Cleft Lip and Palate Program





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On behalf of Children's Hospital Boston's Department of Plastic & Oral Surgery, we are pleased to share the 6th edition of our Cleft Lip and Palate Booklet with you. This booklet is for parents of children born with cleft lip and/or palate. It was first written by the Children's Hospital Cleft Lip/Palate Team more than 25 years ago. Our surgeons and staff work hard to provide the highest quality care for patients and their families, and continuously look for ways to improve our services. This 6th edition is necessary because of advancements in the understanding and care of these special children.

This booklet provides basic information. It is not a substitute for ongoing dialogue between parents and members of the Cleft Lip/Palate Team. Please do not hesitate to ask for more detailed information and never hesitate to ask questions. We are excited to share our Cleft Lip and Palate Booklet with you. We hope the information on the following pages reaffirms the reasons you chose the Cleft Lip and Palate Program here at Children's Hospital Boston.



John B. Mulliken, MD Director, Cleft Lip/Palate Program



John G. Meara, MD, DMD, MBA Plastic Surgeon-in-Chief

The Cleft Lip and Palate Program at Children's Hospital Boston cares for more than 575 patients each year, making it one of the largest in the country. Our training, experience and commitment to innovative care with compassion has made us a national leader in the care of children and adolescents with cleft lip and palate.

Cleft lip/palate is the second most common birth defect in the United States, affecting one in 700 infants. Prenatal ultrasonography will detect cleft lip/palate in the second trimester. Detection at this stage helps parents to understand the condition and prepares them for the treatment of their child. Because the cleft involves the lip, nose and palate, it may affect hearing, speech and the teeth. A dedicated interdisciplinary team of specialists is required to treat these children. Such a team is the heart of our program. In addition to evaluation, repair and long-term management, we also give information and support to our patients and their families.

Insurance Information

Insurance coverage can be confusing and difficult to navigate. Because cleft care can require long-term maintenance, be sure to ask your individual insurance provider what is and is not covered. We define these procedures later in the booklet. It will also be beneficial for you to ask what referrals you might need and what procedures are classified as medical versus dental. Please enroll your child in your dental insurance as dental procedures are typically not covered by medical insurance. If you have further questions, please contact your insurance provider for assistance.

Basic definition

Cleft lip and cleft palate are congenital anomalies of the mouth and lip that occur in the first three months of pregnancy. In a cleft lip, the two sides of the lip do not fuse together as they should during fetal development about five or six weeks into pregnancy. With cleft palate, the roof of the mouth fails to completely form around eight to twelve weeks into pregnancy. A child may be born with a cleft lip, a cleft palate or both.

Diagnosis and presentation at birth

Prenatal ultrasonography can detect cleft lip/palate as early as 16 weeks of gestation. The diagnosis is confirmed at birth when a detailed assessment is made. During the first days of life, your child may have some difficulty feeding due to an inability to latch onto the breast or a bottle and maintain suction. Members of the Plastic Surgical and nursing staff will assist in your child's feeding needs. For more on feeding, turn to page 15.

Care and treatment

Care and treatment of a child with a cleft depends on the severity of the defect. For example, a unilateral (i.e., one side only) incomplete cleft lip may require only one operation early in life and nothing else. A unilateral complete cleft lip may require two procedures. A bilateral (i.e., both sides) complete cleft lip and palate requires a more long-range treatment plan. Traces of the cleft may not completely vanish until the late teenage years, when maturity is reached and facial growth is finished. While these long-term treatment plans may seem daunting at first, it is important to realize that operations are now relatively minor and usually only require a hospital stay of one to three days.

In order to understand the overall management of a child with cleft lip and/ or palate, it is helpful to examine the care that is necessary at various stages of development.

The first year

Depending on the extent of the cleft and the health of the baby, surgical correction is usually completed in the first year of life. Early treatment can include dentofacial orthopedics and surgical repair of the cleft lip and nasal deformity. Cleft palate is closed before the age of one year. Problems with hearing are assessed prior to cleft palate repair. As a child with cleft palate grows, there will be periodic hearing tests, ear exams and the possibility of ear tubes. Your child is evaluated yearly by our interdisciplinary team with a focus on speech and dental development and occlusion, skeletal growth and appearance.

The preschool years

The preschool years are crucial in the development of speech and teeth. First dental evaluations should be made between 12 and 18 months. Approximately 5 to 15% of children who have a repaired cleft palate will need an operation to correct nasal speech. This procedure is usually performed at age five years.

Overview of Cleft Lip and Palate

School age

Your child's secondary (adult) teeth will start to appear during this period. These secondary teeth may not erupt properly. Our surgical team may recommend orthodontics and premaxillary expansion in late childhood. Children whose cleft involves the gum will usually require an alveolar bone graft to help the teeth erupt and permit orthodontic alignment. This normally takes place between 8 and 10 years of age, depending on the dental maturity of the child.

Puberty and beyond

Facial structures may require further correction soon after completion of pubertal growth. Although the nasal deformity is corrected along with the initial lip repair, additional revisions of the nose may be needed. Furthermore, full orthodontic treatment to coordinate bite (occlusion) and relieve crowding and dental rotations may be necessary.

