

PLASTIC & RECONSTRUCTIVE SURGERY

LEC 5

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اختصاص الجراحة التقويمية

CLEFT LIP AND CLEFT PALATE

Cleft lip and cleft palate are the second most frequently occurring of the major congenital anomalies they occur in 1:750--1:1000 ,club foot being the most common

There are two entities of this anomalies:

- **Cleft lip with or without palate (CL/P).**
- **Isolated cleft palate.**

cleft lip and palate CLP (46%),
isolated cleft palate (33%)
isolated cleft lip (21 %).

The majority of bilateral cleft lips (86%) and unilateral cleft lips (68%) are associated with a deft palate.

CLEFT LIP AND/OR PALATE (CL/P)

Embryology:

Primary Palate: The lip, nostril sill, alveolus, and hard palate **anterior** to the incisive foramen

Secondary Palate: The hard palate **posterior** to the incisive foramen and the soft palate

CL is caused by failure of union between medial nasal process and maxillary prominence at 4 to 6 weeks of fetal development.

Demographics:

- Cleft lip and/or palate (CL/P) has a variable racial distribution: Asians> Whites > Blacks.
- CL/P more frequent in males (2:1)
- Left unilateral CL/P is the most common: Left/right/bilateral: 6:3:1.
- **Risk factors**
 - Fetal exposure to substances including phenytoin, EtOH, steroids, phenobarbital,
 - diazepam, and isotretinoin
 - Maternal smoking
 - Parental age, especially advanced paternal age
 - Family history of clefting
- **Most cases are sporadic**, multifactorial
- <15% of all infants with cleft lip and palate have an associated syndrome:
 - Van der Woude's syndrome**
 - Trisomy 21 (Down syndrome).**

ISOLATED CLEFT PALATE (CPO)

Embryology: Interruption of Migration and fusion of the lateral palatal processes of the maxillary prominence between weeks 5 and 12 of gestation result in cleft palate.

Demographics:

- 0.5:1000 births, equal in all races
- More frequently in female: Male/female CPO: 1:2
- More than 40% of isolated cleft palates are part of malformation syndromes: **DiGeorge syndrome, Stickler syndrome**
- **ETIOLOGY**

Genetics

CP: Autosomal recessive with contributing genes

Environment

Smoking: Inconclusive but many studies have implicated its role

Teratogens: Alcohol, isotretinoin increases the risk

Folate and B6 may be protective

Primary retrognathia. Pierre Robin sequence

CLEFT LIP

Cleft lip anatomy:

1-Disruption of continuity, orientation and quality of orbicularis oris muscles

2-Cupid bow and lip rotated upward on both the lateral and medial –cleft side.

3-The alveolus and nostril floor are open in complete cleft lip.

4-The premaxilla is rotated and protruding especially in bilateral cleft lip.

5-Associated cleft lip nasal deformity e.g. flatten alar dome on affected side, shortened columella especially bilateral cases

CLASSIFICATION OF CLEFT LIP

A. Severity/extent

1. Microform CL (“forme fruste” or “minor cleft lip”

- a. Vermilion notching, scar-like line or depression, lip shortening
- b. ± Nasal deformity, usually mild
- c. Surgery may or may not be indicated based on severity

2. Incomplete CL

- a. Intact nasal sill (termed “Simonart band”)
- b. Intact alveolar ridge

3. Complete CL

- a. Clefting of the lip, nostril sill, and alveolus
- b. Wider than incomplete clefts with greater cleft nasal deformity

4. Complete CLP

- a. CL deformity is same as above

- b.** Includes CP (posterior to incisive foramen)

B. Unilateral versus bilateral

- 1. Unilateral CL**

- 2. Bilateral CL**

- a. Central prolabium and premaxilla**

- d. More likely to be complete, wide clefts.**

The critical factors for evaluating unilateral complete clefts are the position of the lesser and greater alveolar segments, the vertical height of the lateral lip element, and the degree of associated nasal deformity.

MANAGEMENT OF CLEFT LIP

A. Initial evaluation

1. Reassure parents and family
2. Explain surgical goals and timing of interventions
3. Evaluate for associated anomalies
4. Monitor for appropriate weight gain

B. Preoperative molding may be used to bring cleft segments together to minimize tension during repair

- **1-Elastic head cap:** used in first week of life especially for projected premaxilla in bilateral cleft lip.
- **2-Maxillary orthopedic:** for collapsed maxillary arches at (1-2) weeks of age.

