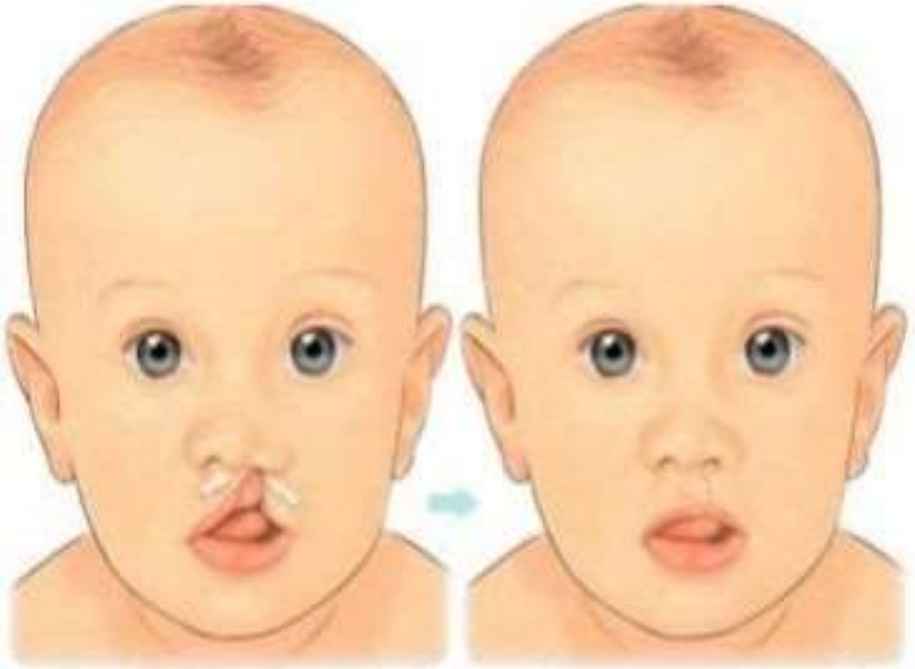
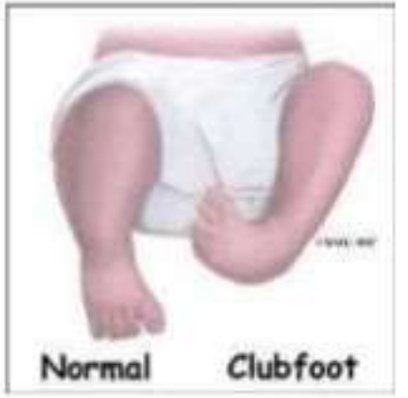
CLEFT LIP AND PALATE

By- Dr. Sonal Anchlia

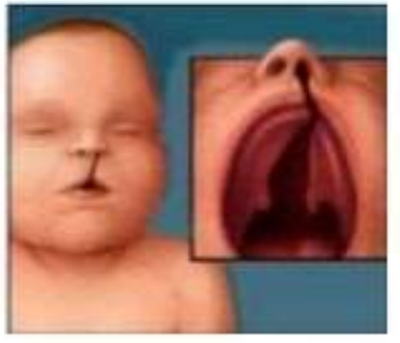


INTRODUCTION

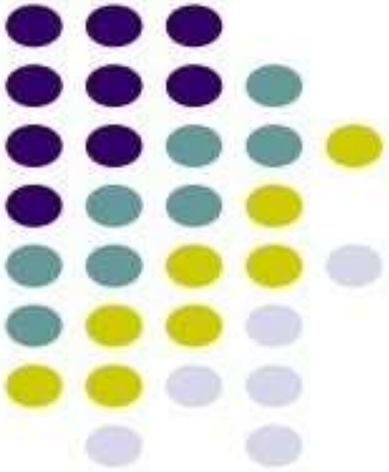
- Cleft lip and palate is the second most common congenital anamoly after clubfoot



- Amoung the 15 types of orofacial clefting, cleft lip and palate is the most common one.



GOALS OF SURGICAL CARE



GOALS OF SURGICAL CARE:

1. Normalized esthetic appearance of the lip and nose.
2. Intact primary and secondary palate.
3. Normal speech, language, and hearing.
4. Nasal airway patency.
5. Class I occlusion with normal masticatory function.
6. Good dental and periodontal health.
7. Normal psychosocial development.

INCIDENCE

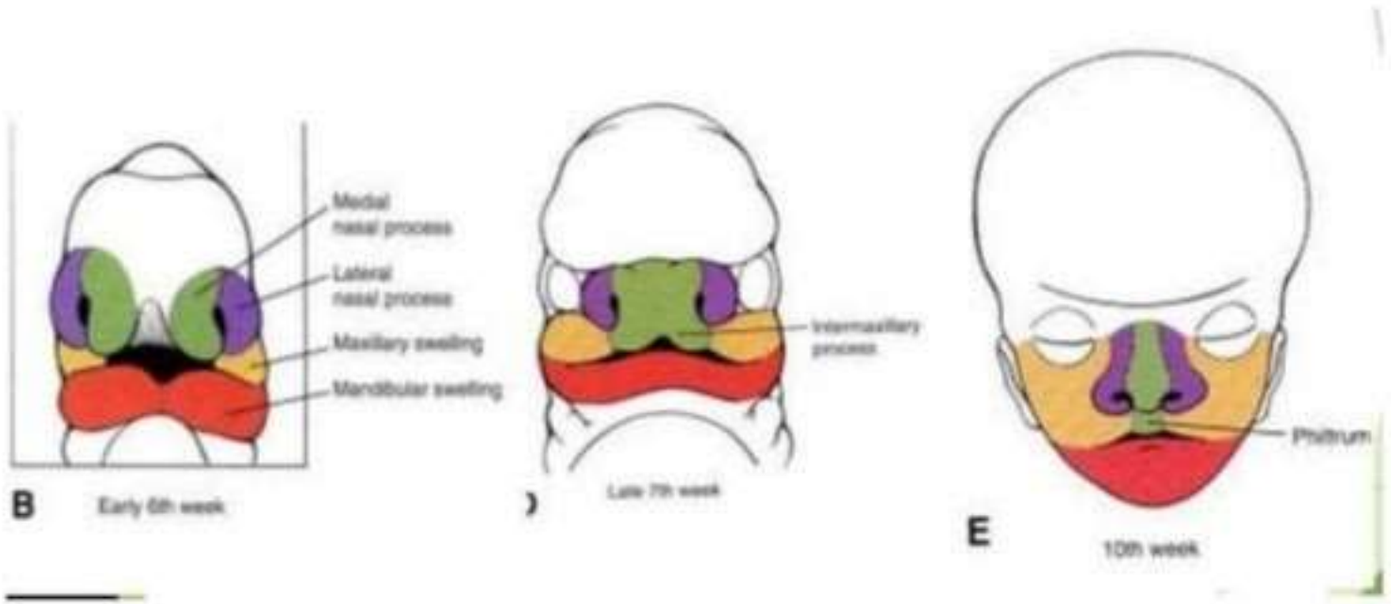
More common clefts are –

- Unilateral clefts
- Males female ratio is 2:1
- Left sided

Males more affected by cleft lip

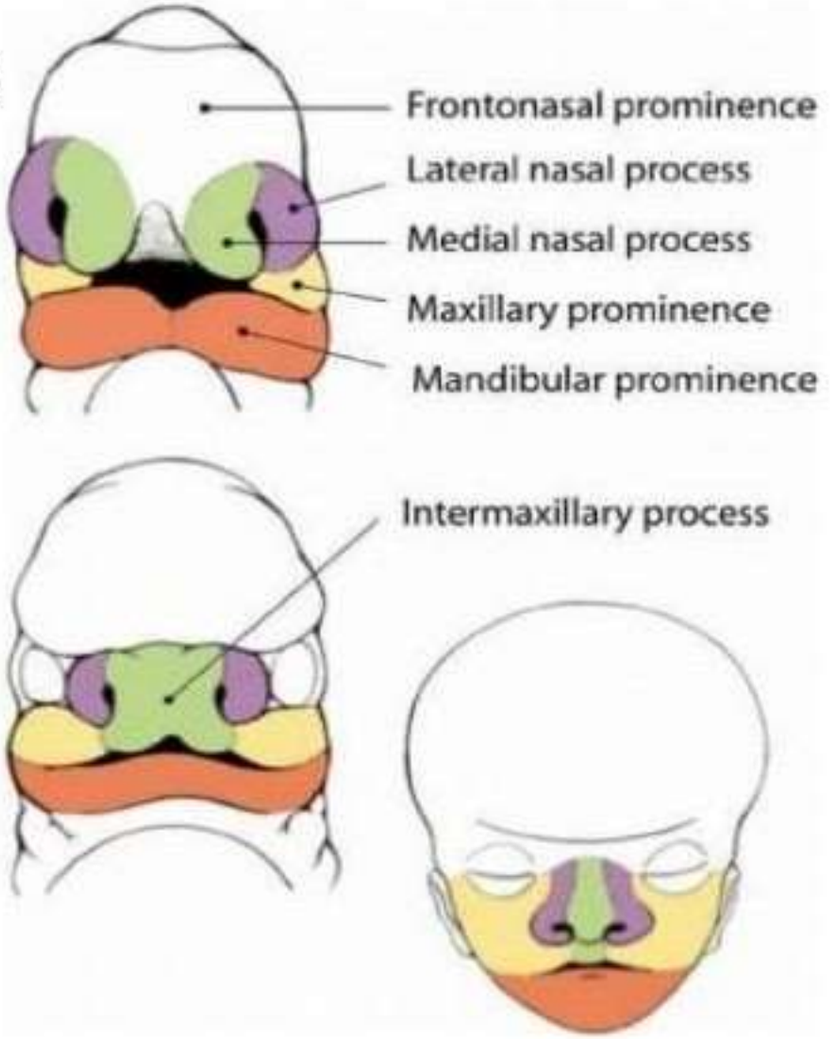
Females more affected by cleft palate

EMBRYOLOGY


Medial nasal process
Lateral nasal process
Maxillary swelling
Mandibular swelling
B
Early 6th week
Intermaxillary process
Philtrum
E
10th week

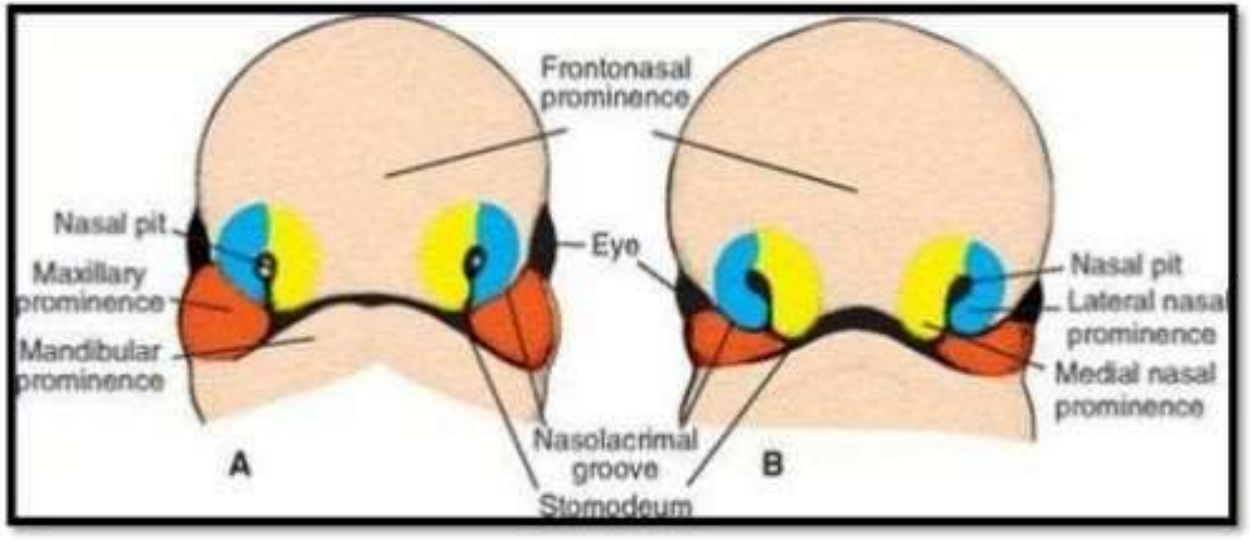
EMBRYOLOGY

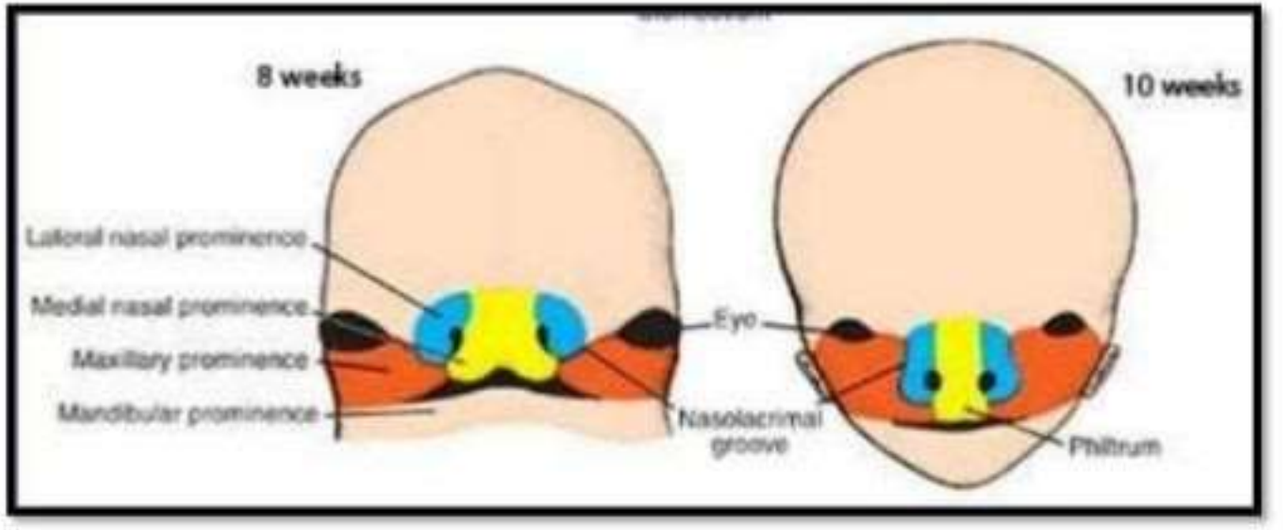
- Development of facial structures starts at the end of 4 th week
- The lateral swellings will form the alae of the nose.
- The medial swellings will give rise to four areas:
  1. The middle portion of the nose.
  2. The middle portion of the upper lip.
  3. The middle portion of the maxilla.
  4. The maxillary swellings will approach the medial and lateral nasal swellings but remain separated from them by well marked grooves.



