

CLEFT LIP/PALATE

KEY FIGURES:

Unilateral cleft lip
Bilateral cleft lip
Cleft palate

Cleft lip: preoperative
and postoperative
Pierre Robin syndrome

Also known as “harelip,” cleft lip with or without a cleft palate is the most common craniofacial birth defect. It is beyond the scope of this text to describe the specific surgical procedures used to correct this anomaly. In fact, the treatment of cleft lip/palate remains a challenging problem even for plastic and reconstructive surgeons.

All health care providers, especially those in rural settings, should have an understanding of basic background information so that they can educate parents and ensure that the child receives proper treatment.

Definitions

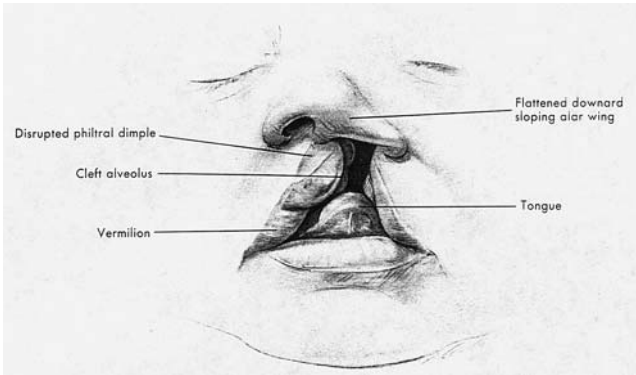
Cleft Lip

A cleft lip results when the developing tissues of the lip do not completely fuse. The lip is divided into two parts, and an incorrect alignment of the lip muscles (orbicularis oris) results.

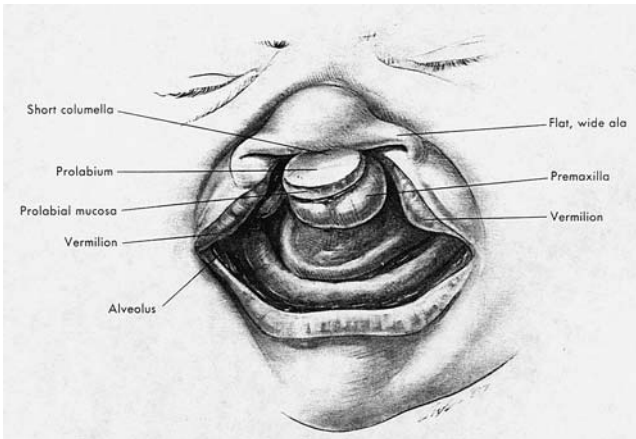
A cleft lip primarily involves the upper lip, although rare forms of facial clefting can involve the lower lip. The typical cleft lip often involves the nose, with resultant distortion of the nostrils and nasal sill.

Cleft Palate

The palate is essentially the roof of the mouth. It is composed of two parts, the hard palate and the soft palate. The teeth erupt in the anterior hard palate (called the alveolar ridge), and the posterior hard palate serves as the base of the nasal cavity.

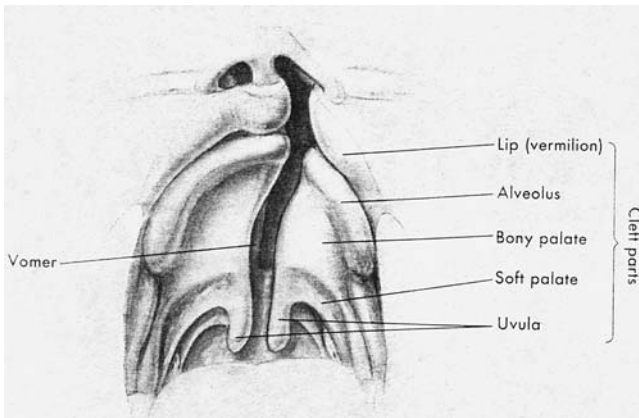


Unilateral cleft lip. The upper lip is twisted and shortened vertically. Cupid's bow is incomplete, the vermilion tapers cephalad, and the white line extends into the vestibule. In this case an alveolar cleft is present, creating a defect in the nasal floor. The alar rim is significantly distorted. (From Jurkiewicz MJ, et al (eds): *Plastic Surgery: Principles and Practice*. St. Louis, Mosby, 1990, with permission.)



Bilateral cleft lip. Severely shortened columella, wide flattened alae, and jutting, often rotated premaxilla are hallmarks of the deformity. (From Jurkiewicz MJ, et al (eds): *Plastic Surgery: Principles and Practice*. St. Louis, Mosby, 1990, with permission.)