C. Timing of CL repair

1. **3 months of age**, generally accepted
2. **“Rule of Tens”** (historical criteria) for suitability for surgery
 - a. 10 weeks old
 - b. 10 pounds
 - c. Hemoglobin 10 mg/dL

OPERATIVE TREATMENT

A. Goals of unilateral repair

1. Lengthen medial lip element
2. Reconstitute orbicularis oris
3. Restore Cupid's bow, aligning white roll and wet-dry vermillion
4. Correct nasal deformity (primary rhinoplasty).

Unilateral CL repair variations. Note most are Z-plasty-based reconstructions

1. **Straight-line repair (Rose-Thompson)**



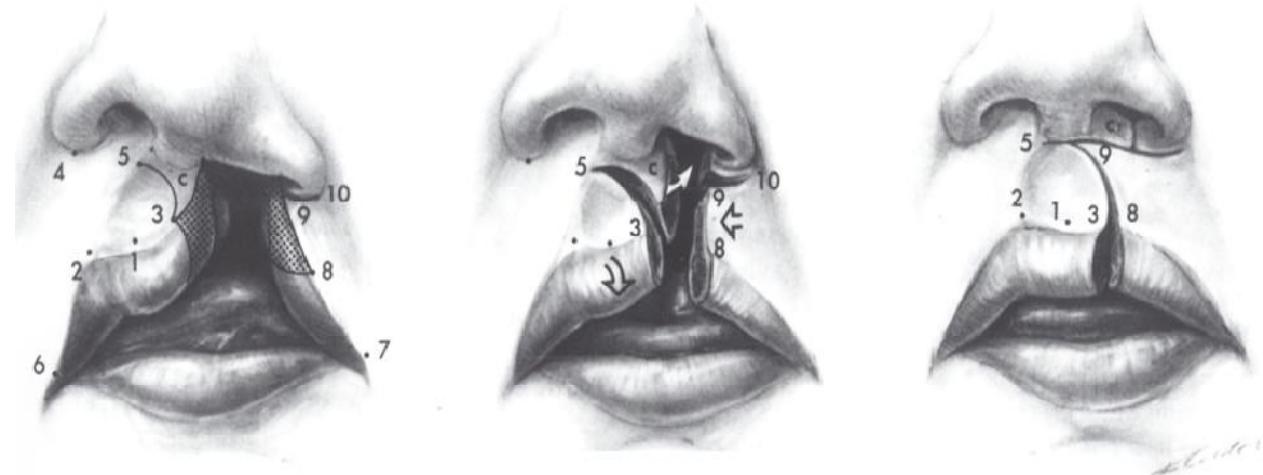
2. **Quadrangular flap (Le Mesurier)**



3. Triangular flap (Randall-Tennison, Trauner, and Skoog):



4. Rotation advancement (Millard): most commonly used repair in which the medial lip is rotated downward to fill the cleft defect, and lateral lip is advanced to fill the defect that occur in place of medial lip ,with small pennant shaped C-flap used either to create nasal sill or lengthened the columella.



Post operative care:

- 1) feeding with a catheter-tip syringe catheter for the first 10 days postoperatively to minimize strain on the muscle and skin sutures. Diet is advanced to full strength formula.
- 2) Suture line care consists of regular cleansing with half strength hydrogen peroxide followed with a light coating of antibiotic ointment.
- 3) Sutures are removed on or after the third postoperative day. After suture removal, taping and silicone scar gel is encouraged

cleft palate

Anatomy:

1-Hard palate: it is the bony part consists of primary and secondary palate which separated by incisive foramen. It consist of :

- Premaxilla, extended to incisive foramen.
- Paired maxilla.
- Palatine bone

2-soft palate(velum): contains muscle involved in velopharyngeal closure which:

1-extrinsic muscles:

2-intrinsic muscle: musculus uvulae

- Levator palate muscle.
- Tensor veli palatini muscle.
- Palatopharyngeus
- Palatoglossus
- Salpingopharyngeus
- Superior constrictor

All are supplied by vagus and glossopharyngeal nerve except the tensor veli palatini which supply by trigeminal nerve.

Vascular and nerve supply of hard palate is through the greater palatine artery and nerves through the greater palatine foramen.