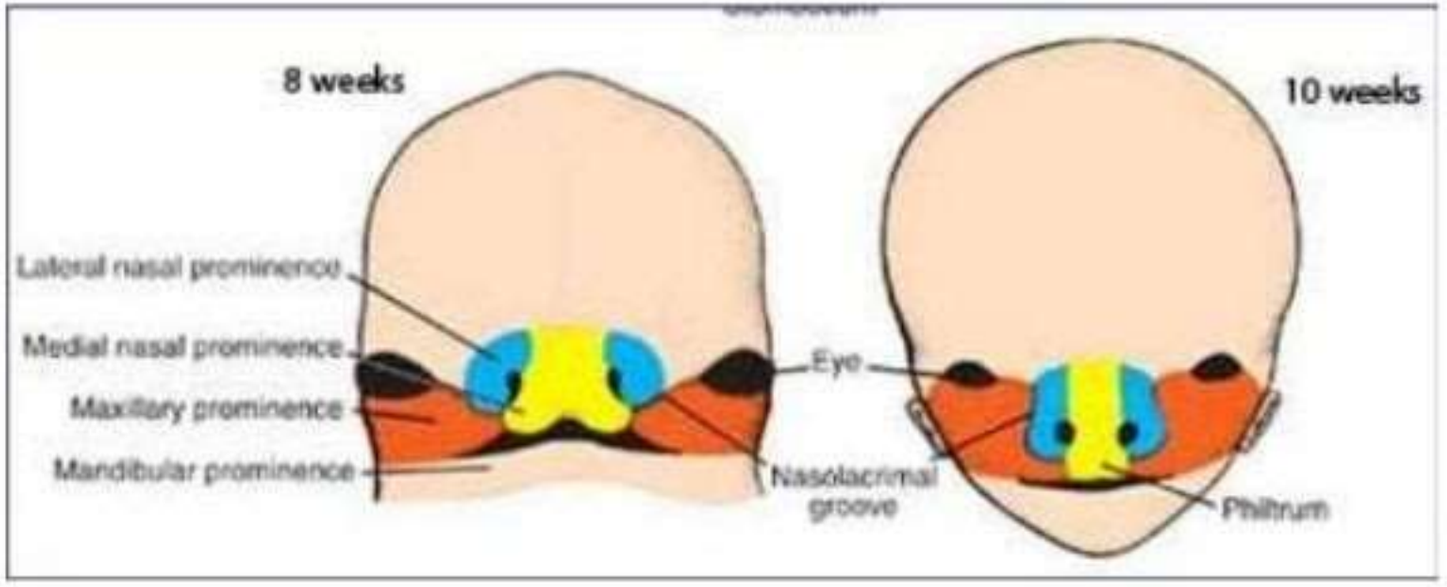
- These swellings simultaneously merge with each other and with the maxillary swellings laterally.

- The **upper lip** is formed by the **two medial nasal swellings and the two maxillary swellings.**





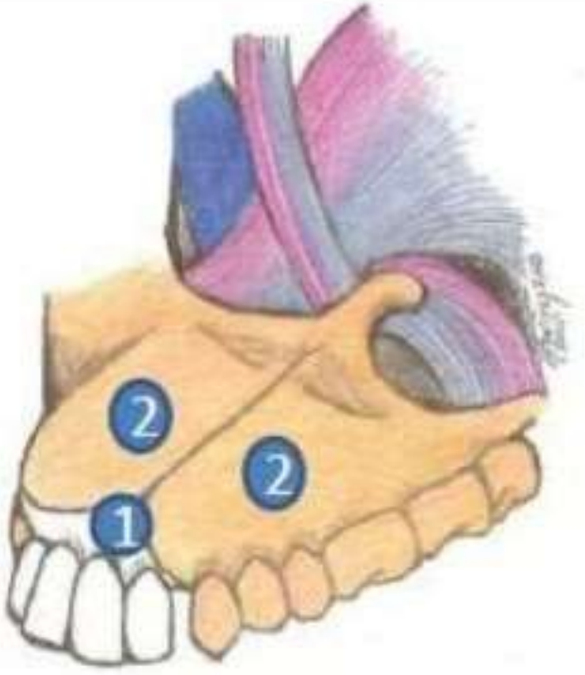
- The maxillary and lateral nasal process separated by nasolacrimal groove/duct
- **Frontonasal process** – bridge of the nose
- **Medial nasal process** – tip of nose and philtrum of upper lip
- **Lateral nasal process** – ala of the nose

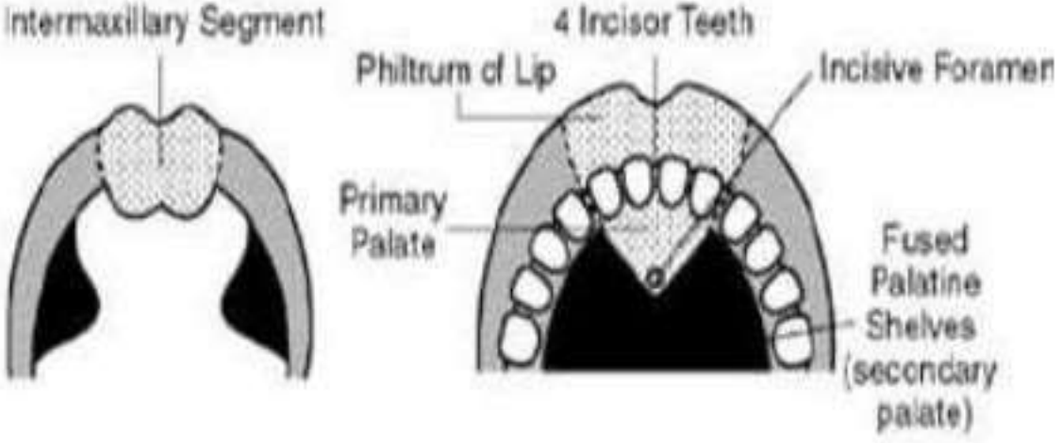


DEVELOPMENT OF THE PALATE

It develops from two parts:

1. The primary palate.
2. The secondary palate



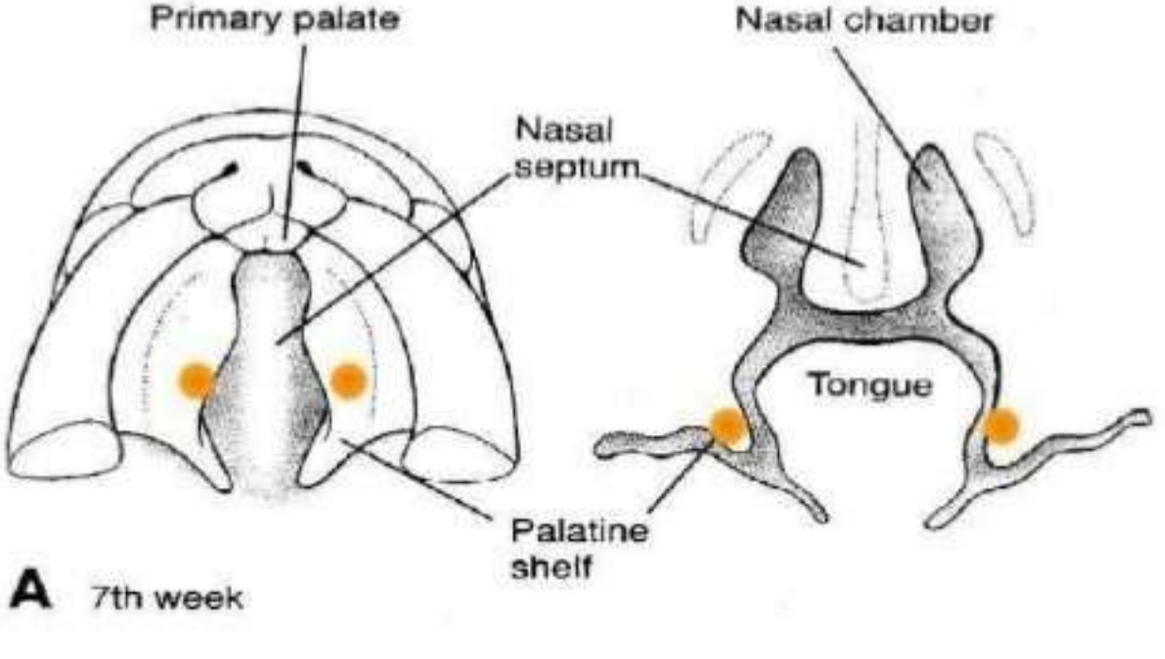


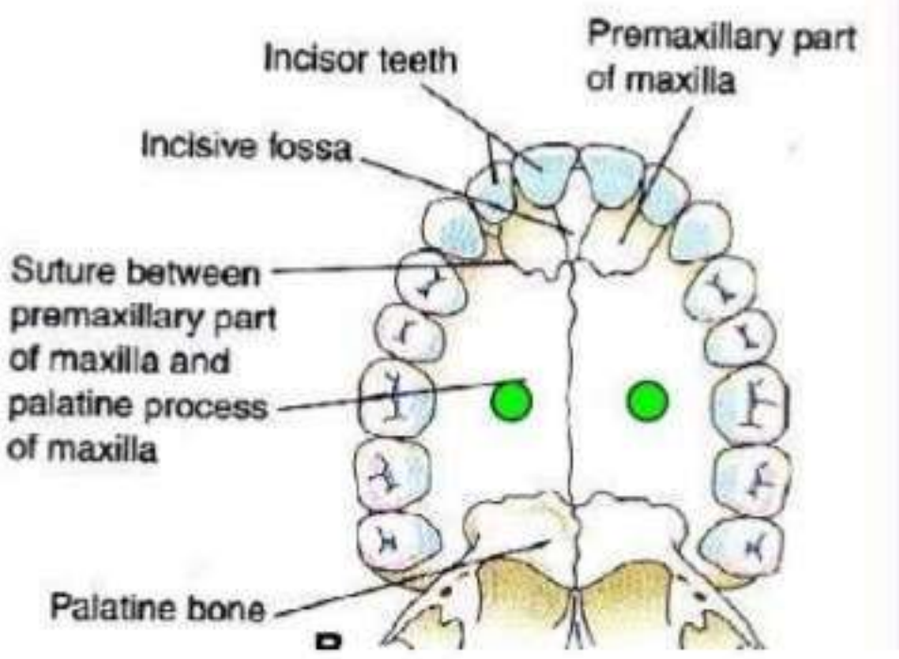
PRIMARY PALATE

- The primary palate appears earlier than the secondary palate at the beginning of the 6th W.I.U.
- The primary palate develops from the inter-maxillary segment.
- At first the primary palate and the central parts of the upper lip are formed as one unite.
- At the 8th week IU the primary palate is separated from the upper lip by the formation of the vestibular lamina.

SECONDARY PALATE

- The secondary palate is that part of the palate posterior to the incisive fossa.
- It formed of the remaining part of the hard palate and the soft palate .

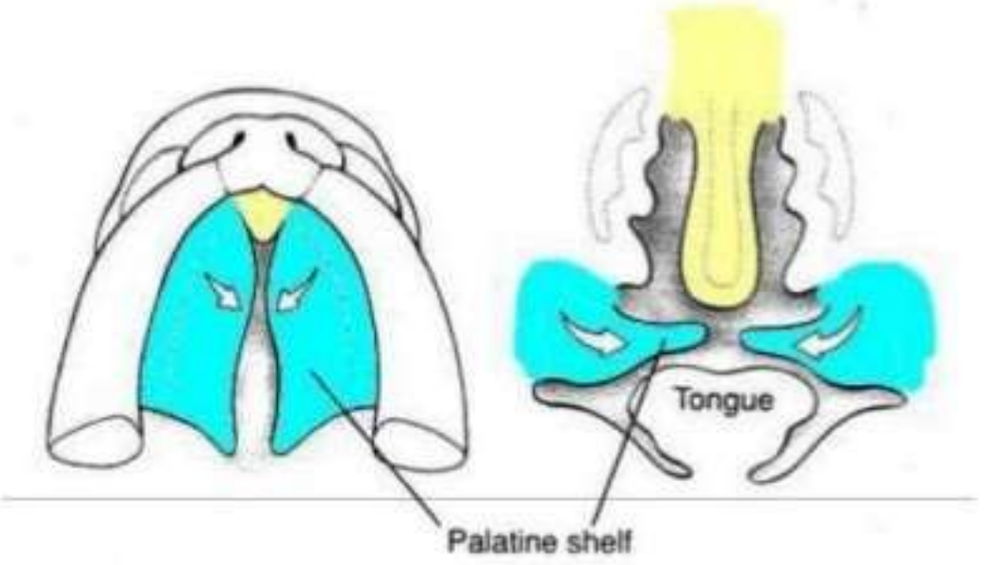




Palatal Shelf Elevation

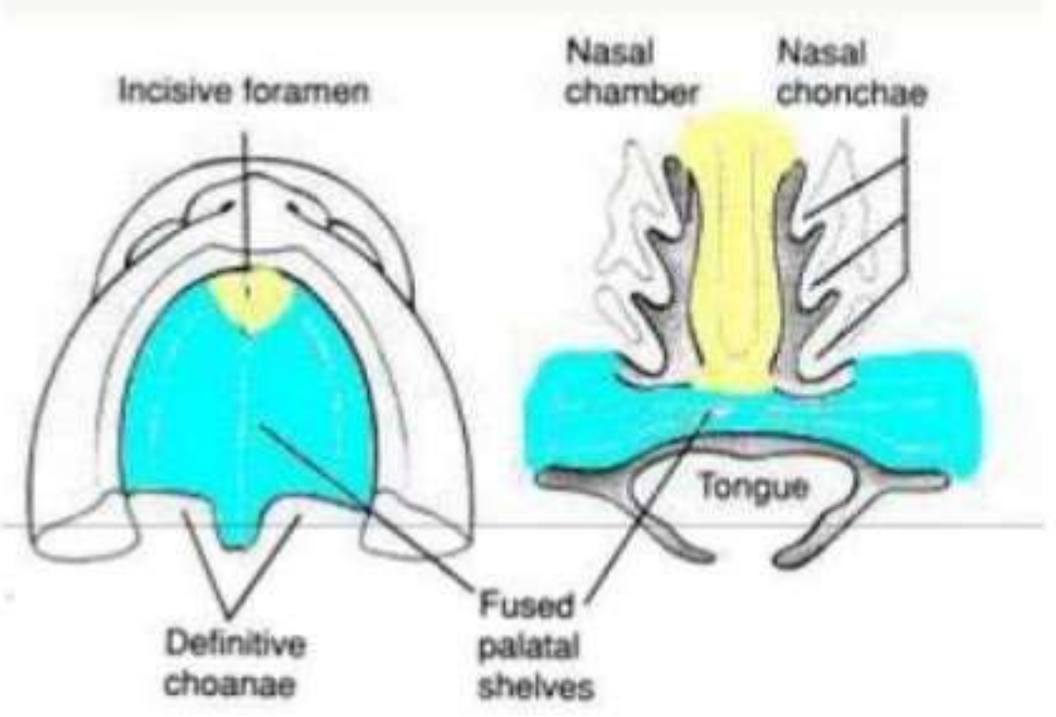
- From 8-9 weeks I.U., the lateral palatine shelves slide or roll over the tongue and acquire a horizontal position (shelf elevation).

