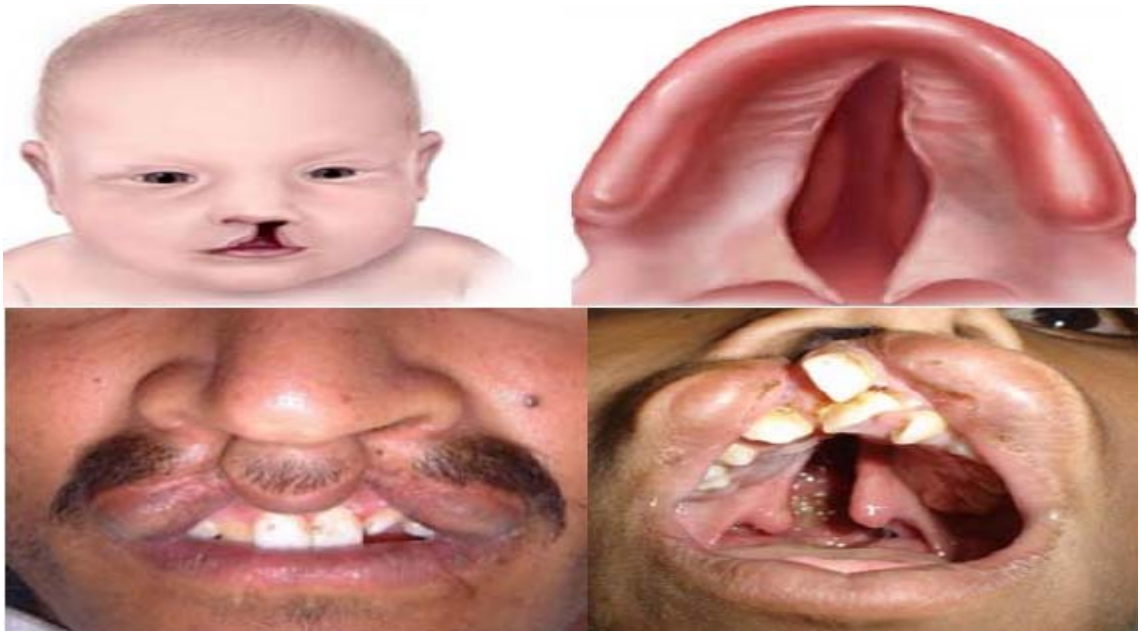
Cleft Lip and Palate



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Introduction

Cleft lip and palate is a common congenital anomaly. Clefting is associated with many problems including cosmetic deformities, dental abnormalities, speech, swallowing and growth difficulties. Proper management depends upon collaboration of many medical specialties to restore anatomical and physiological normality and prevent or minimize complications

Epidemiology

Cleft lip and palate is the most common congenital anomaly involving the head and neck region while it comes second to the clubfoot deformity. The incidence of cleft anomaly differs according to the ethnicity, occurring most often among Asians, Latinos and Native Americans (1 in 500), while it is 1 in 700 in European ethnicity and the highest incidence is among persons of African ethnicity (1 in 1000).

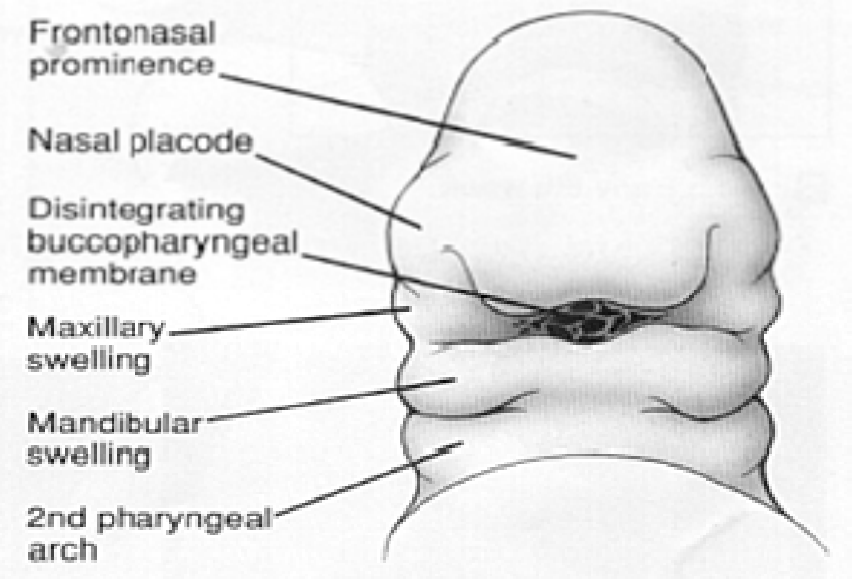
Cleft lip occurs more frequently in males (male to female ratio is about 2:1), with the deformity is usually more severe in male, while isolated cleft palate is more likely to occur in females.

