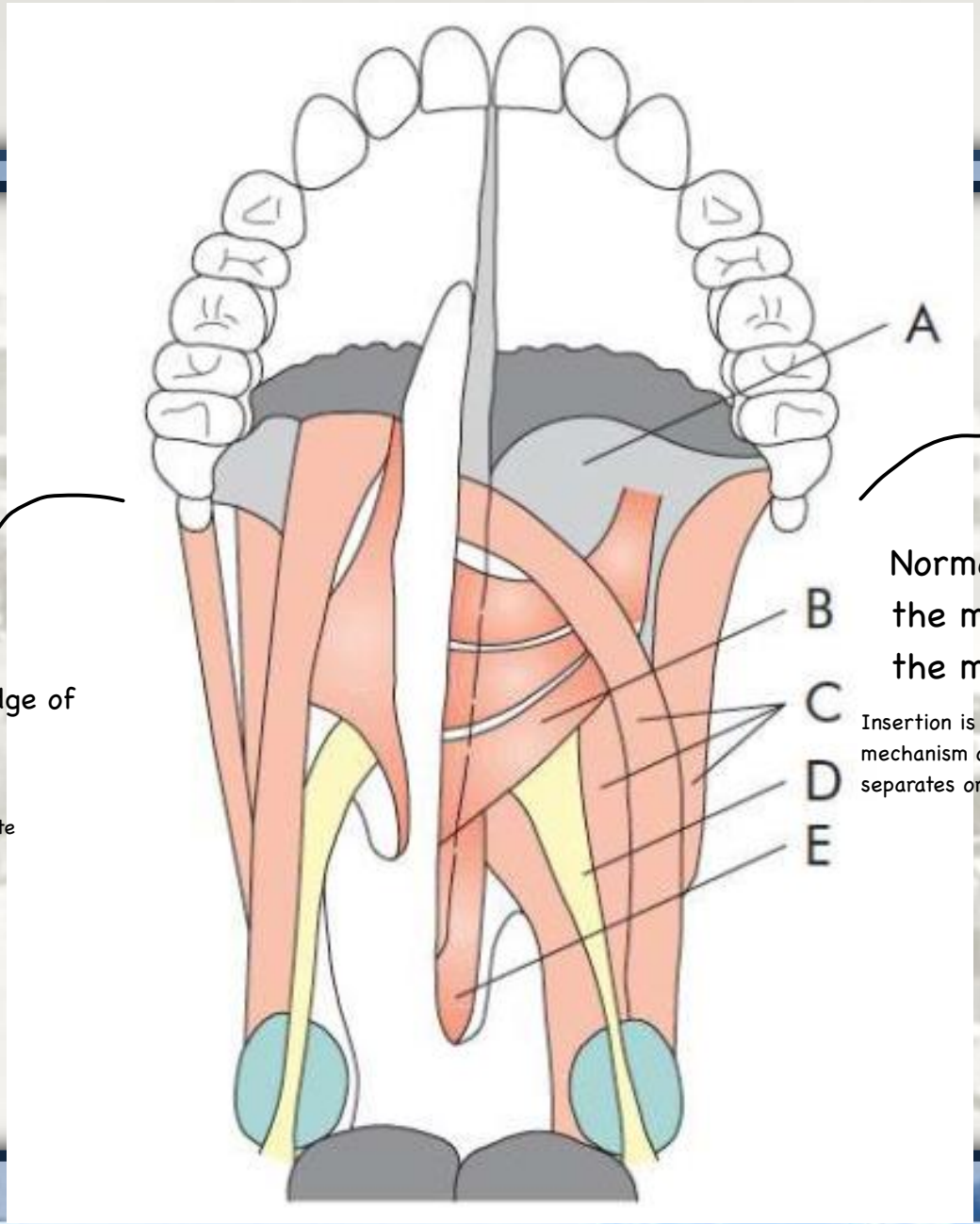
Flying away premaxilla :
There is no columella

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

Submucosal clefting : Palate is "seen" intact , but there is line called zuna pellocida , bifid uvula



Abnormal insertion : towards posterior edge of hard palate

Abnormal function of soft palate

Normal insertion of the muscles : towards the midline

Insertion is very important for the mechanism of soft palate that separates oral cavity from nasal cavity

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
    اللسان راجع لورا
    Lower jaw is undersized
  - Associated with several syndromes (stickler, velocardiofacial syndrome, etc.)
  - Management – prone positioning (severe cases sometimes require tracheostomy
    treat obstructive sleep apnea syndrome (OSAS
    - Mandibular distraction