- This process occurs when the shelves have developed sufficient strength to slide over the tongue.



Fusion of the Primary & Secondary Palates

- The palatine closure begins from the 9th to the 12th W.I.U. After the shelves are in a horizontal position and they attain their final growth, shelf fusion occurs.



TERMINOLOGY

1. **The primary palate** comprises the anatomic structures anterior to the incisive foramen (alveolus, maxilla, piriform, lip).

2. **The secondary palate** comprises the anatomic structures between the incisive foramen and the uvula.

3. So, complete cleft of the primary and secondary palates involves the maxilla, alveolus, hard palate, and soft palate.

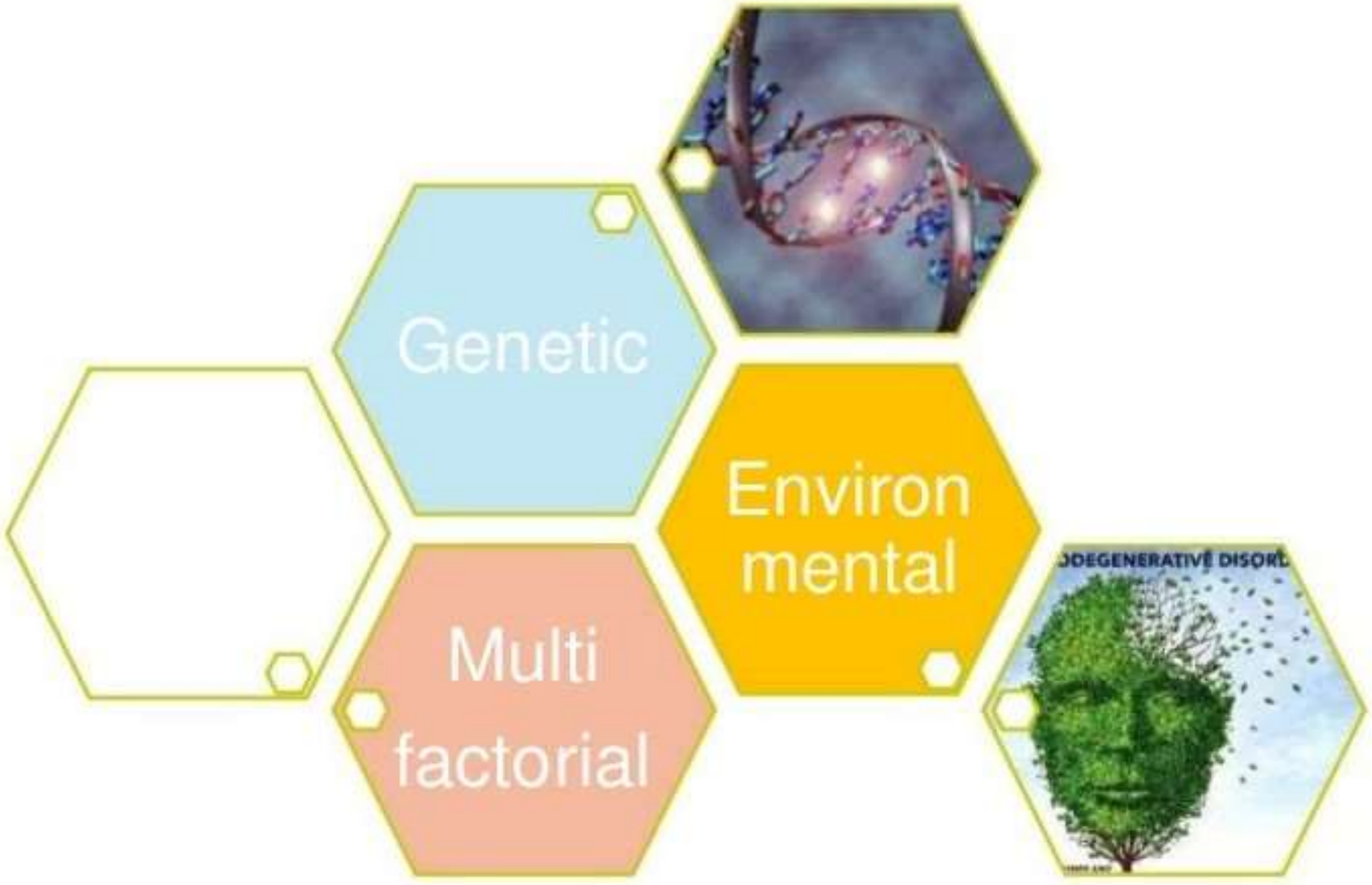
4. Isolated cleft palate involving the hard and soft palate (without affecting the alveolar ridge) → **complete cleft of the secondary palate.**

5. Cleft involving only the soft palate (and not the hard palate or alveolus) → **incomplete cleft of the secondary palate.**

FORMATION OF CLEFTS

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

ETIOLOGY


Genetic
Environ
mental
Multi
factorial

1] Genetic

- Genetic disorders are classified into 4 groups

  1. Chromosomal
  2. Single gene
  3. Multifactorial – oral clefts
  4. Mitochondrial

2] Multifactorial because :

1) Chances increases if more than one family member if affected
2) More the severity, greater the chances of recurrence in sibling
3) Higher risk if affected individual is of less affected sex
4) Risk decreases in remotely related individuals
5) Consanguinity increases the rate because of sharing of genes

3] Environmental factors

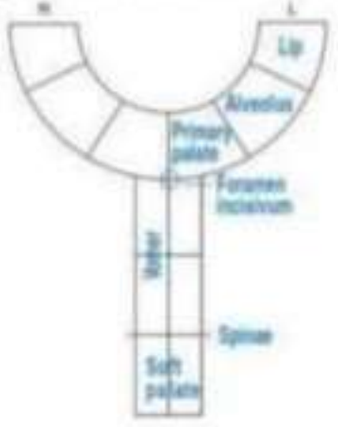
- Maternal smoking or tobacco exposure
- Viral infections
- Poor nutrition
- Certain Medicinal drugs
- Teratogens like:
  - ➢ Rubella virus, Cortisone/ steroids, Mercaptopurine, Methotrexate, Valium, Dilantin

PREDISPOSING FACTORS

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

CLASSIFICATION

The Striped-Y Classification system



CLASSIFICATION

Bixler divided oral clefts into three groups

1. Syndromic / single-gene / chromosomal or environmental
   - 1% of CLP AND 8% of isolated cleft palate
2. Familial
   - 25% of CLP and 12% of isolated cleft palate
3. Isolated / non-familial
   - 75% of CLP and 80% of isolated cleft palate

Davis and Ritchie's classification (1922)

Group I: Prealveolar process cleft (clefts affecting the lip)

1. Unilateral (right/left: complete/incomplete)
2. Bilateral (right: complete/incomplete; left: complete/incomplete)
3. Median (complete/incomplete)

Group II: Postalveolar process cleft (clefts affecting the palate)

1. Soft palate
2. Hard palate

Group III: Alveolar process cleft (any cleft involving the alveolar process)

1. Unilateral (right/left: complete/incomplete)
2. Bilateral (right: complete/incomplete; left: complete/incomplete)
3. Median (complete/incomplete)

Victor Veau's classification (1931)

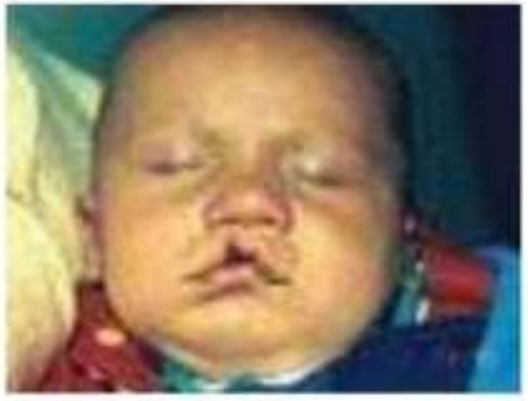
A] Cleft lip

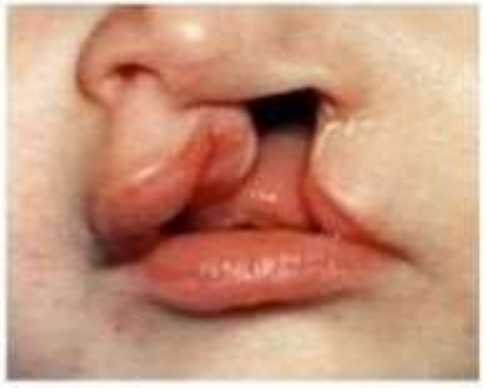
Class I : U/L notching of vermillion border, not extending into the lip.

Class II : cleft extending into the lip, but not including the floor of the nose.

Class III: extending into the floor of the nose.

Class IV: any b/l cleft of the lip, whether incomplete or complete.





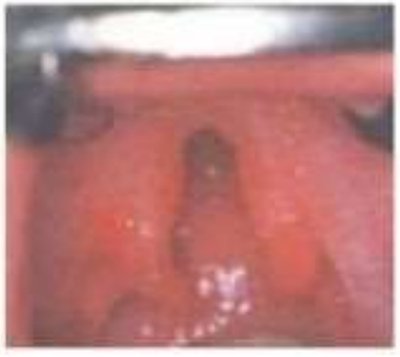
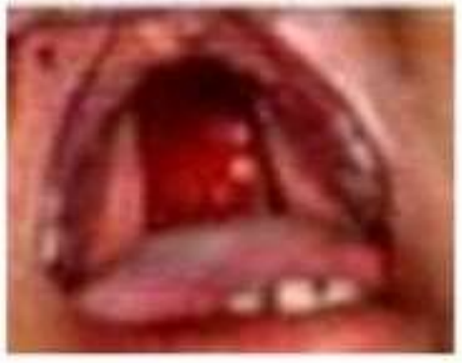
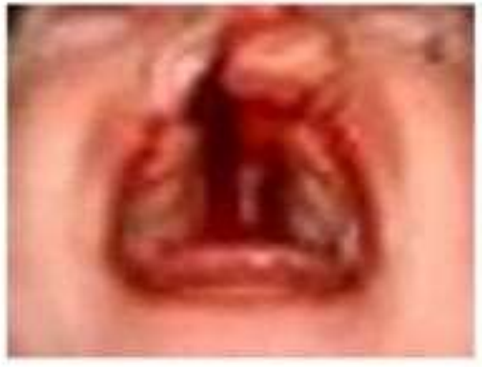
B] Cleft palate

Class I : soft palate

Class II : soft/hard palate extending no further than incisive foramen.

Class III: complete unilateral cleft, extending from uvula to incisive foramen, then deviating to one side

Class IV: two clefts extending forward from the incisive foramen into the alveolus.


Fogh- Andersen's classification (1942)

1. H arelip (single or double)
2. H arelip with cleft palate
3. Isolated cleft palate
4. R are atypical clefts, e.g., median cleft lip

Kernahan and Stark's classification (1958)

1. Clefts of structures anterior to the incisive foramen
2. Clefts of structures posterior to the incisive foramen
3. Clefts affecting structures anterior and posterior to the incisive foramen

American Cleft Palate – Craniofacial Association (ACPA) classification (1962)

1. Clefts of the prepalate (cleft of lip and embryologic primary palate)

a. Cleft lip (cheiloschisis)
b. Cleft alveolus (alveoloschisis)
c. Cleft lip, alveolus, and primary palate (cheiloalveoloschisis)

2. Clefts of the palate (cleft of the embryologic secondary palate)

a. Cleft of the hard palate (uranoschisis)
b. Cleft of the soft palate (staphyloschisis or veloschisis)
c. Cleft of the hard and soft palate (uranostaphyloschisis)

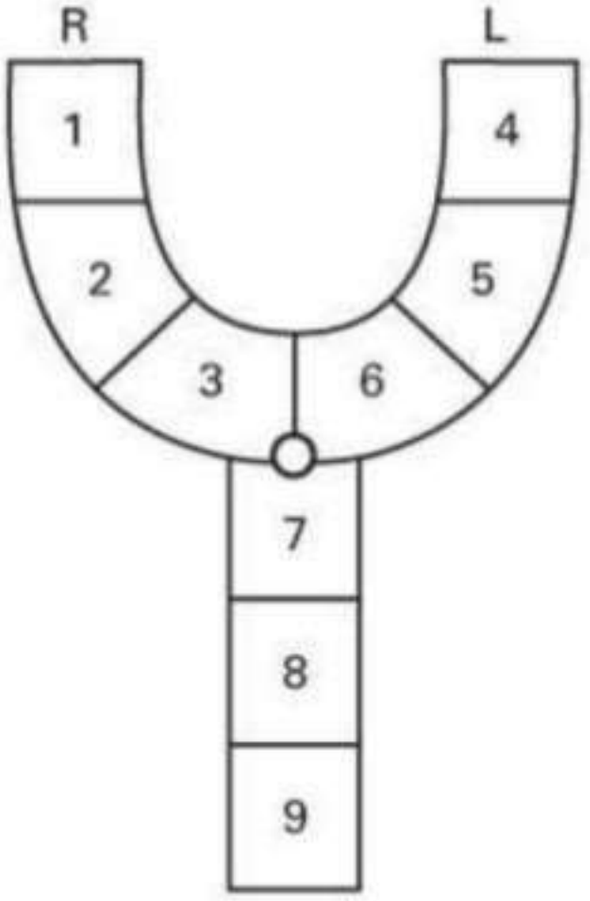
3. Clefts of the prepalate and palate (alveolocheilopalatoschisis)

4. Facial clefts other than prepalatal and palatal

a. Cleft of the mandibular process
b. Naso-ocular clefts
c. Oro-ocular clefts
d. Oroaural clefts

Kernahan and Stark's stripped Y classification (1971)

1 R lip
2 R alveolus
3 R palate anterior to the incisive foramen
4 L lip
5 L alveolus
6 L palate anterior to the incisive foramen
7 Anterior hard palate
8 Posterior hard palate
9 Soft palate



Spina's classification (1973)