Half of cleft patients have combined cleft lip and palate, while 30% of cases have cleft palate and the remaining 20% of cleft children have cleft lip with normal palate.

Cleft lip can be unilateral or bilateral and may range from a small notch in the upper lip to a complete separation of lip extending to the floor of the nose. Unilateral cleft lip occurs commonly on the left side (left to right ratio is 2:1). Bilateral cleft lip is much less common than unilateral cleft lip (ratio is 9:1).

Embryology of Cleft lip and Palate

The majority of facial development occurs from the 4rd through 12th week after conception and involves ectoderm and mesenchyme in the region of the neural plate. The nose, lips and palate are derived embryologically from 5 embryonic processes: (1) the median frontonasal process, (2) the paired maxillary processes and (3) the paired mandibular processes.



Cleft lip results from failure of one or both of the maxillary prominences to fuse with the medial nasal prominences, which normally occurs by the end of the 4th week of gestation.
a. Unilateral cleft lip results from failure of the maxillary prominence to merge with medial nasal prominence on the affected side.
b. Bilateral cleft lip results from failure the maxillary prominences to merge with the medial nasal prominence on both sides
c. Median cleft lip results from failure of the two medial nasal prominences to merge together from both sides.

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The palate is a result of the fusion of the 2 lateral palatine processes that come from the maxillary processes (secondary palate) with the median palatine process (Premaxilla) that arises from the frontonasal process (primary palate). Failure of this fusion results in cleft.

There are several factors contribute to the development of cleft lip and/or palate in a child. These may be genetic or environmental factors.

Chromosomal abnormalities

- Pierre Robin sequence
- Treacher Collins syndrome
- Trisomy 13,
- Trisomy 18
- Trisomy 21”Down syndrome”
- Velocardiofacial syndrome

Environmental Factors

- Alcohol, Smoking, Hypoxia
- Viral infections (rubella)
- Teratogens (steroids, anticonvulsants, phenytoin, alcohol)
- Folic acid deficiency
- Exposure to ionizing radiation
- Intrauterine bands or adhesions