The soft palate is the posterior portion of the roof of the mouth. The soft palate is mobile and is composed of several muscles important for normal speech and proper function of the eustachian tubes (associated with the middle ear).



Unilateral complete cleft lip and palate. (From Jurkiewicz MJ, et al (eds): *Plastic Surgery: Principles and Practice*. St. Louis, Mosby, 1990, with permis-

Presentation and Incidence

Presentation varies widely. The child may be born with a unilateral or bilateral cleft lip with a normal palate, a cleft palate (soft only or hard and soft) with a normal lip, or a unilateral/bilateral cleft lip with a cleft palate. The most common presentation is left-sided unilateral cleft lip with cleft palate. Male infants are affected more often than female infants.

The majority of affected infants are otherwise healthy and normal intellectually. However, there is a 25% incidence of additional anomalies, including neurologic and cardiac abnormalities as well as club foot. Evaluate the child closely so that other problems are not missed.

Cleft lip/palate is the most common congenital facial abnormality. The incidence in Caucasian populations is 1–1.5/1000 live births; in African and African-American populations, < 0.5/1000 live births; and in Asian and Hispanic populations, 2–3/1000 live births.

Embryologic Development and Etiology

During fetal development the lip and palate are formed during the first trimester (days 30–60 of gestation). A cleft develops when something interferes with the normal processes of fusion and mesodermal penetration of the frontonasal processes and maxillary processes of the face of the embryo. Essentially, instead of growing together to form a normal lip and palate, the embryonic tissues remain separate, causing the cleft to develop.

Parents without clefts who have one child with a cleft lip/palate have a 5% chance of having another child with a cleft lip/palate (compared with the usual 0.14% risk in parents with no family history of cleft). If both one parent and one child have a cleft or if two normal parents have two children with clefts, the likelihood of a cleft in another child increases to 15–20%. These data seem to suggest a genetic component.

The development of cleft lip/palate, however, probably is due to a combination of multiple factors—for example, folic acid deficiency, advanced maternal/paternal age, use of anticonvulsants (phenytoin or phenobarbital), alcohol intake, and possibly smoking. A viral etiology also has been suggested.

It is quite common for the parents of a child with a cleft to give a history that includes none of the above factors. In some rural cultures, a cleft palate is thought to represent a sign of evil or wrong-doing on the part of the family. Parents are often quite guilt-ridden, thinking that they did something to cause the defect. *Therefore, it is important to assure parents that they did nothing wrong to cause the child's abnormality.*

Immediate Concerns

Adequate Nutrition

The initial concern for an infant with cleft lip/palate is ingestion of adequate calories and fluids to maintain health and allow proper growth.

An infant's ability to suck is related to two factors: the ability of the external lips to perform the necessary sucking movements and the ability of the palate to allow the necessary build-up of pressure inside the mouth so that foodstuff can be propelled into the mouth.

Infants with cleft lip/palate have sufficient external lip muscle movements. Therefore, an isolated cleft lip usually does not interfere with the child's ability to suck, although it may take some practice not to lose a lot of milk out of the cleft lip defect.

In contrast, a child with a cleft palate with or without a cleft lip has difficulty sucking properly. The cleft in the palate impedes the proper build-up of pressure inside the mouth. For the same amount of effort, the infant with a cleft palate does not ingest as much milk as a normal infant. This increase in the work of feeding may lead to insufficient intake of calories for proper growth and health.

Helpful Strategies

The child should be fed with the head upright at about a 45° angle.

Usually bottle feeding is needed. If possible, use a nipple made for premature infants because it has a larger opening than normal. An alternative is to cut an X in the tip of a regular nipple to enlarge the opening. The larger-than-normal opening allows more liquid to flow out of the nipple with less suction.

A squeezable bottle also facilitates getting the milk into the infant's mouth.

If you are in an area with access to a pediatric orthodontist, an appliance can be made to fit into the infant's mouth and cover the cleft defect. This appliance, called an obturator, greatly improves the infant's ability to suck and ingest calories with less energy expenditure.

Do not be alarmed when milk comes out of the infant's nose. The palate serves to separate the nasal cavity from the oral cavity. The presence of a cleft in the palate removes this separation so that food and liquids easily pass from the mouth and come out of the nose.