Group I: Preincisive foramen clefts

a. Unilateral
b. Bilateral
c. Medial

Group II: Transincisive foramen clefts

a. Unilateral
b. Bilateral

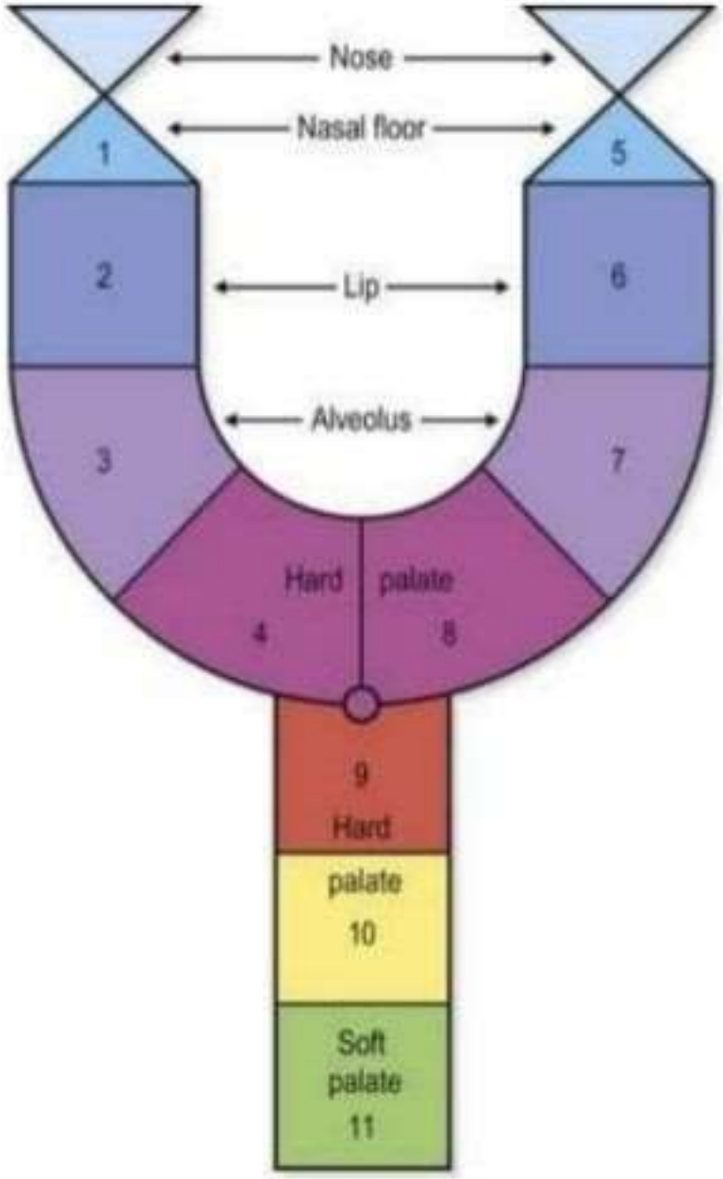
Group III: Post incisive foramen clefts

a. Total
b. Partial

Group IV: Rare facial clefts

Kernahan and Stark's stripped-Y:

Modification by Ehlsaky (1973) and Millard (1976)



LAHSHAL classification by Okriens (1987)

- Lahshal is a paraphrase of the anatomic areas affected by the cleft.

L – lip

A – alveolus

H – hard palate

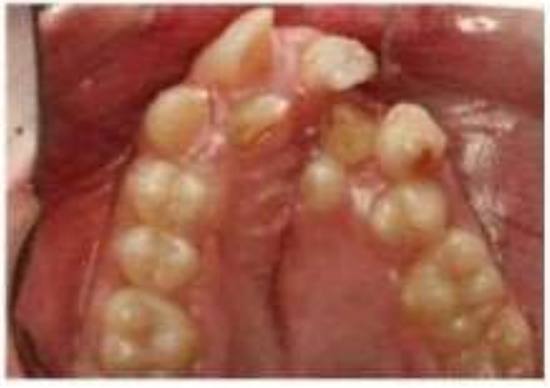
S – soft palate

H – hard palate

A – alveolus

L – lip

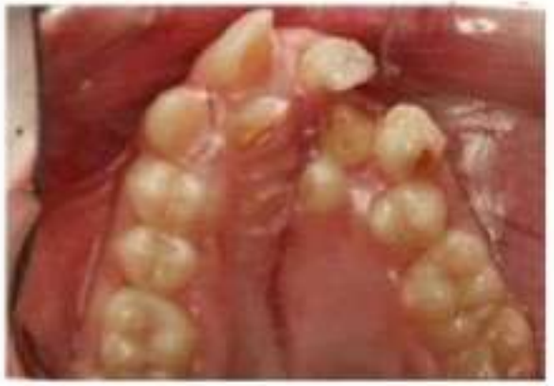
PROBLEMS ASSOCIATED WITH CLEFT LIP & PALATE

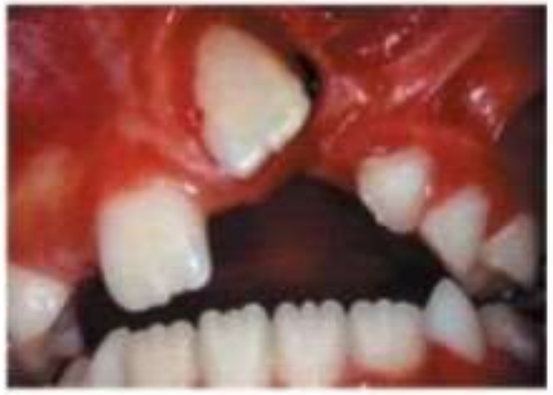


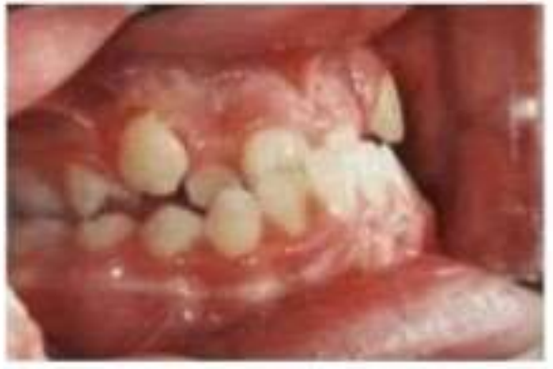

Dental
Skeletal
Nasal
Feeding
Ear problems
Speech
Associated anomalies

1. DENTAL PROBLEMS

- Tooth agenesis, hypodontia (most common)
- Supernumerary teeth (2 nd most common)
- Enamel hypoplasia (CI)
- Crossbites
- Ectopic eruption, transposition
- Taurodontism, dilacerations







2. SKELETAL PROBLEMS

- Maxillary deficiency
- Mandibular prognathism
- Class III malocclusion
- Concave profile

3. NASAL PROBLEMS

Alae flared, Columella pulled to non cleft side.

4. EAR PROBLEMS:

Recurrent Middle Ear infection, Possible Deafness.

5. SPEECH PROBLEMS:

Retardation of consonants, Hypernasality, Articulation Defects, Hearing problem

6. ASSOCIATED ANOMALIES:

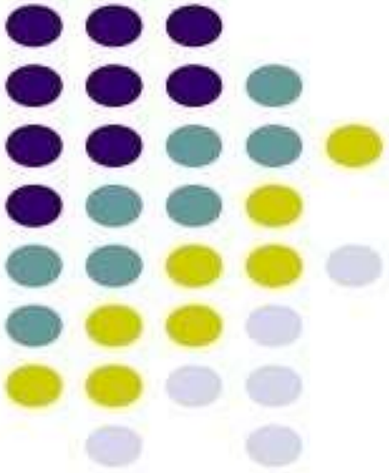
Congenital heart defects, Mental retardation

7. FEEDING PROBLEMS

- Infants with cleft palate can have difficulty feeding due to the inability to form an adequate seal between the tongue and palate for creation of sufficient negative pressure to suck fluid from a bottle.
- Specialized nipples and bottles are necessary to improve feeding immediately after birth.

- **The most useful devices combine:**
    1. Oversized nipples with reservoir spaces and large openings.
    2. Squeezable bottle to push fluid into the nipple assembly.

MANAGEMENT OF CLEFT LIP & PALATE



MULTIDISCIPLINARY MANAGEMENT

TIMING AND SEQUENCING OF SURGICAL-ORTHODONTIC TREATMENT

Procedure	*Timing*
Cleft lip repair	After 10 weeks
Cleft palate repair	9–18 months
Pharyngeal flap or pharyngoplasty	3–5 years or later based on speech development
Maxillary/alveolar reconstruction with bone grafting	6–9 years based on dental development
Cleft orthognathic surgery	14–16 years in girls, 16–18 years in boys
Cleft rhinoplasty	After age 5 years but preferably at skeletal maturity; after orthognathic surgery when possible
Cleft lip revision	Anytime once initial remodeling and scar maturation is complete but best performed after age 5 years

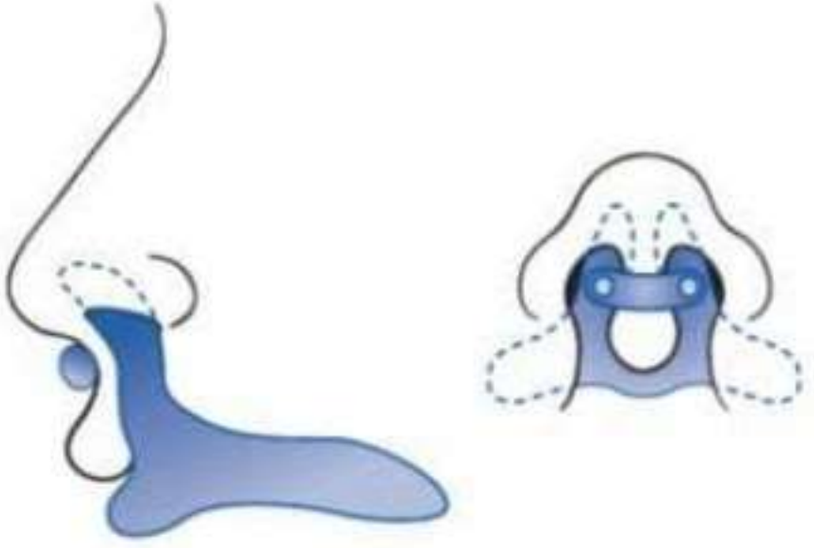
PRESURGICAL TAPING

- Facial taping with elastic devices is used for application of selective external pressure and may allow for improvement of lip and nasal position prior to the lip repair procedure.

- Some surgeons prefer presurgical orthopedic (PSO) appliances rather than lip taping to achieve the same goals.

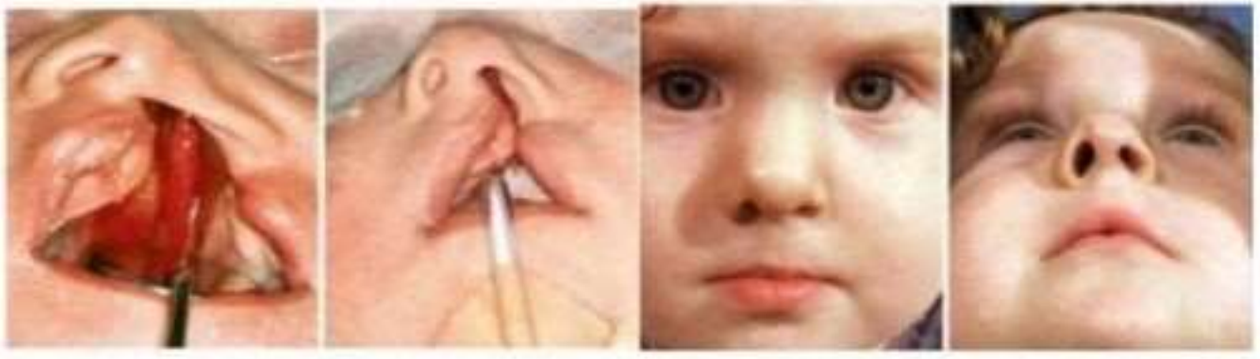
PRESURGICAL ORTHOPEDICS

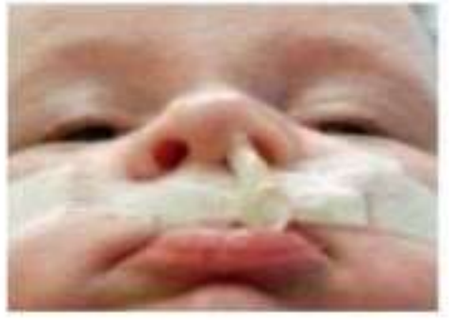
- Appliances are composed of a custom-made acrylic base plate that provides improved anchorage in the molding of lip, nasal, and alveolar structures during the presurgical phase of treatment.

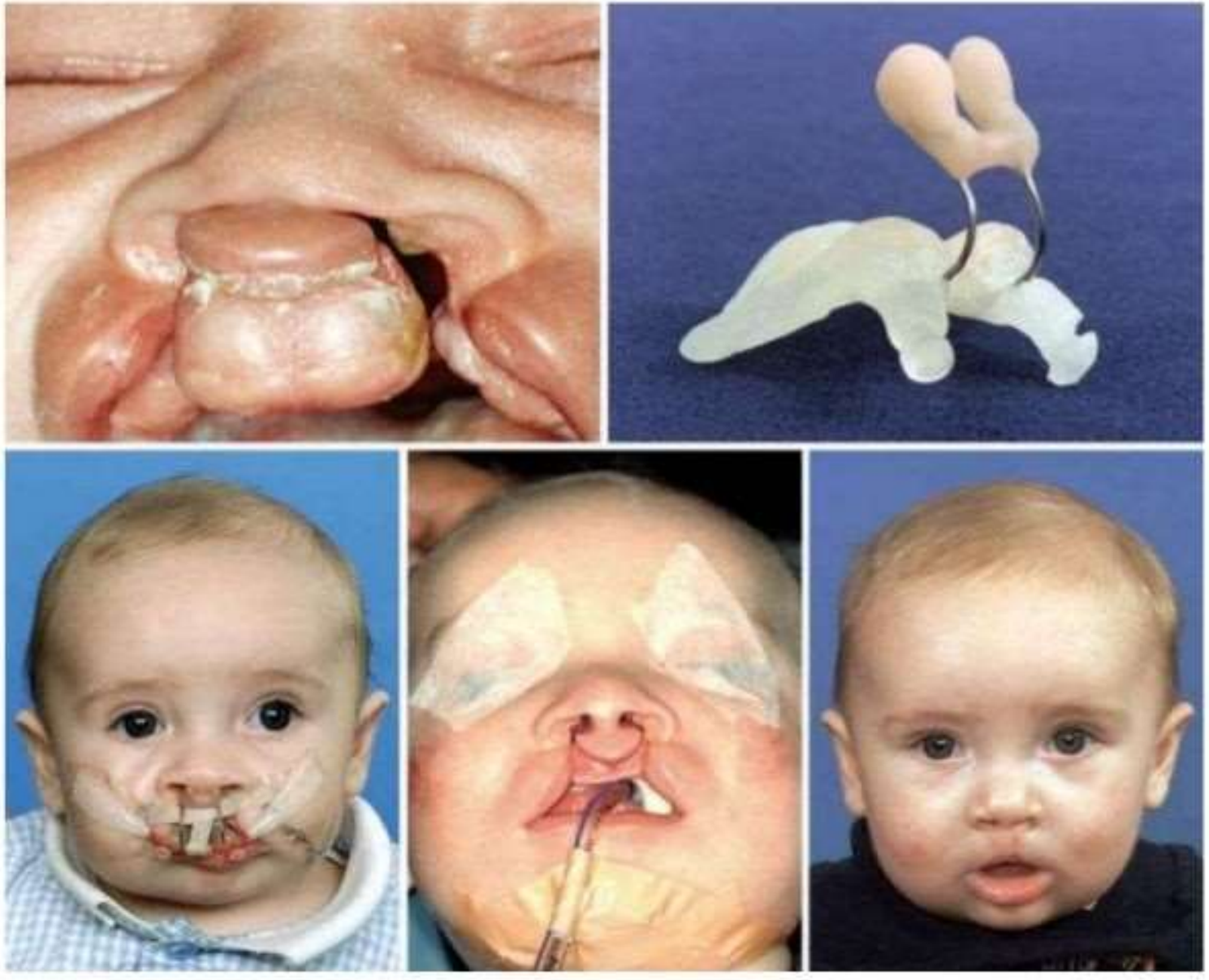


Objectives:

- Reduce severity of initial cleft deformity
- Columella – Nonsurgical lengthening (in bilateral clefts) and uprighting (in unilateral clefts)
- Reduction in the width of the alveolar cleft segments until passive contact of the gingival tissues is achieved.