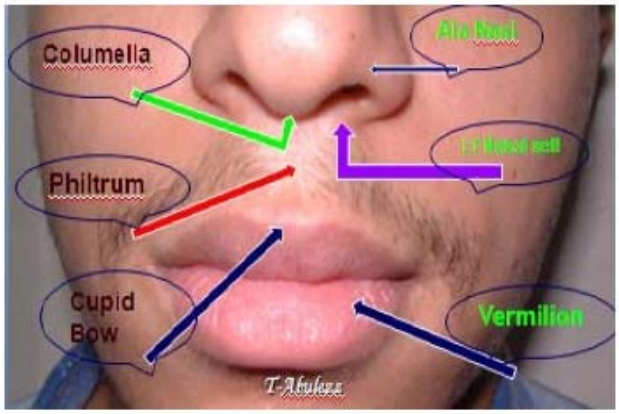
Anatomy of the Cleft Deformity

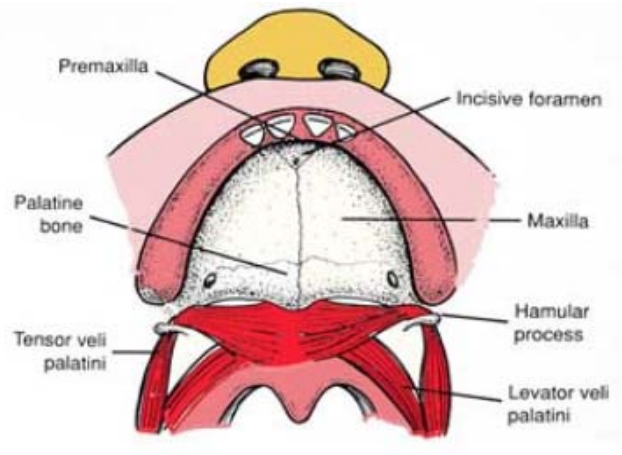
Patho-anatomy of the cleft Deformity

Glossary:

• **Alveolar ridge** (alveolus): the ridge in the upper jaw where the gums are and from which the teeth will erupt.
• **Primary Palate**: the front part of the hard palate anterior to Incisive Foramen, sometimes called **Premaxilla** (hard part) or **Prolabium** (soft part = central part of the upper lip).
• **Secondary Palate**: the back part of the hard palate and all of the soft palate
• **Hard palate**: the bony part of the roof of the mouth
• **Soft palate**: the soft, muscular part of the Palate

The degree of the deformity depends on the severity of the cleft lip or palate. Cleft is usually associated with many problems including cosmetic deformities, dental abnormalities, speech abnormalities, swallowing and growth difficulties.





A. Defects associated unilateral cleft lip.

1. Floor of nose communicates freely with the oral cavity.
2. The maxilla is hypoplastic on the cleft side.
3. The columella is displaced to the normal side.
4. The columella is shortened on the side of the cleft.
5. Nasal ala on cleft side is laterally and inferiorly displaced (flaring) and retrodisplaced.

6. The muscles of the obicularis oris do not form a complete sphincter but instead are directed upward parallel to the margins and terminate beneath the ala nasi (laterally) and the base of the columella (medially).
7. Inferior turbinate often hypertrophied on cleft side.

B. Defects associated with bilateral cleft lip.

1. Floor of nose is absent bilaterally.
2. Central portion of the alveolar arch is rotated forward and upward out of the area.
3. Obicularis oris muscle deformity similar to unilateral cleft lip in lateral segments (ie inserts into the ala nasi) but there are no muscle fibers in the prolabial (medial) segment.
4. Prolabium skin for lip is underdeveloped.
5. Central portion of the lip contains no lip muscle or lip vermillion.
6. The columella is short.
7. Nasal tip is widened and flattened.
8. Septum and nasal spine are forward in relation to the retrodisplaced alar bases.

C. Defects associated with the cleft palate.

1. Open roof of mouth communicating with nasal cavity.
2. Mucosal deficiency always present except in a submucous cleft palate.
3. The muscles of the soft palate are usually hypoplastic.
4. The soft palate muscles have an abnormal insertion into the posterior margin of the remaining bony palate rather than the midline raphe.

D. Facial skeleton defects associated with clefts.

1. Hypoplastic maxilla on cleft side.
2. Malalignment of alveolar arches.
3. With bilateral cleft, premaxilla often grossly deficient in bone.

E. Dentation abnormalities.

1. Supernumerary teeth 20%
2. Dystrophied teeth 30%
3. Congenitally missing teeth 50%
4. Malocclusion (nearly all patients)