After the completion of skeletal growth the patient may have a malocclusion that requires orthodontic treatment with advancement of the upper jaw. Final correction of the external nose and septum may be needed after the orthognathic procedure. Naturally, not all children will require all these procedures. It depends on the type and extent of the original cleft.

In this booklet, members of our team have focused on communicating information that will help make the process as smooth as possible for parents and baby.

Causes and genetics

Why was my child born with a cleft lip and/or cleft palate?

Cleft lip and cleft palate are caused by an interaction between genetic and environmental factors. Some children born with a cleft have a parent or a distant relative with a cleft. Usually there is no family history of cleft lip or palate. Cleft lip and isolated cleft palate are considered separate genetic entities. Both are multifactorial conditions, meaning there are many factors that can cause a baby to be born with a cleft.

Although genetic factors are involved in the formation of cleft lip and cleft palate, environmental factors also need to be considered. Many medications, especially anticonvulsants, have been shown to increase the chance of a baby being born with cleft lip and/or cleft palate. Other factors that have been shown to increase the risk include maternal illnesses, tobacco or alcohol use during pregnancy and possibly vitamin deficiency.

Genetic factors refer to the inherited information in genes. We have about 30,000 to 50,000 genes in each of our cells, which, in early life, direct the development of the embryo. These genes go on to determine physical characteristics as well as risks for different diseases throughout life. There are many genes that interact in an abnormal way to cause cleft lip and/or cleft palate. Some genetic changes can lead to isolated cleft lip and/or cleft palate. Other genetic changes result in syndromes that can include other findings in addition to a cleft.

Approximately 10% of babies born with cleft lip, with or without cleft palate, have other physical differences; however, approximately 30% of babies born

Overview of Cleft Lip and Palate

with isolated cleft palate have other physical differences. Therefore, some babies born with cleft lip and/or cleft palate should be evaluated by a geneticist to rule out a possible syndrome by examination and appropriate genetic testing. This evaluation is critical for optimizing the care of the child by addressing other potential medical concerns as early as possible. Also, if a genetic evaluation uncovers a genetic disorder, the chance of having another child with cleft lip and/or cleft palate may be increased.

What are my chances of having another child with a cleft lip and/or cleft palate?

The chance of having more than one child with a cleft lip and/or cleft palate is different for each family. Members of the Cleft Lip and Palate team and the geneticist will provide detailed answers to this question.

Once a genetic syndrome has been ruled out, the condition is referred to as isolated cleft lip and/or cleft palate. There are, as yet, no tests to determine the genetic changes in the absence of a recognized syndrome.

If there is one affected person in the family with a cleft, the likelihood of having a child with a cleft lip and/or palate is 2 to 5%. If there is a second affected person in the family, either another sibling or a parent, the chance for future children to have cleft lip increases to 10-14% and the risk for isolated cleft palate rises to 8%

Do children with a cleft lip always have a cleft palate?

Not necessarily. A child can be born with just a cleft lip, just a cleft palate, or a cleft lip and cleft palate together.

What is a cleft lip?

A cleft lip is an incomplete union of skin and muscle between the nose and lip. The lip is formed during the first four to six weeks of pregnancy. During this time, skin and muscle normally grows in from both sides of the face to join with skin that grows down from the tip of the nose. If the growth and union of these parts are not complete, the baby is born with a cleft lip.

Are all cleft lips the same?

There are several types of cleft lip. The lip, nose and palate can be involved to varying degrees. The major types of cleft lips are:

- unilateral (to the left or right of midline, but not both; left side is more common)
 - incomplete (partial but not full union of skin/muscle)
 - complete (total absence of skin/muscle fusion)
- bilateral (to the left and right of midline)
 - complete (this is most common with bilateral clefts)
 - incomplete
 - asymmetrical (complete on one side and incomplete on the other side)



Unilateral incomplete cleft lip



Unilateral complete cleft lip/palate



Bilateral complete cleft lip/palate

Can my child's cleft lip be repaired?

Your child's cleft lip can be closed. After the operation, your child's mouth and nose will be near normal in appearance and will function normally.

When will my child's cleft lip be repaired?

Your child's cleft lip is usually closed before six months of age. Cleft lip and cleft palate are repaired in separate operations. The plastic surgeon on your team will talk with you about the best operative plan for your child.

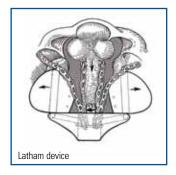
How is my child's cleft lip repaired?

The plastic surgeon uses the existing muscle and tissues of your child's lip and nose to close the cleft. The repair takes one or two operations depending on the type of cleft lip. The repair is performed in the operating room under general anesthesia. Your child stays in the hospital for one or two nights after each operation. We encourage one parent to stay with the child. The nurse will teach you how to care for your child after each procedure. There is information in this booklet about feeding your child before and after cleft lip repair.

An incomplete cleft lip (unilateral or bilateral) is usually repaired with one operation when the child is about three to five months of age. During this operation the nasal asymmetry is also corrected.