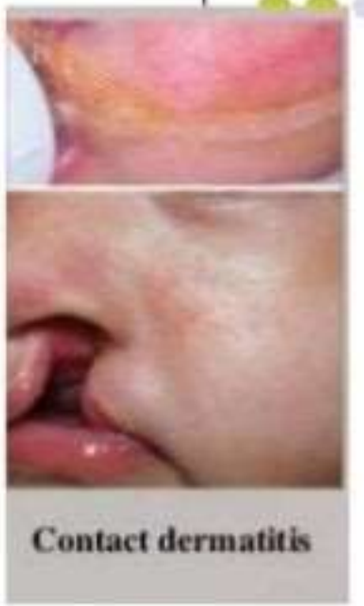






Complications and disadvantages:

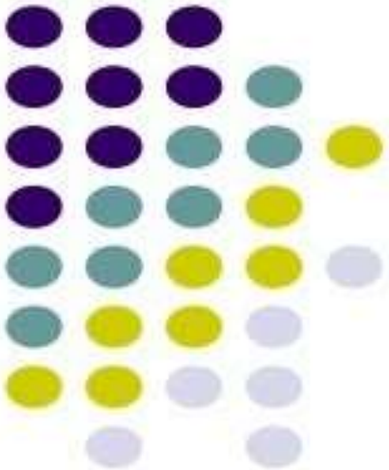
- Irritation of the oral mucosal or gingival tissue
- Ulceration of intraoral tissues
- The intranasal lining of the nasal tip can become inflamed
- Skin irritation due to tape usage
- Parent compliance required
- Moulding plate may get dislodged and obstruct the airway



Contact dermatitis

Overactivation of the nasal stent may produce bruising or petechiae in the dome area.

CLEFT LIP

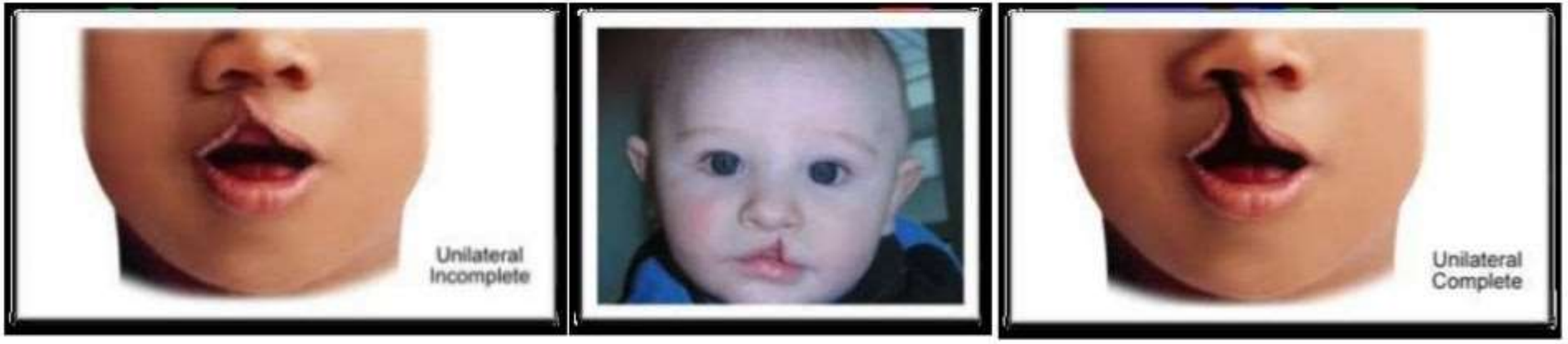


CLEFT LIP REPAIR

- Goal: improve facial aesthetics by restoring nasal and lip contour
- Timing: 3 to 6 months
- **Millards "RULE OF TEN"**
  - 10 weeks (age)
  - 10 pounds (weight)
  - 10 gm/dl (Hb)

1- Unilateral Cleft Lip Repair

- Clefts of the lip and nose that are unilateral present with a high degree of variability.
- The repair technique is usually performed after 10 weeks of age.



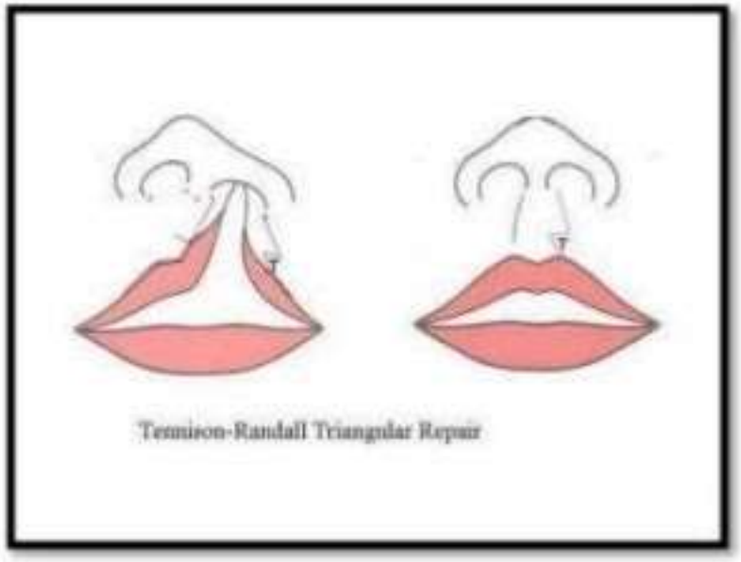
1- Unilateral Cleft Lip Repair

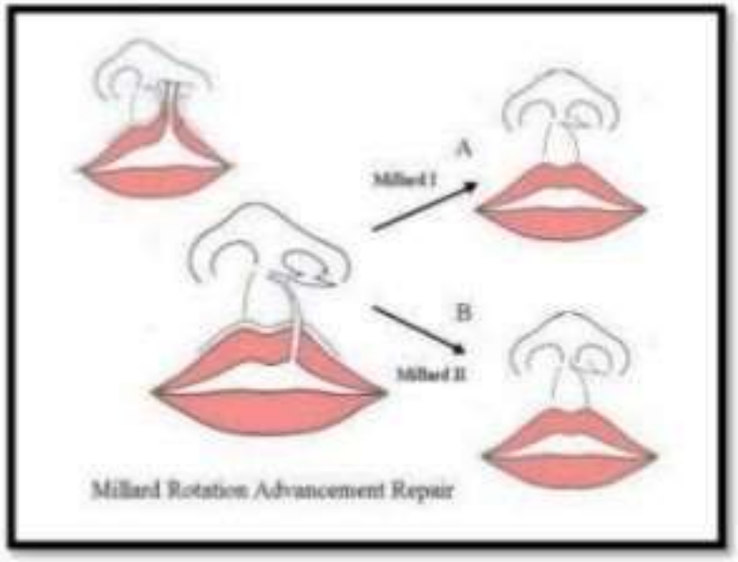
The basic premise of the repair is to create a three layered closure of skin, muscle, and mucosa that approximates normal tissue and excises hypoplastic tissue at the cleft margins.

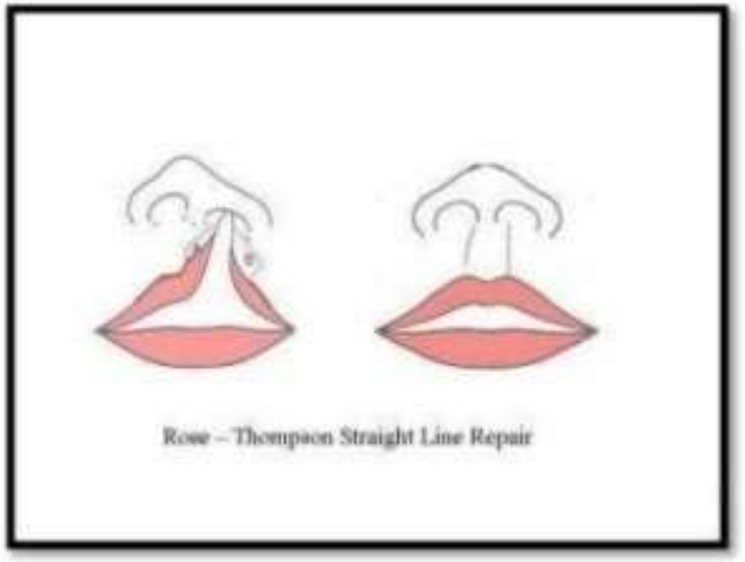
Critical in the process is the reconstruction of the orbicularis oris musculature into a continuous sphincter.

Techniques:

1. Tennison – Randall (Z-Plasty)
2. Millards rotation advancement repair
3. Rose – Thompson straight line repair
4. McComb`s techniques

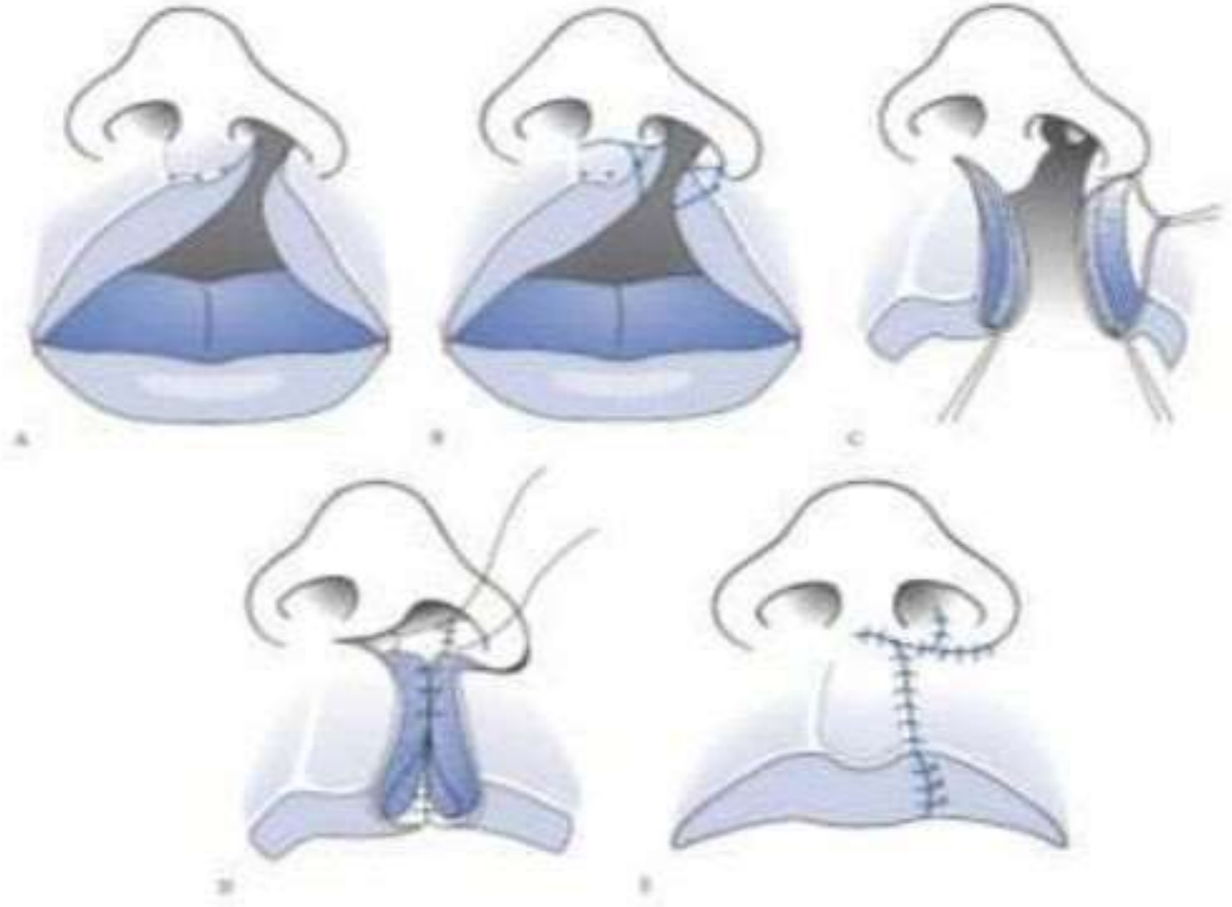






McComb`s techniques

McComb`s technique that has become popular, consisting of dissecting the lower lateral cartilages free from the alar base and the surrounding attachments through an alar crease incision. This allows the nose to be bolstered and/or stented from within the nostril to improve symmetry.