Classification of Cleft lip and Palate

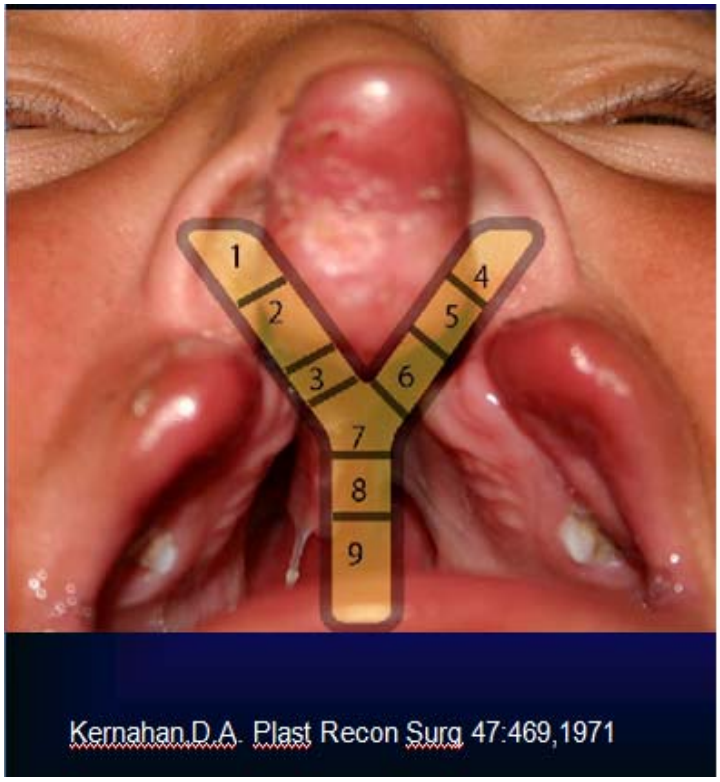
International Confederation of Plastic and Reconstructive Surgery classification

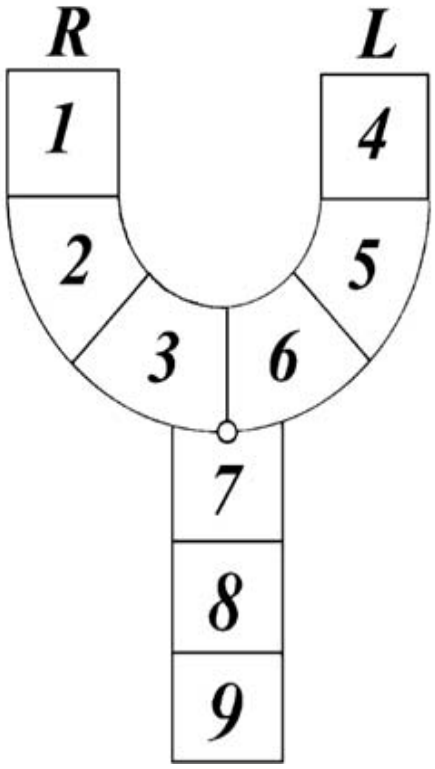
- Group I – Defects of the “Primary Palate” involving the lip or alveolus
- Group II – Clefts of the “Secondary Palate” involving the (hard palate, soft palate, or both.
- Group III –Combination of clefts involving the primary and secondary palates

Kernahan and Stark symbolic classification

This system provides a graphic classification scheme using a Y-configuration, which can be divided into 9 areas, as follows:

- Areas 1 and 4 – Lip (right and left)
- Areas 2 and 5 – Alveolus (right and left)
- Areas 3 and 6 – Palate between the alveolus and the incisive foramen (right and left)
- Areas 7 Hard palate
- and 8 – Soft palate
- Area 9 –Submucous cleft