      Method to increase the bone of the jaw

Feeding Difficulties

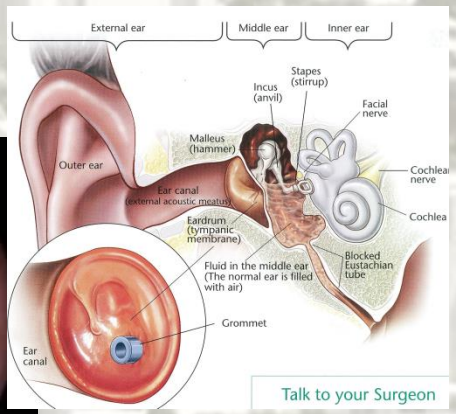
- Critical aspect in management
  - Nutrition and feeding
  - Cleft palate limits the ability to suck due to the common cavity
  - Cleft lip alone
  - Special bottles
  - Premaxillary orthopedics

Otological Manifestations

- Abnormal insertion of tensor veli palitini
- 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)

To drain fluid in middle ear

Otitis media with effusion

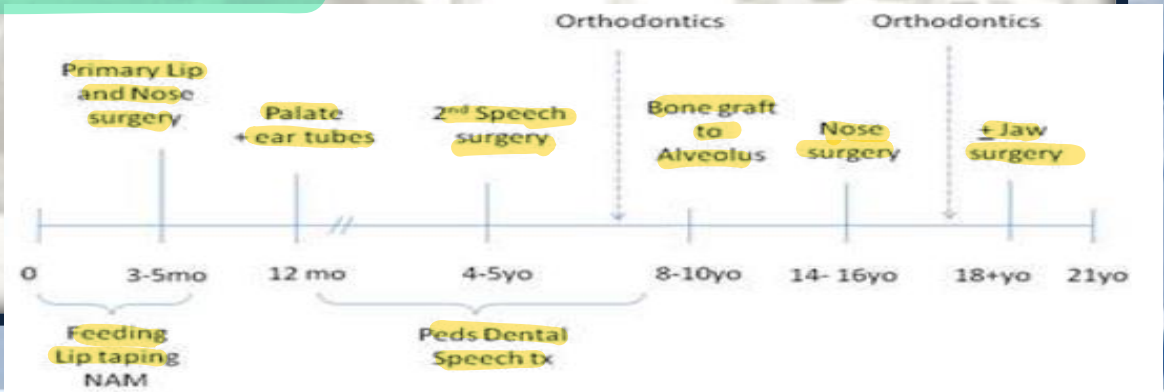


Speech Development

- 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



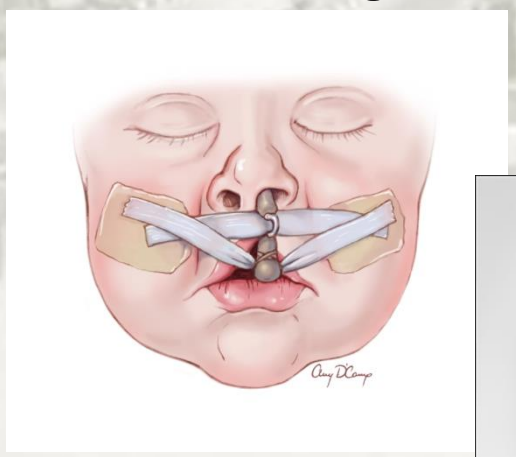
Presurgical

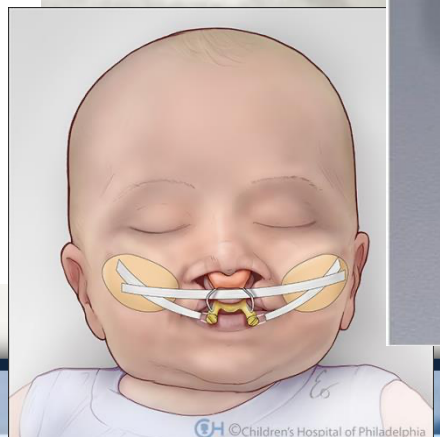
- Wide cleft lip or premaxilla protrusion
  - Advantageous to narrow the cleft and mold the premaxilla before proceeding with surgery
- Taping
  - Effective in reducing the width of the cleft in a
- nonsurgical manner
  - Strip of hypoallergenic tape is placed with tension across the cleft and secured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