Secondary blood supply through the lesser palatine artery and nerve through lesser palatine foramen

Classification of cleft palate

1. **Bifid uvula only**
2. ***Submucous cleft triad:** Intact mucosa with aberrant musculature
 - a. **Bifid uvula**
 - b. **Hard palate notch (palpable on exam)**
 - c. **Zona pellucida: Pale midline mucosa due to diastasis of soft palate (velar) musculature**
3. **Cleft velum only (soft palate, Veau type I)**
4. **Cleft of velum and bony palate (soft and hard palate, Veau type II)**
5. **cleft lip and palate**
 - i. If unilateral complete, **Veau III**
 - ii. If bilateral complete, **Veau IV**

- *the majority of the patients with submucous cleft palate are asymptomatic, although approximately 15% will develop velopharyngeal insufficiency (VPI)*
- *The infant with isolated cleft palate is examined carefully for manifestations of the Pierre Robin sequence (micrognathia, glossoptosis, and airway obstruction). The micrognathia and associated glossoptosis Causes obstruction of the palatal shelves as they swing from a vertical to horizontal orientation during palate fusion resulting in the characteristic wide "horseshoe" cleft palate . If the Pierre Robin sequence is present, the majority of cases can be treated with positioning and anti-reflux medications. In more severe cases, treatment may include nasopharyngeal airway protection, gavage feedings, and apnea monitoring. A small percentage of Pierre Robin patients require surgical intervention such as tongue-lip adhesion, distraction lengthening of the mandible or tracheostomy. Because of airway concerns, palatoplasty may be delayed for several months in Pierre Robin patients compared with other cleft palate closures.*

Early Considerations in Cleft Palate Care:

FEEDING: the cleft prevents the child from developing adequate suction. In general, however, swallowing mechanisms are normal; therefore, if the milk or formula can be delivered to the back of the child's throat, the infant will feed effectively. Most patients require assistance through the use of nipples with large cross-cut fissures, squeezable bottles, and baby should hold in 45° degree to decrease regurgitation into the nose, and feeding should take longer time.

Breastfeeding is usually not successful, unless milk production is very abundant.

AIRWAY: Maintenance airway by prone position during sleeping specially in **Pierre Robin sequence**

MIDDLE EAR DISEASE: The abnormal attachment of the muscles of the soft palate in a cleft palate alters the tension on the pharyngeal drainage of the Eustachian canal, increasing the incidence of ear infections. Myringotomy and grommet tube placement is performed in the majority of infants at the time of either the lip repair or the palate repair to prevent the development of hearing abnormalities

ASSOCIATED DEFORMITIES: Associated deformity occurs in >40% of children with isolated cleft palate and <15% of children with cleft lip and palate.

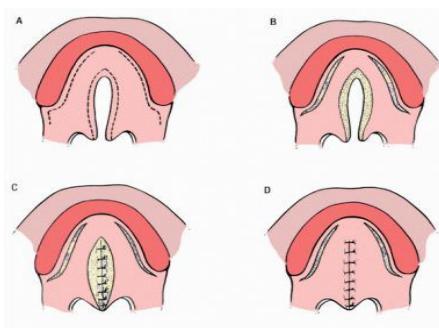
Surgical correction:

The optimum timing of cleft palate repair balances the benefit of normal velopharyngeal function to optimize speech development against the potential disadvantage of impaired facial growth secondary to early surgical trauma

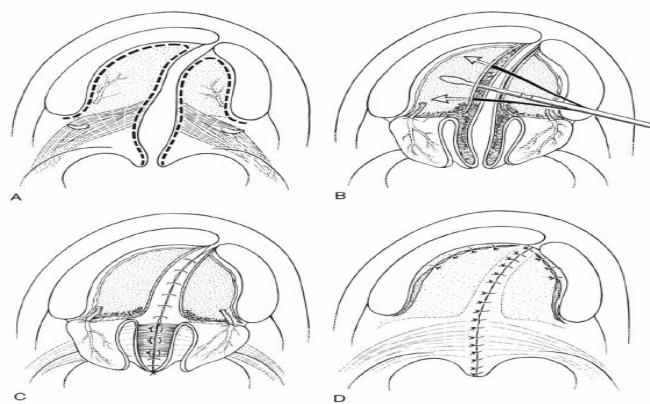
- **Time usually between ages of 11 to 12 months.**
- In children with airway issues, such as that associated with micrognathia of Pierre Robin sequence, the surgery can be delayed until age 14 to 18 months to allow further mandible growth

Surgical procedures are:

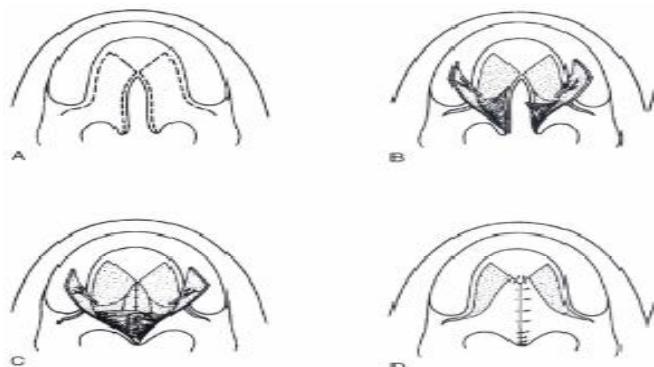
1-Bilateral bipedicled mucoperiosteal flap (Von Langenbeck repair): the flaps are elevated and then closed at midline, nasal mucosa first and oral mucosa last. This technique is not involving elongation of palate.