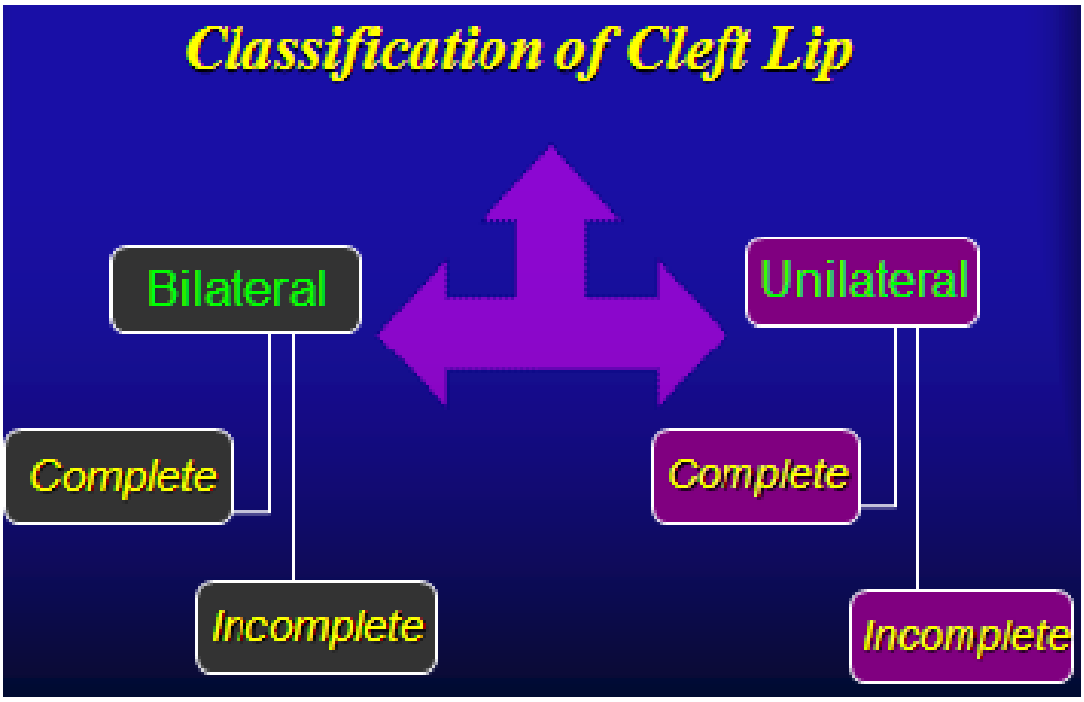
Veau classification

The Veau classification system divides the cleft lip and palate into 4 groups, which are as follows and illustrated in the image below:

- Group I – Defects of the soft palate only
- Group II – Defects involving the hard palate and soft palate
- Group III – Defects involving the soft palate to the alveolus, usually involving the lip
- Group IV – Complete bilateral clefts

Classification of cleft lip

- Cleft lip may be **unilateral** or **bilateral**.
- The cleft is described as **complete** when there is no separation between the oral opening and the nasal opening, while it is **incomplete** when there is any tissue separating the oral opening from the nasal sell.



Problems related to cleft lip and palate

Feeding

Newborn babies with cleft lip can usually breastfeed. Babies with a cleft palate may have more difficulty breastfeeding because they cannot create a negative pressure in their mouths. They may take in too much air during feeding (aerophagia), feed slowly or suffer from milk regurgitation through their noses. These problems if not corrected with cause malnutrition with failure to thrive of the baby.

Respiratory tract infection

Cleft palate children are usually more prone to upper respiratory tract infection. Factors predisposing to this are multiple; of which is the post feeding regurgitation of the acid-mixed milk through the nose which can induce aseptic inflammation of the nasal mucosa. When this child is having malnutrition with lowered immunity, the aseptic inflammation turns to bacterial infection that may spread towards the middle ear through the Austachian tube or descend downwards to the bronchi and lung causing repeated attacks of infection. Fever will worsen the nutritional deficiency as it raises the metabolic rate of the baby. There are many anatomical factors making cleft children more prone to repeated otitis media, of which

- Failure of the opening mechanism of Eustachian tube
- Eustachian tube is shorter than other children
- Deficient attachment of tensor veli palatini muscle
- Angle of entry into nasopharynx allows increased reflux of liquids

(Bluestone 1999)

Hearing

Children with cleft lip and palate are more likely to develop recurrent otitis media because the

Eustachian tube becomes distorted and malfunctioning due to the abnormal muscle arrangement of the cleft palate.

Speech

Normal development of the lips and palate are essential for a child to properly form sounds and speak clearly. Children with cleft lip usually develop speech normally. There may be some abnormal pronunciation of “labial” letters e.g. b, f and p. Children with cleft palate usually have “nasal” tone beside defective pronunciation of letter D, T, K, G, Q, N and M.