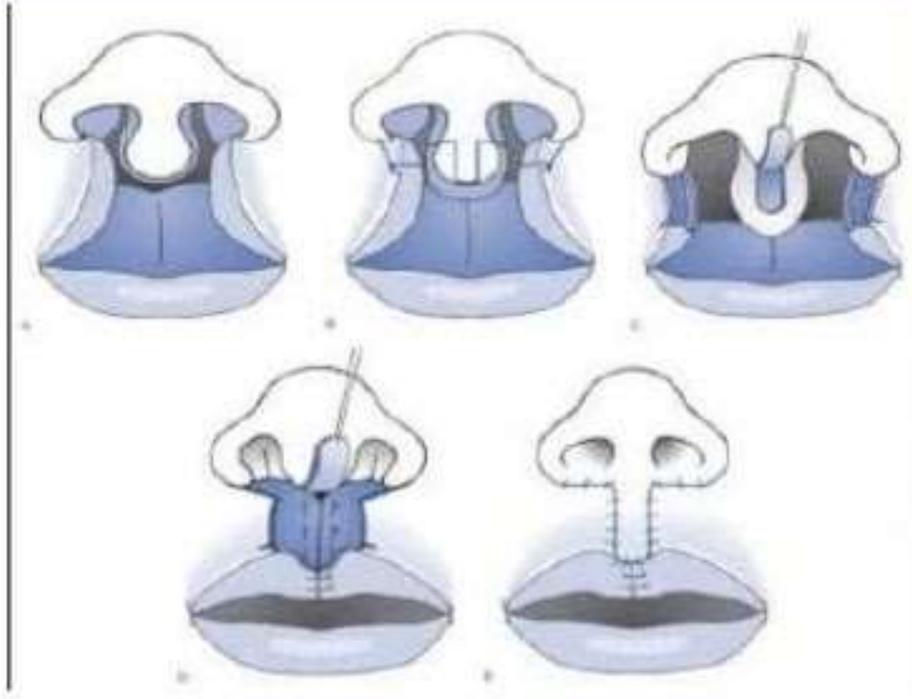
2- Bilateral Lip Repair

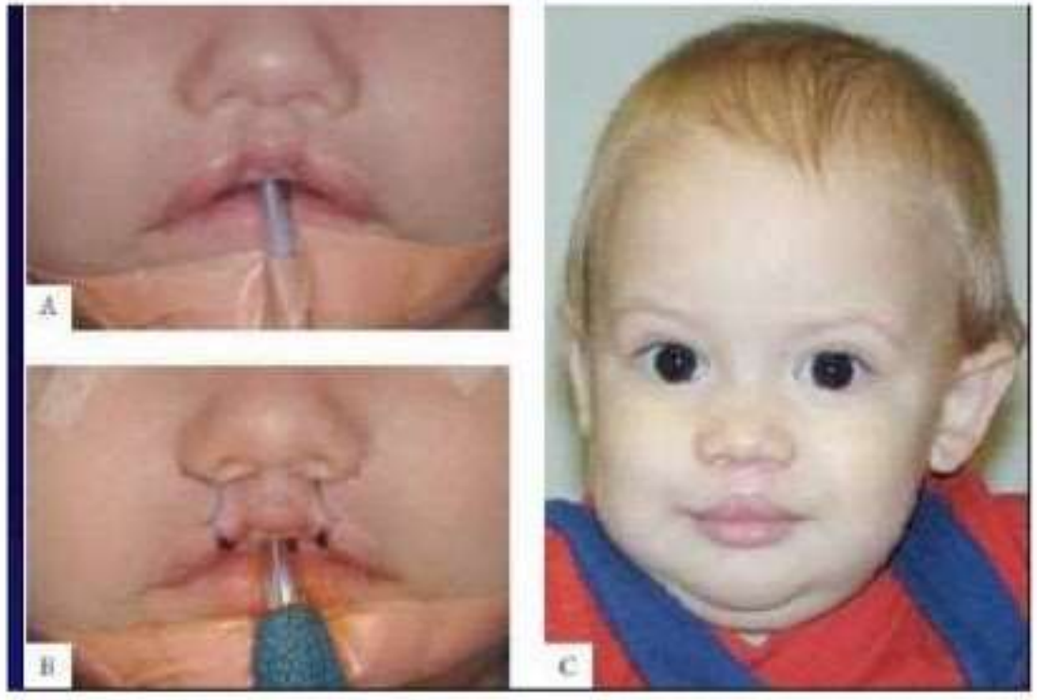
Bilateral cleft lip repair can be one of the most challenging technical procedures performed in children with clefts.

The lack of quality tissue present and the widely displaced segments are major challenges to achieving exceptional results, but superior technique and adequate mobilization of the tissue flaps usually yields excellent esthetic results.

Bilateral Complete

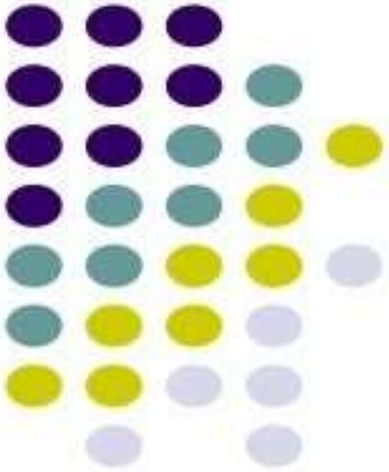
2- Bilateral Lip Repair





The bilateral cleft of the lip and maxilla shown here is complete and highlights the hypoplastic tissue along the cleft edges.

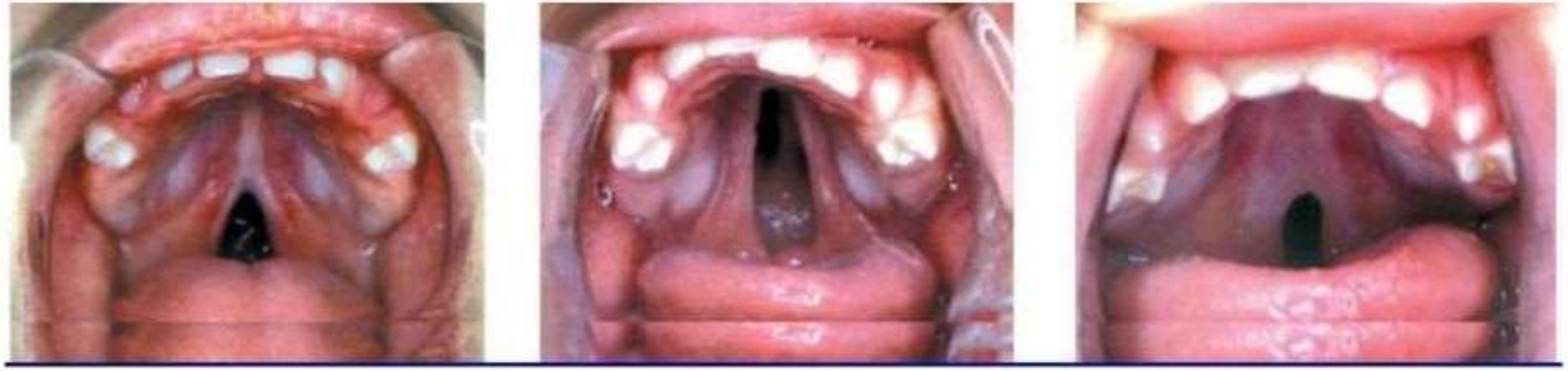
CLEFT PALATE



Cleft Palate Repair

two main goals of cleft palate repair during infancy:

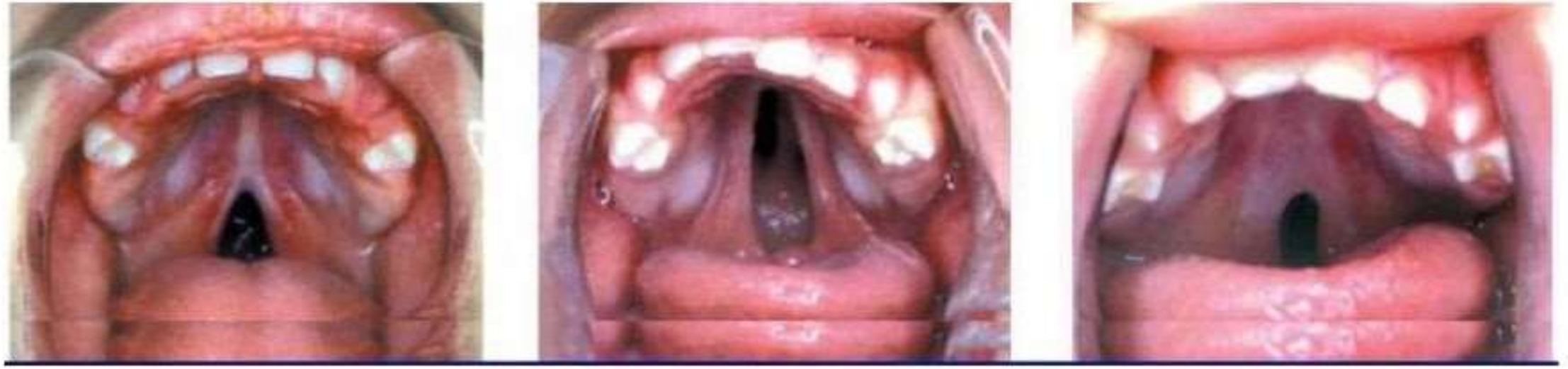
1. The water-tight closure of the entire oronasal communication involving the hard and soft palate.
2. The repair of the musculature within the soft palate that is critical for normal creation of speech.



Cleft Palate Repair

- The exact timing of repair of a palate cleft is controversial. Generally the velum must be closed prior to the development of speech sounds that require an intact palate.

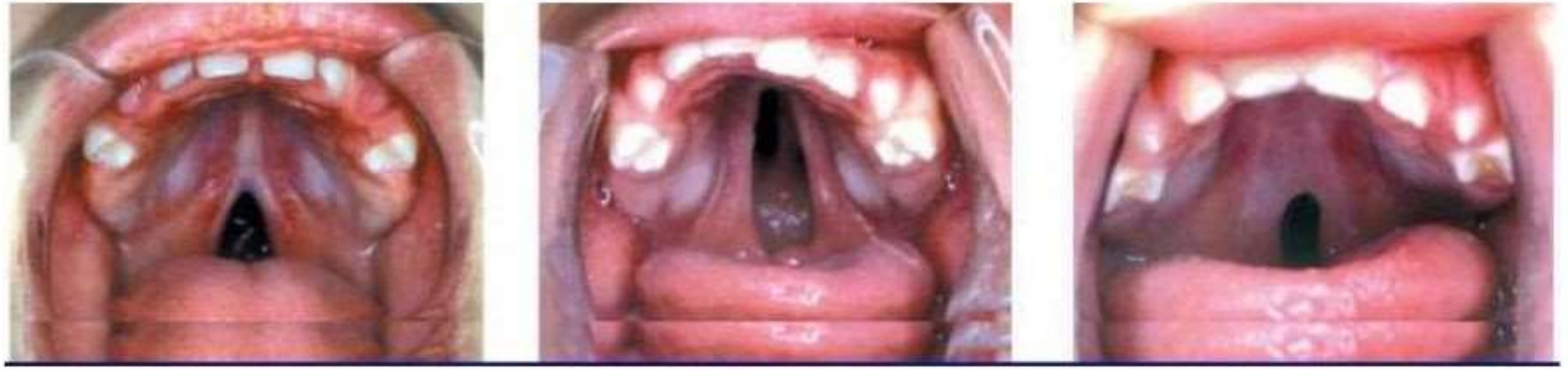
- Average this level of speech production is observed by about 18 months of age in the normally developing child.



Cleft Palate Repair

- If the repair is completed after this time, compensatory speech articulations may result.

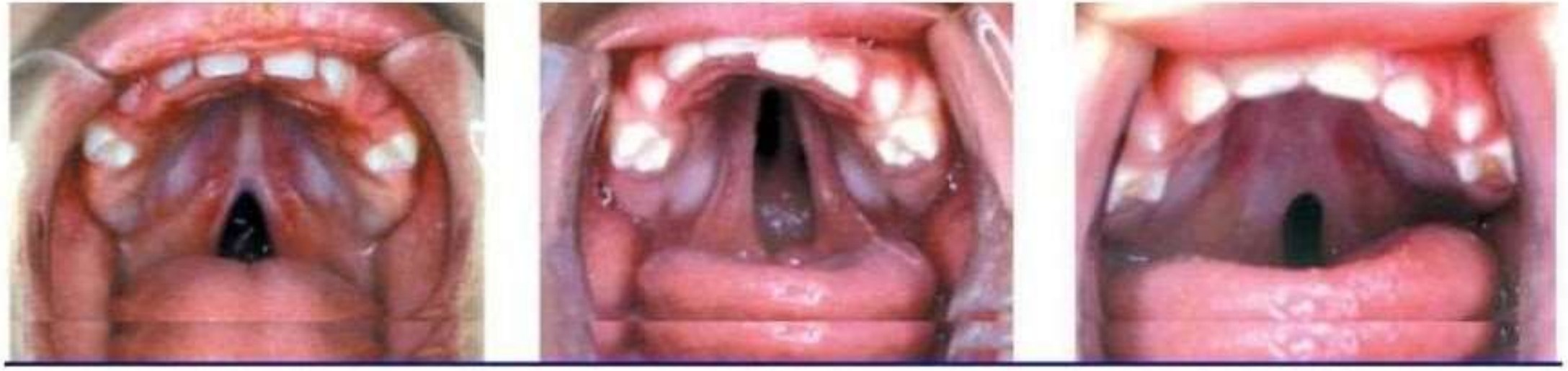
- Repair completed prior to this time allows for the intact velum to close effectively, appropriately separating the nasopharynx from the orophayrynx during certain speech sounds.



Cleft Palate Repair

- When repair of the palate is performed between 9 and 18 months of age, the incidence of associated growth restriction affecting the maxillary development is approximately 25%.

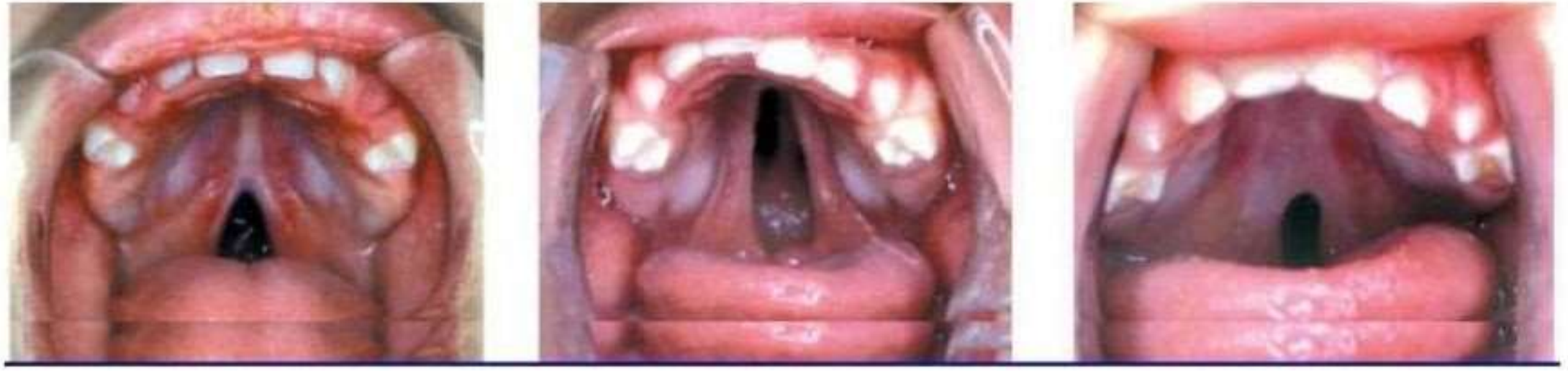
- Cleft palate reconstruction requires the mobilization of multilayered flaps to reconstruct the defect due to the failure of fusion of the palatal shelves.