Initial Corrective Operations

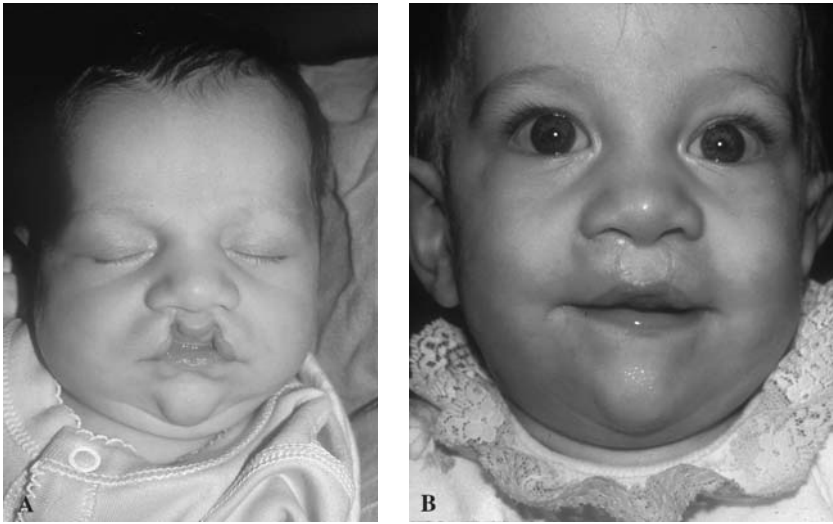
Correction of the cleft lip/palate deformity is not a life-preserving medical necessity. However, nontreatment often results from lack of access to specialists rather than a conscious decision to leave the child untreated.

A cleft lip/palate is a challenging deformity to repair. Usually, the lip is repaired at around 3 months of age. In full-term, healthy infants, however, repair may be undertaken at an earlier age.

In utero repair of the cleft lip defect is a subject of ongoing research. Clefts usually can be seen on prenatal ultrasound, and it has been shown that fetal tissues heal without the scars usually seen in infants and adults.

The palate is repaired at a separate operation, usually when the infant is around 12 months of age. In areas with limited access to specialists, however, the lip and palate can be repaired at the same time. In fact, even in the developed world where plastic surgeons are abundant, some authorities believe that simultaneous repair of the lip and palate may yield better results than separate repairs.

Often, if the infant has recurring ear infections, pressure-equalizing (PE) tubes are needed. Usually they are inserted at the time of palate repair.



Infant with an incomplete bilateral cleft lip. A, Preoperative appearance. B, Appearance after one operation.

Other Possible Operations

Unfortunately, even under the best of circumstances, further operations, available primarily to patients in the developed world, may be required for definitive treatment. The combination of cleft lip and cleft palate requires additional surgery more often than cleft lip or cleft palate alone.

Even if the palate is properly repaired, the child may have significant speech difficulties. Often they can be corrected with speech therapy, but approximately 20% of children with a repaired cleft palate require an additional operation to improve palatal function, usually between the ages of 4 and 6 years.

Irregularities of the lip often are noticeable even after repair. Some irregularities that are present when the child is 1–2 years of age are due to tight scarring and improve with time. Revisions usually are delayed until the child is 4–6 years of age.

In addition, significant deformity of the nostrils may require correction, usually when the child is 4–6 years of age. Surgery on the nose can be done at the same time as lip revisions.

Most clefts of the anterior hard palate (alveolar ridge) are repaired when the canine tooth on the side of the cleft begins to erupt, usually around age 7–8 years.

Optimally, a pediatric dentist or orthodontist should be continually involved with the care of children with cleft palate to get the teeth in as normal a position as possible.

If the maxilla (upper jaw) does not grow normally, corrective surgery may be indicated. The procedure, called a Le Fort I osteotomy, usually is delayed until the child is much older, often in the later teen years.

Table 1. Overview of Operations that May be Required for Cleft Lip/Palate

Approximate Age	Operation	Comments
3 mo	Cleft lip repair	Should be delayed for small children (< 10 lb or 4 kg) to decrease risks from anesthesia.
12 mo	Cleft palate repair	Can be delayed to 18 mo because of airway concerns, but should be done before infant starts to talk.
3–12 mo	PE tubes	Depending on number of ear infections, PE tubes can be placed at time of lip repair or palate repair.
4–6 yr	Improvement of palatal function	Improves child's speech; about 20% of children with cleft palate repair require additional surgery.
4–6 yr	Lip revision, minor nostril corrections	These procedures can be done at the same time.
7–8 yr	Repair of anterior (alveolar) hard palate	Usually done when canine tooth begins to erupt; can be done successfully in older child (10–12 yr old).
> 17–18 yr	Le Fort I osteotomy: upper jaw surgery	Maxilla usually does not grow normally in child born with cleft palate and may need to be cut and repositioned to correct relationship of upper jaw to lower jaw (mandible).
> 17–18 yr	Rhinoplasty; more complex nose surgery than performed before	Nose may need significant work to achieve more normal appearance and function. Procedure may include cartilage grafts, bone repositioning, and correction of deviated septum.