Dental problems

Sometimes a cleft involves the gums and jaw, affecting the proper growth of teeth and alignment of the jaw. In cleft alveolus, teeth near the cleft may be missing, or they may

come through at different angles to normal. Sometimes there are extra teeth. There may also be problems with the growth and direction of teeth

Management of Cleft problem

The management of children with cleft lip and palate presents many challenges but also many rewards. Affected individuals present a multiplicity of problems and effective management involves a multidisciplinary cleft team. This is a group of individuals from different specialist backgrounds who work closely together.

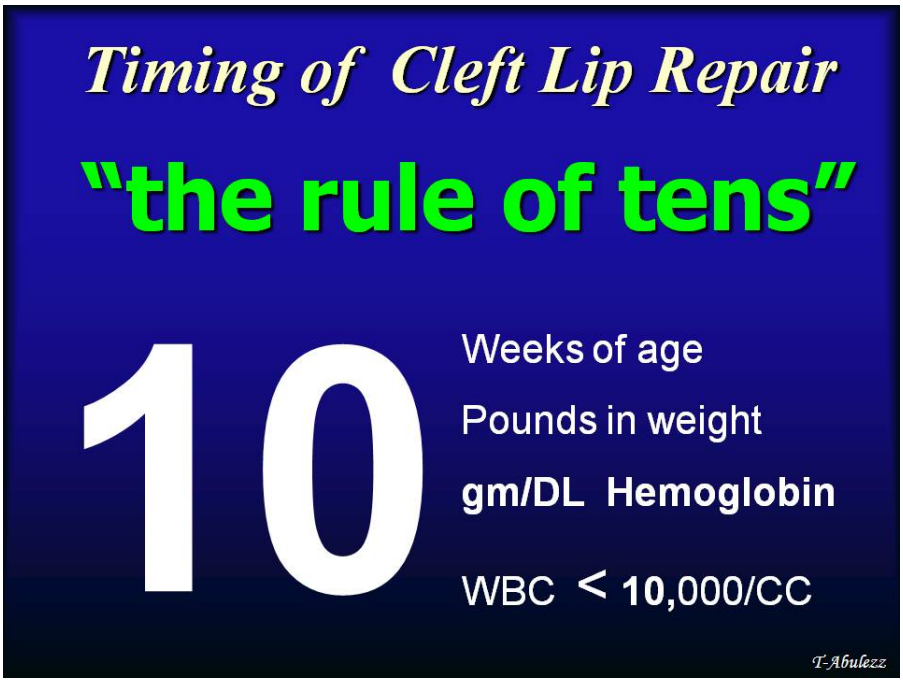
The main objective of the surgical interference is to achieve:

- Normal appearance
- Normal speech
- Normal dentition and facial growth

Surgical Repair of Cleft lip

Timing: The optimal timing of the surgical repair of cleft lip is before the eruption of the milk teeth. In most centers, the repair is performed at 3 months of age. Still there is some centers advocate surgery in the early neonatal period, with a theoretical benefit in the scar appearance and nasal cartilage adaptability, thus minimizing the nasal deformity.

The golden rule of ten still applies in most centers to minimize anesthetic risks: perform surgical repair of cleft lip when the child has a hemoglobin of 10 g, weight of 10 lb, and is aged 10 weeks and the leucocytic count is not more than 10,000 cells/cc.