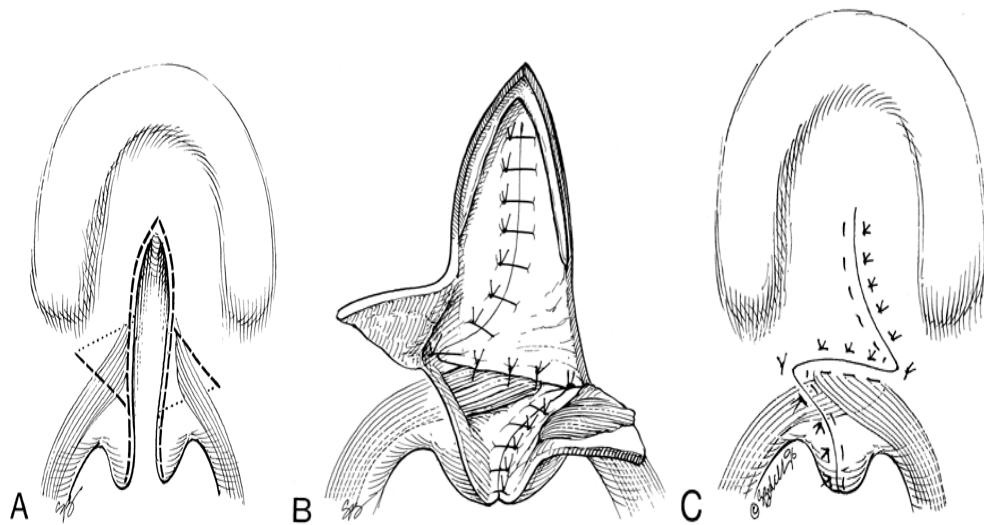
2-Two flap palatoplasty (Baradach): its modification of Langenbeck technique, where the incision extend along the alveolar margins to edge of cleft.making the flap entirely dependent on circulation from greater palatine artery. This technique is not involving elongation of palate.



3-V-Y elongation technique (Veau-Wardil-kilner). This technique is involving elongation of palate.



4-Furlow technique: soft palate elongation with double reversing Z-plasties



Postoperative care:

Elbow restraint are worn continually for 4-6 weeks.

The airway is observed and the child placed on oxygen monitor.

Patient are given liquid only diet for 3 week and child is transitioned to soft diet for an additional 3 weeks.

Surgical complication of cleft palate surgery:

- 1-Fistula: most common in wide bilateral cleft palate.**
- 2-Airway obstruction may occur secondary to postoperative bleeding.**
- 3-Midfacial hypoplasia**
- 4- Speech problem due to velopharyngeal incompetence**

Velopharyngeal incompetence:

Incomplete closure of soft palate against the posterior pharyngeal wall during speech , this lead to escape the air from oropharynx up through nasopharynx which lead to hypernasal speech.

Management:

- 1-Preoperative increasing the pharyngeal muscles strength by asking the baby to blow.**
- 2-Using the procedure which elongate the soft palate e.g. V-Y advancement and double opposite Z-plasty.**
- 3-Using mymucosal flap from posterior pharyngeal wall that suture to posterior soft plate.**
- 4- posterior pharyngeal wall augmentation using autologus tissues or alloplastic materials to reduce the size of velopharyngeal orifice.**

Summary of management of cleft lip and palate by age:

AGE	Treatment
Prenatal -----	Prenatal imaging, diagnosis
Newborn-----	Feeding assessment, medical assessment, genetic counselling, treatment information
0-3mo -----	Presurgical orthopedics
3 mo -----	Primary cleft lip repair and tip rhinoplasty \pm gingivoperiosteoplasty
12 mo-----	Primary cleft palate repair with intra velar veloplasty \pm bilateral myringotomy and tubes
Diagnosis of ----- velopbaryngeal insufficiency (3-4 y)	Secondary palate lengthening or pharyngoplasty, speech obturator
School-age years -----	Treatment of secondary lip and nasal deformities
7-9 y (mixed dentition)---	Secondary alveolar bone graft
Puberty -----	Definitive open rhinoplasty
Skeletal maturity-----	Le Fort I \pm mandible orthognathic surgery