PE = pressure-equalizing.

Visiting Surgeon Programs

Skilled plastic surgeons and other health professionals in several international programs travel to remote areas and perform cleft lip/palate repair and other reconstructive procedures. These high-volume initiatives serve tens of thousands of patients. There is no need for rural families to lose hope for their child.

Listed on the following page are just a few organizations that you can contact to gain more information or to arrange a visit to your area.

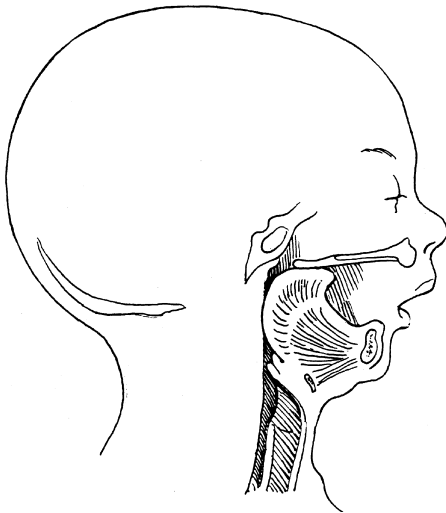
Table 2. Visiting Surgeon Programs

Organization	Contact Information	Background Information
Operation Smile	(757) 321-7645 Fax: (757) 321-7660 6435 Tidewater Drive Norfolk, VA 23509 www.operationssmile.org	Operating since 1982 Members travel in U.S. and through- out world
Interplast	(650) 962-0123 Fax: (650) 962-1619 300-B Pioneer Way Mountain View, CA 94041 www.interplast.org e-mail: DirMedServcs @Interplast.org	Operating since 1969 Members travel throughout world
American Society of Plastic Surgery- Reconstructive Surgeons Volun- teers Program	(847) 228-9900 Fax: (847) 228-9131 444 E. Algonquin Rd Arlington Heights, IL 60005 www.plasticsurgery.org	Formed in 1988 Clearinghouse that offers information about many organizations that volunteer time and skills worldwide

Pierre Robin Syndrome

Pierre Robin syndrome is another congenital anomaly to be aware of. These patients may also have a cleft palate, so be sure to evaluate all neonates with a cleft palate for signs of this syndrome.

Pierre Robin syndrome describes an infant with a small, posteriorly displaced mandible that causes the tongue to seem too large for the mouth. During fetal development a cleft palate or a highly arched



Pierre Robin syndrome. Note the small lower jaw and the resultant position of the tongue in the back of the throat. The tongue can cause potentially life-threatening airway obstruction. (From McCarthy J (ed): Plastic Surgery. Philadelphia, W.B. Saunders, 1990, with permission.)

palate forms to accommodate the relatively large tongue. Early recognition of newborns with Pierre Robin syndrome is mandatory to avoid serious complications.

Immediate Concerns

Affected infants may present with episodes of **severe respiratory distress**. When the infant is in an upright position or with the head forward, the tongue falls forward and the airway opens. If the infant is placed in the supine position (lying down, face up—the usual position in which an infant is placed to sleep), the tongue falls backward and blocks the airway, causing severe respiratory distress. If unrecognized, this syndrome can result in death.

Treatment

- The infant must be monitored closely and kept in the prone (face-down) position for sleep.
- The child should sit upright in a somewhat forward position when eating.
- Occasionally the tongue needs to be stitched to the lower lip to prevent the tongue from falling backward.
- Rarely, in very severe cases, the infant may require a tracheotomy to maintain an adequate airway.
- If the infant has a cleft palate, repair is usually delayed until the child is a few years old because of airway concerns.
- As the infant grows, the mandible grows. Respiratory concerns become less problematic. Usually, no other treatment is required.

Bibliography

1. Mackool RJ, Gittes GK, Longaker MT: Scarless healing. *Clin Plast Surg* 25:357–366, 1998.
2. Stal S, Klebuc M, et al: Algorithms for the treatment of cleft lip and palate. *Clin Plast Surg* 25:493–507, 1998.