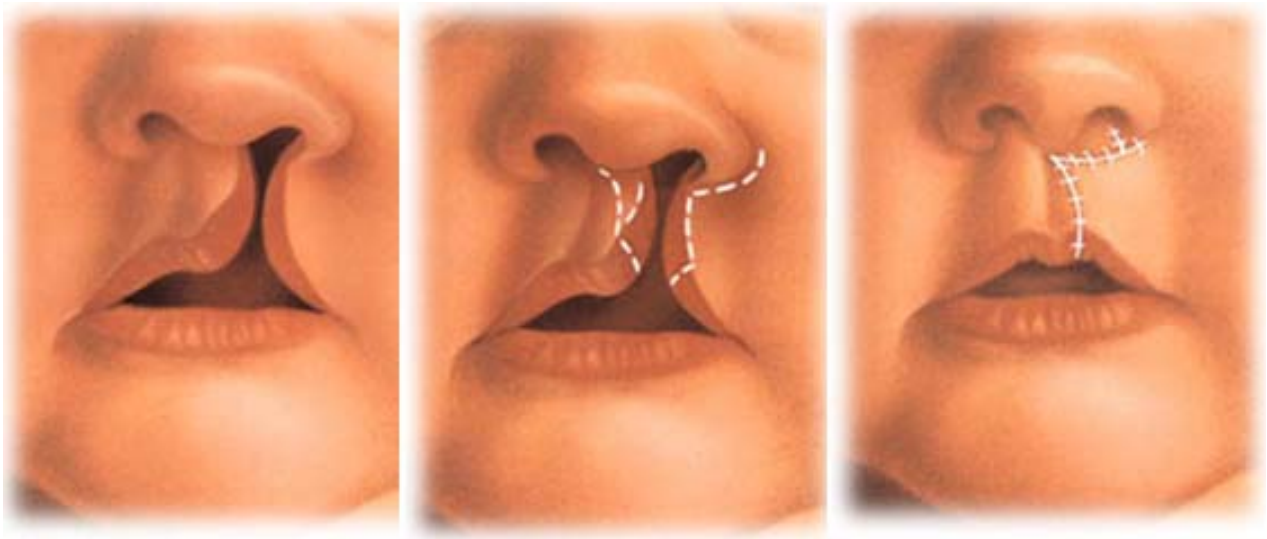


A preliminary lip adhesion is a procedure that may be used in selected patients preceding definitive lip repair, especially when the defect is wider than 1 cm. The nasal deformity is an integral part of the cleft lip. Depending on the severity, primary nasoplasty may be done at the time of the primary lip repair.

Repair of Unilateral Cleft Lip

Goal: Symmetric shaped nostrils, nasal sill, and alar bases; well defined philtral dimple and columns; natural appearing Cupid's bow; functional muscle repair

- Rotation-advancement (Millard)
- Triangular (Tennison-Randall)
- Quadrangular



Millard's Unilateral lip Repair

Repair of Bilateral Cleft Lip

Repair of a bilateral cleft lip is frequently more complex and challenging than repair of a unilateral cleft lip and it may be repaired in one or two stages.

Surgical Repair of cleft palate

The goal of surgery is to create a palate that works well for speech without interfering with the development of the growing facial skeleton.

Timing: Surgical repair of the cleft palate falls in controversial. Traditionally it is done in a single-stage repair involving closure with mucoperiosteal flaps between 9 and 18 months of age. However, recently a multistage approach is advocated but with the soft palate repaired early at the age of 3-6 months and the hard palate is closed after 2 years of age.

Surgical Techniques: The basic surgical techniques included the following: von Langenbeck, 3-flap (V-to-Y), and double reverse z-plasty (Furlow) palatoplasties.

Von Langenbeck technique

Bipedicle mucoperiosteal flaps of both the hard palate and the soft palate are used to repair the defect. After their elevation, the flaps are advanced medially to close the palatal cleft.

Schweckendiek "staged" technique

This is a staged repair. In this technique, the soft palate is first repaired when the patient is young (typically 3-4 mo), and this is followed with hard palate closure is usually performed when the patient is 18 months, but it may be delayed until the patient is 4-5 years. The vomer flap is usually useful with wide clefts.

Two-flap technique "Bardach Method"

The 2-flap technique involves 2 posteriorly based flaps that extend the length of the defect. The
flaps are rotated medially to close the defect.– Medial incisions made, which separate oral and nasal mucosa, Lateral incisions made at junction of palate and alveolar ridge, flaps are elevated preserving greater palatine artery, the velar muscles are dissected from posterior palate then close is made in 3 layers.