Cleft Palate Repair

- surgeon must also reconstruct the musculature of the velopharyngeal mechanism.

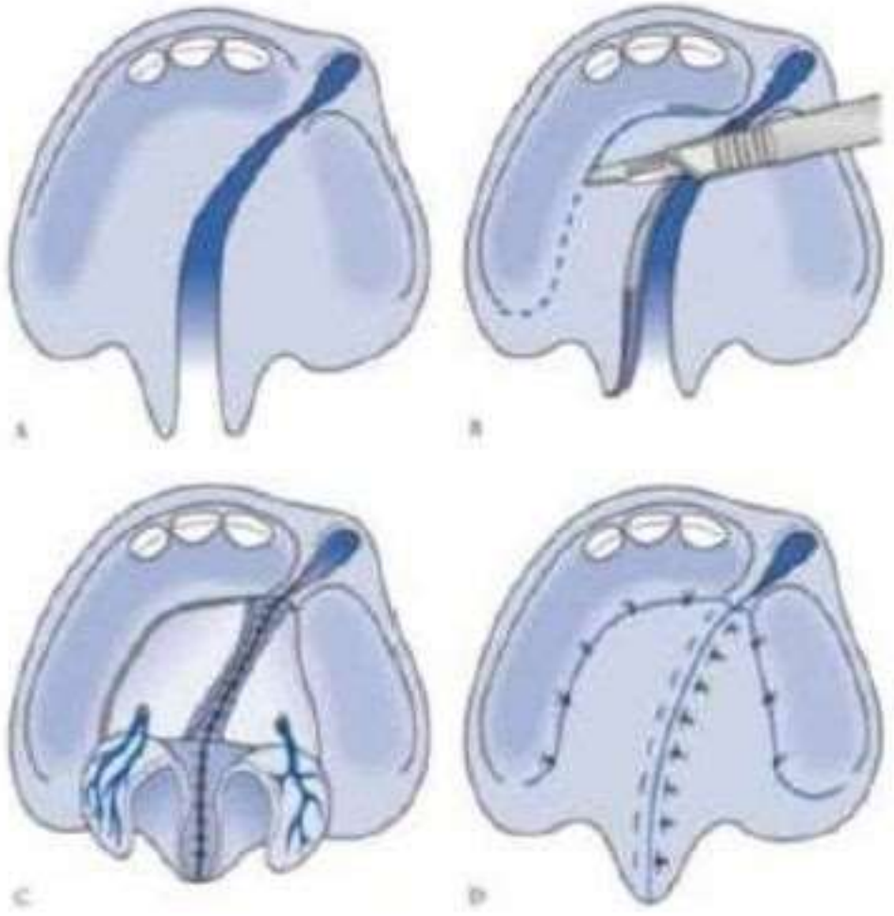
- The musculature of the levator palatini is abnormally inserted on the posterior aspect of the hard palate and therefore must be disinserted and reconstructed in the midline.



Bardach two-flap palatoplasty

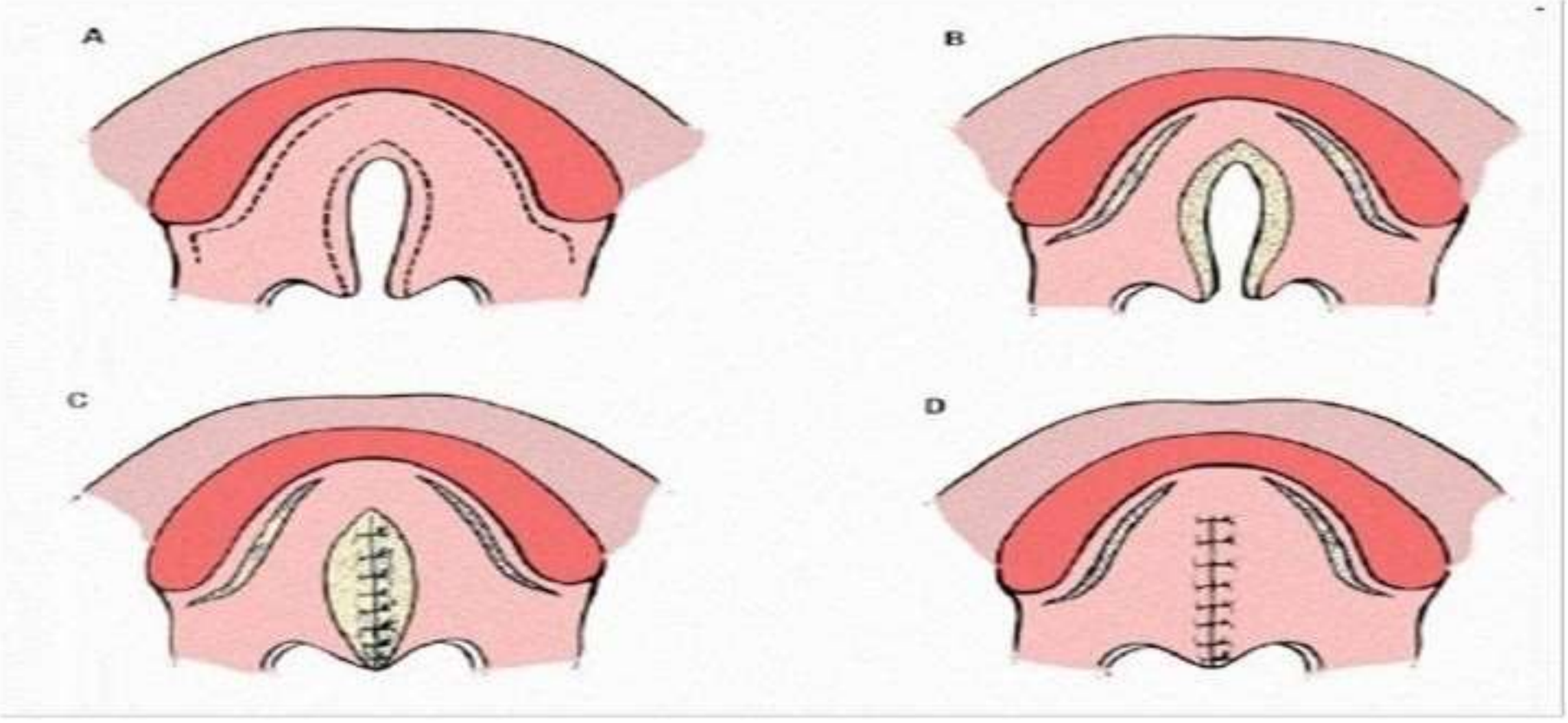
Uses two large full thickness layered dissection and brought to the midline for closure

A, Unilateral cleft of the primary and secondary palates, from the anterior vestibule to the uvula.
B, This tech.requires two large full-thickness mucoperiosteal flaps to be elevated from each palate shelf. The anterior of the cleft is not reconstructed until the mixed dentition stage.
C, layered closure is performed palatoplasty by reapproximating the nasal mucosa.
D, Once the nasal mucosa and musculature of the soft palate are approximated, the oral mucosa is closed in the midline



THE VON LANGENBECK TECHNIQUE

The von Langenbeck technique is similar to the Bardach palatoplasty but preserves an anterior pedicle for increased blood supply to the flaps.



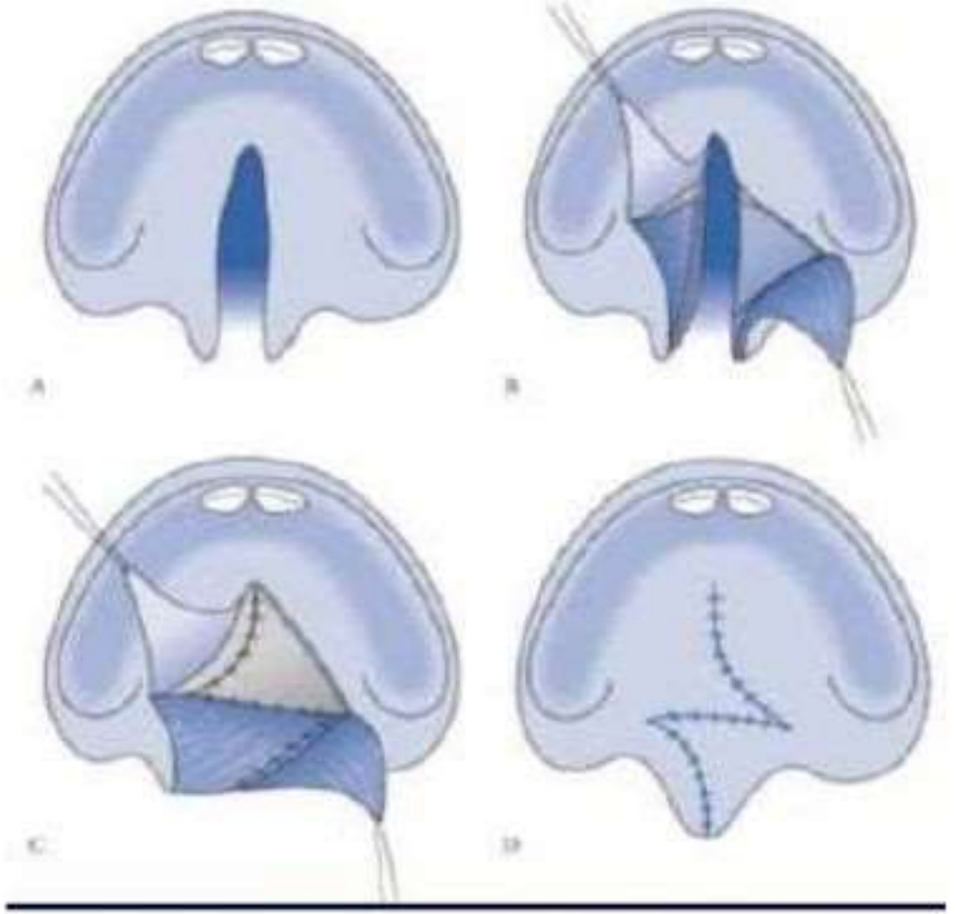
Furlow double-opposing Z plasty

Common technique attempts to lengthen the palate by taking advantage of a Z-plasty technique on both nasal mucosa and oral mucosa.

A, complete cleft of the secondary palate (both hard and soft). **B,** Z-plasty technique requires that separate Z-plasty flaps be developed on the oral and then nasal side.

C, The flaps are then transposed to theoretically lengthen the soft palate. A nasal side closure is completed in the standard fashion anterior to the junction of the hard and soft palate.

D, The oral side flaps are then transposed and closed in a similar fashion completing the palate closure.



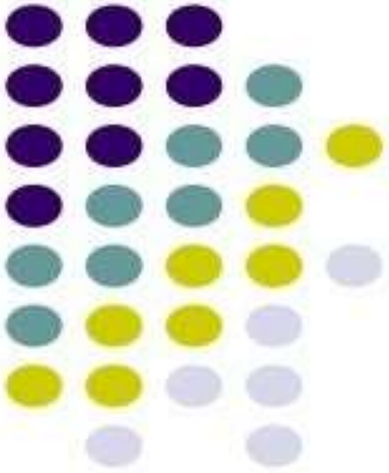
PHARYNGEAL FLAP

In very wide clefts, we used pharyngeal flap at the primary palatoplasty procedure to assist in closure.

The disadvantages of the pharyngeal flap are:

1. Bleeding.
2. Snoring.
3. Obstructive sleep apnea.
4. Hyponasality.

Secondary Cleft Palate Surgery for Management of Velopharyngeal Dysfunction



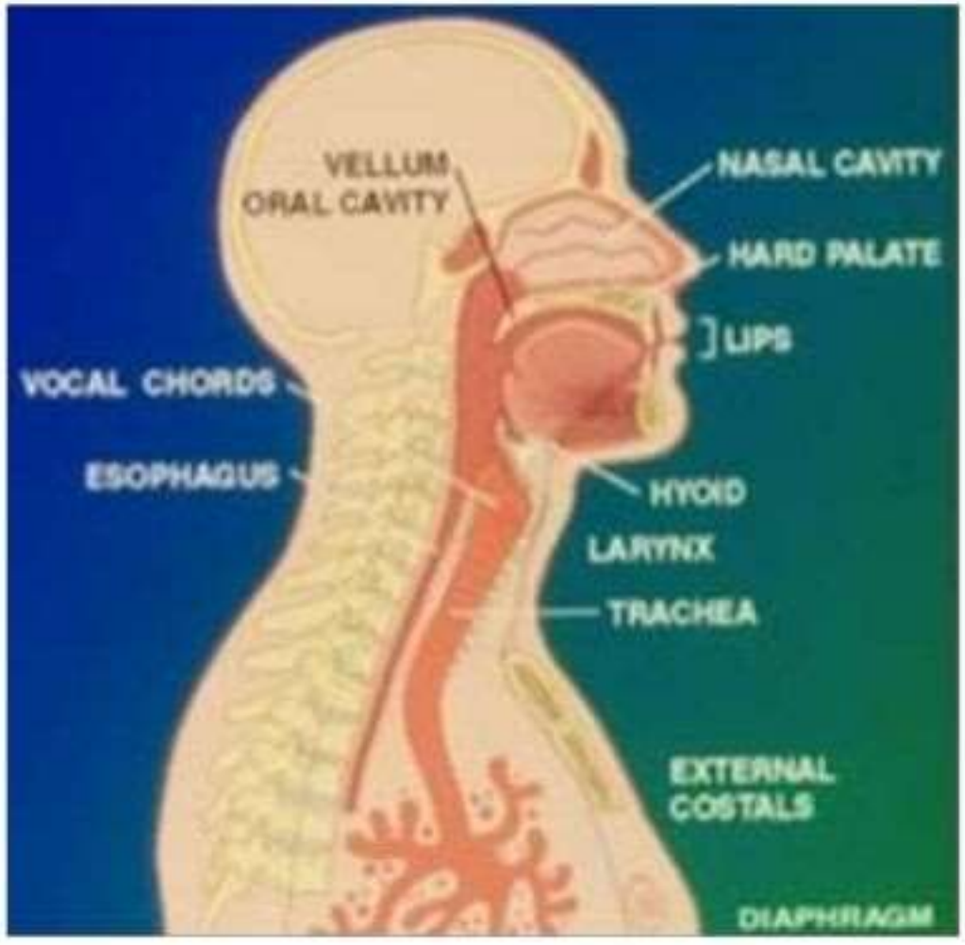
VELOPHARYNGEAL DYSFUNCTION

Introduction

The combination of the soft palate and pharyngeal wall musculature form the velopharyngeal (VP) mechanism.