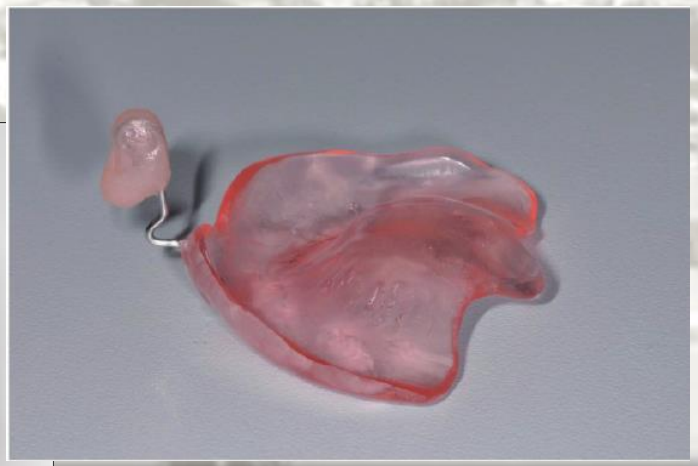
disadvantage

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

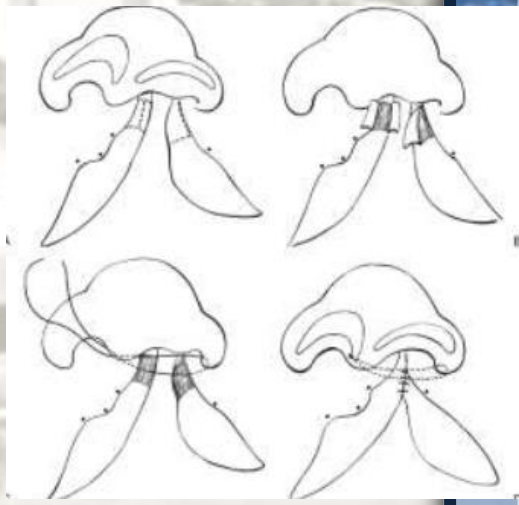






Presurgical

- 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



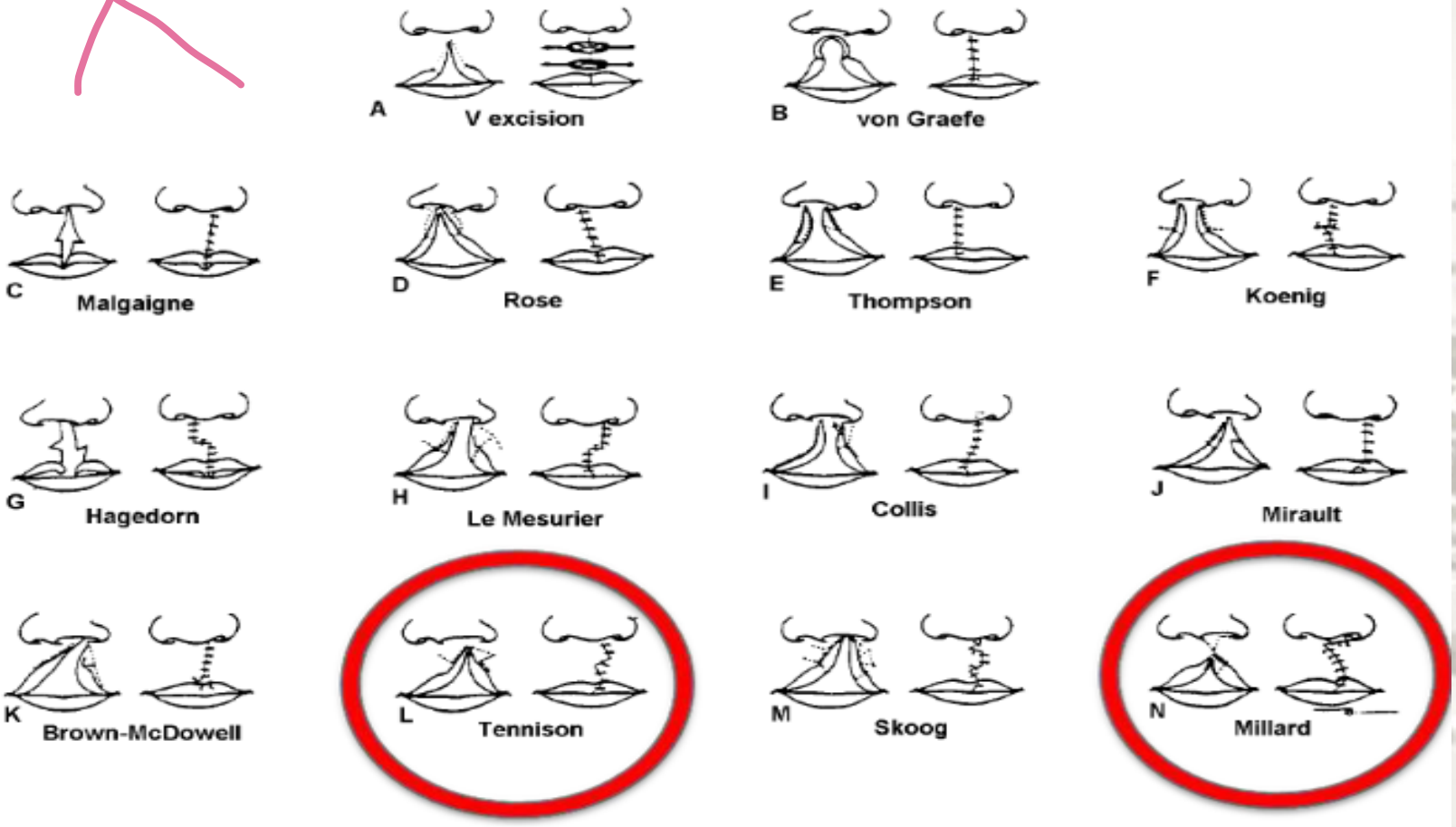
Cleft Lip Repair

- 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

Dish face deformity

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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