V-Y (Wardill-Kilner-Veau) "push-back" technique

Incisions are made along the free margins of the cleft and extended anteriorly from the apex of the cleft to where the canine teeth erupt. Mucoperiosteal flaps are elevated from the nasal and
oral surfaces of the bony palate. The palate is closed in 3 layers: nasal mucosa, the muscle layer,
and oral mucosa.

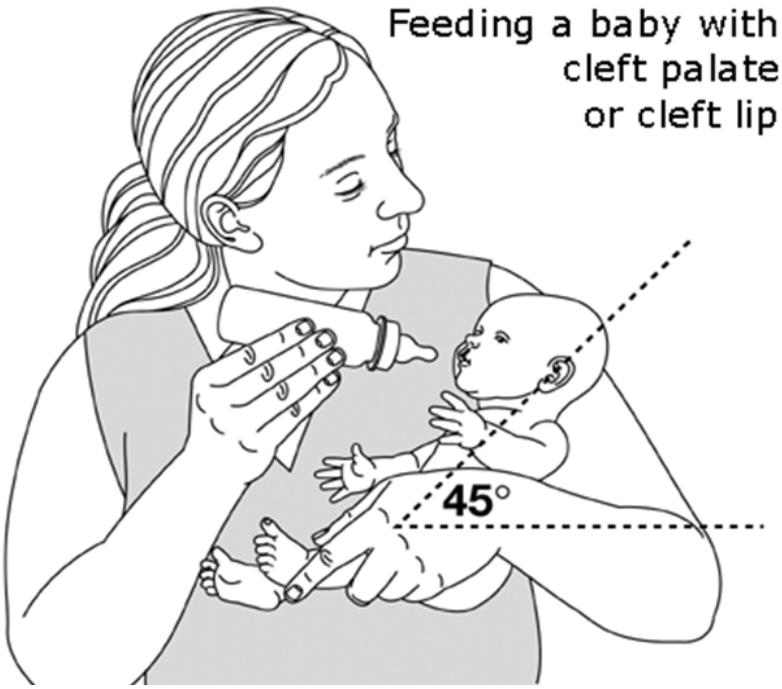
Double reverse (Furlow) Z-plasty

In 1986, Furlow described a technique to lengthen the velum and to create a functioning levator
muscle sling. This is a good method when the cleft is narrow or if a submucous cleft exists. The

technique involves opposing z-plasties of the mucosa and the musculature of the soft palate.

Feeding a baby with cleft palate

Most babies with a cleft palate are fed with a bottle. Various bottles and nipples can be used to assist with feeding. A cleft feeding nipple must have a wide opening enough to allow the formula to flow easily into the baby's month cavity, but it must not be so large as to cause choking. Several cleft palate feeders are available from several manufacturers. The most useful feeder usually has a long soft nipple with a crosscut hole attached to a flexible plastic bottle that can be squeezed to increase the flow of liquid.



With regard to positioning, a semi-upright position, as upright as possible, is best. This position helps prevent food and liquid from entering the nasal cavity. Upright positioning may also decrease Eustachian tube reflux, which may lead to otitis media.

The cleft palate baby is best fed with small frequent meals and the mother is instructed to keep the baby in the upright position and gently pat the upper back after the meal and not to put him/her to bed until the swallowed gas can be driven out.

Treating Otitis media

In children with a cleft palate, a grommet tube is usually inserted into the eardrum with the lip repair around the 3rd moth of life to prevent collection of middle ear effusion and damage by repeated infection.

Speech Therapy

Children with cleft palate should be evaluated by a speech-language pathologist before palate surgery (about 7-9 months) to determine the impact of the palate on speech and what to expect in speech development after surgery. After the cleft palate surgery, these children need speech therapy to develop normal articulation.

Dental and Orthodontic therapy

Most children with cleft lip and/or palate require orthodontic treatment as early as 6 years of age. This orthodontic treatment involves various phases, typically starting with palatal expansion done to normalize the width of the palate. Later, braces are put on to place the teeth in their proper position. Some kids with a cleft might be missing a permanent tooth, which can be replaced with a dental implant. Others may need some teeth to be removed to prevent overcrowding.