The VP mechanism functions as:

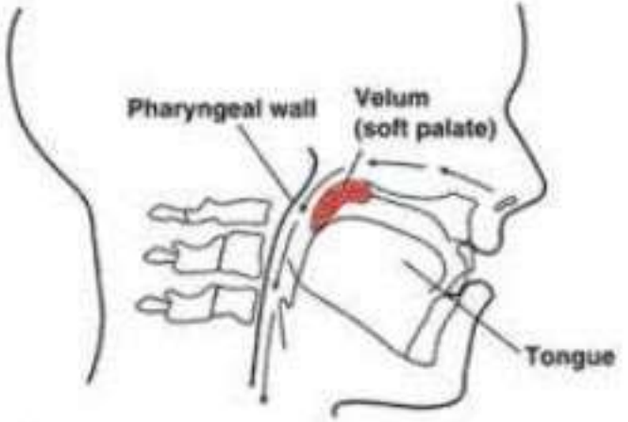
- sphincter valve to regulate airflow between the oral and nasal cavities.
- create a combination of orally based and nasally based sounds.



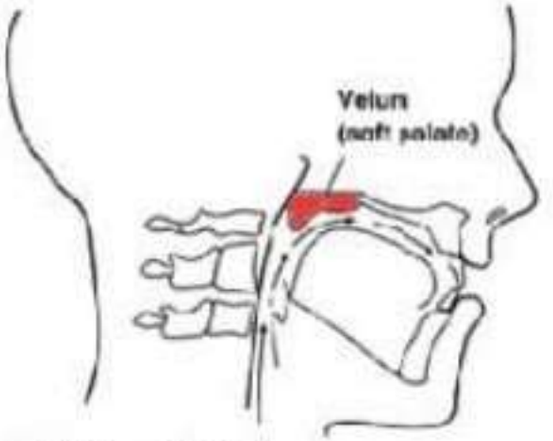
VELOPHARYNGEAL DYSFUNCTION

Clefting of the secondary palate causes:

- division of the musculature of the velum into separate muscle bellies.
- abnormal insertions of the muscles along the posterior edge of the hard palate.



Velum - normal position



Velum - raised

Velopharyngeal insufficiency (VPI)

Definition

Inadequate closure of the nasopharyngeal airway port during speech production.

Etiology

- Inadequate surgical repair of the musculature.
- Congenital defects.

Diagnosis

- With the use of videofluoroscopy and nasopharyngoscopy.
- Videofluoroscopy are used to radiographically examine the upper airway with the aid of an oral contrast material.
- Nasopharyngoscopy allows for direct visualization of the upper airway and specifically the VP mechanism from the nasopharynx.

Velopharyngeal insufficiency (VPI)

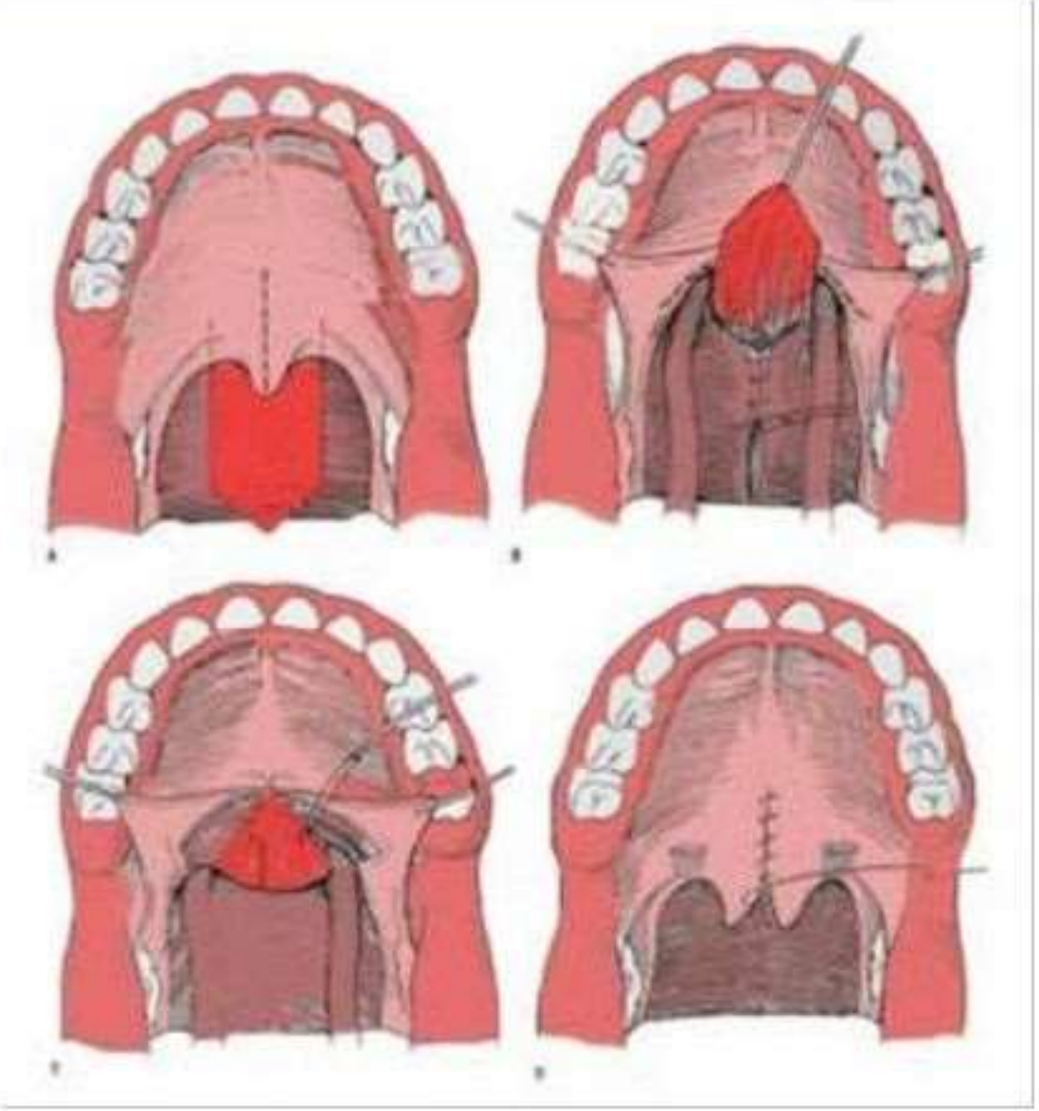
Timing for Surgery

- VPI causes hyper nasal speech.
- Ranging from 2.5 to 5 years of age.

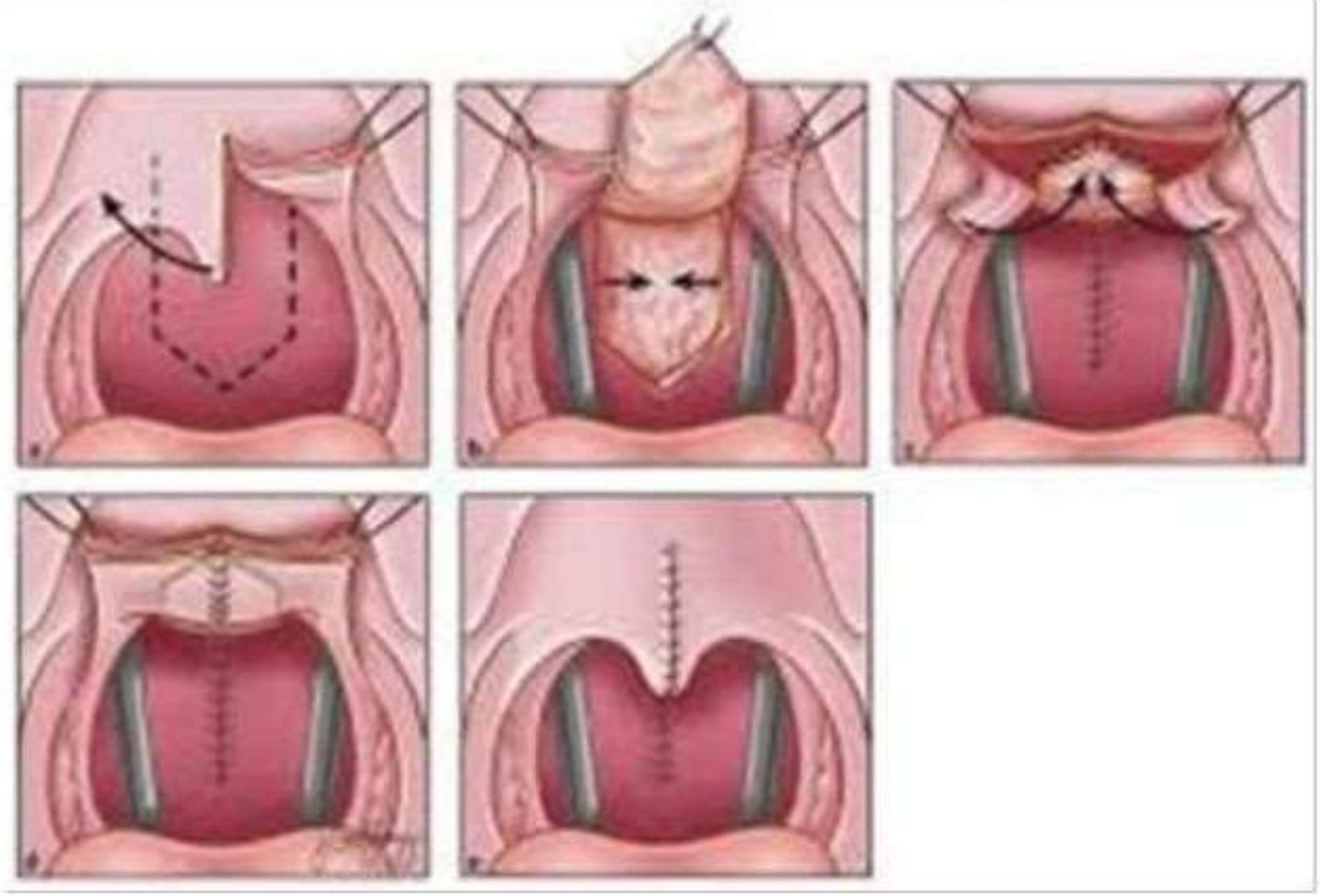
Operative Techniques

1. The superiorly based pharyngeal flap.
2. The sphincter pharyngoplasty.
3. The dynamic sphincter pharyngoplasty

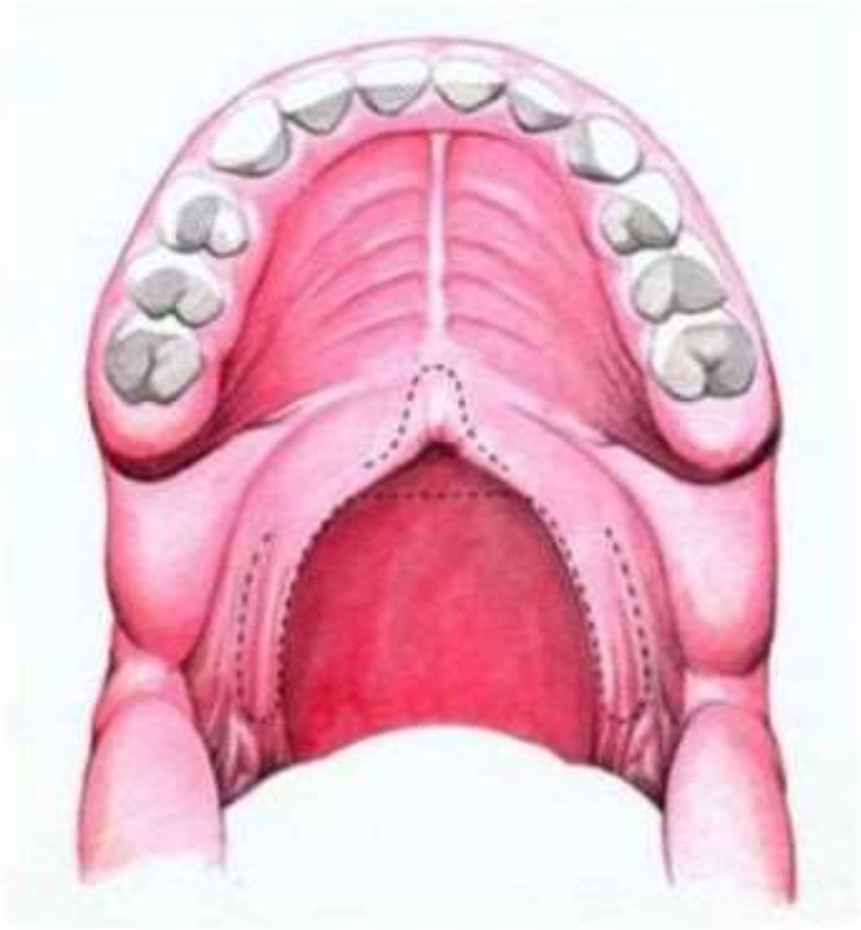
1. The superiorly based pharyngeal flap



Inferiorly based pharyngeal flaps

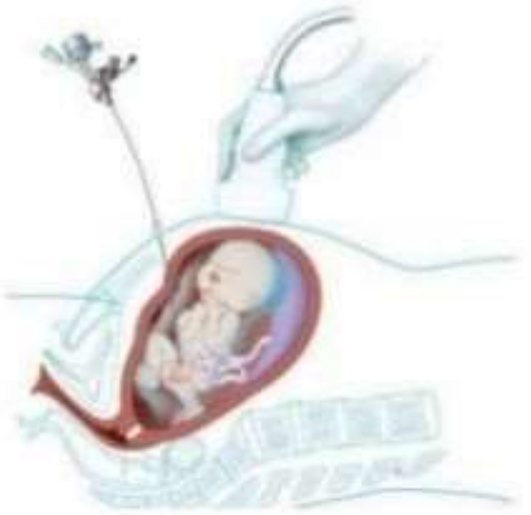


The dynamic sphincter pharyngoplasty



RECENT ADVANCES

- Fetal surgery – done in intrauterine life (prior to 20 weeks)
- Non-life threatening defects like cleft lip, cleft palate, Pierre Robin syndrome, Treacher-Collins syndrome, craniofacial microsomia
- Open fetal surgery
- Feto-endoscopic approach




CLEFT
THANK YOU
THANK YOU