Is pre-surgical orthodontic treatment required?

Repair of unilateral complete cleft lip (associated with a complete cleft palate) is done in stages. The first procedure involves the insertion of a dental appliance called a Latham device. At 3-to-6 weeks of age, the pediatric dentist makes an impression of your child's gums which is then used to make the appliance. At 6-to-12 weeks old, the dentist inserts the appliance in your child's mouth: this is done in the operating room under general anesthesia. The appliance is then manipulated a little every day to slowly bring the gums closer together and improve the surgical repair of the lip. The appliance stays in the mouth for 6-to-8 weeks.



It will be removed in the operating room just before closure of the cleft lip.

Is a "lip-nasal adhesion" procedure required before the cleft lip repair?

In certain circumstances, the first operation for a unilateral complete cleft lip is a "lipnasal adhesion." This operation is performed at about 3 months of age, and involves:

- a simple closure of the lip
- the first stage of nasal correction
- when possible, closure of the gum cleft (called "gingivoperiosteoplasty")



Left unilateral complete cleft lip/palate



After lip-nasal adhesion



Patient at age 8



Submental vertex view

When is the complete repair made?

The second operation is a more complete repair of the cleft lip and correction of the nose. This second operation takes place roughly 12 weeks after the lip-nasal adhesion (5-to-6 months of age).

Repair of a bilateral complete cleft lip usually requires one operation at about 4-to-6 months of age. The nose and gums are repaired at the same time as the lip closure.





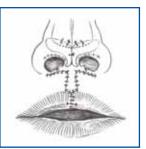


Unilateral complete cleft lip/palate

After labial adhesion

After dermal nasolabial repair





Repair of a bilateral complete cleft lip

Will my child look normal after the cleft lip is repaired?

After the operation, the lip, nose and face are swollen for a few days. The scar may be red for several weeks. It will take 6-to-12 months for the scar to soften and fade. The scar will never completely disappear, but in time, it is usually difficult to see. Your child's lip and nose will be nearly normal in appearance.

Is the nose corrected as part of the cleft lip repair?

Nasal asymmetry is commonplace with a cleft lip. Although nasal correction is part of the initial lip repair, sometimes it is difficult to permanently correct the nose in one operation.







Unilateral incomplete cleft lip, before and after repair







Bilateral incomplete cleft lip, before and after repair







Bilateral complete cleft lip/palate, before and after repair

As my child gets older, will another operation of the lip or nose be needed? Revision of your child's lip and/or nose may be indicated before beginning school or during adolescence, but sometimes revision of the lip or nose is never necessary.

Although repaired at the time of lip repair, children whose cleft lip involves the alveolus, or the gum line, usually need another operation to help the permanent teeth to erupt and to allow orthodontic manipulation. This operation is called an "alveolar bone graft". It is usually performed when the child is 8-to-10 years old. Orthodontic preparation is needed for this procedure. These topics are discussed in more detail later in this booklet.

Do children with a cleft palate always have a cleft lip?

Not necessarily. A child can be born with just a cleft lip, just a cleft palate or a cleft lip and cleft palate together.

What is a cleft palate?

A cleft palate is an opening in the roof of the mouth. The palate forms during the first 8 to 12 weeks of pregnancy. During this time, bone and muscle grow in from both sides of the upper jaw and join to form the roof of the mouth and the floor of the nose. If the fusion of these two shelves of bone and muscle is not complete, the baby is born with a cleft palate, i.e., an opening between the mouth and the nose.

Are all cleft palates the same?

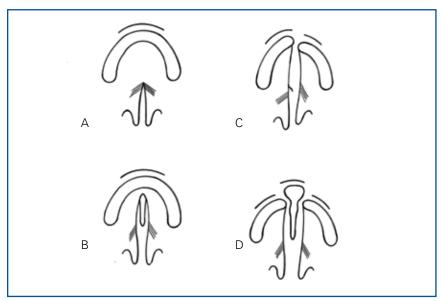
The palate is composed of two parts, a muscular part (soft palate) and a bony part (hard palate). The soft and hard palate can be involved to varying degrees.

Can my child's cleft palate be repaired?

Yes. The opening in the hard and soft palate is usually closed in one operation.

When will my child's cleft palate be repaired?

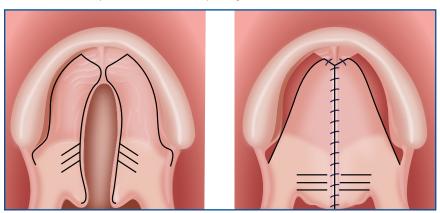
The cleft palate is closed between 8 and 11 months, before your baby makes his the first attempt to speak. A plastic surgeon on our team will talk with you about the best operation and timing for your child.



Types of cleft palate - (A) soft palate; (B) bilateral complete cleft palate; (C) unilateral complete cleft lip and palate; (D) bilateral complete cleft lip and palate. Lines indicate the abnormal direction of palatal muscle before repair.

How is my child's cleft palate repaired?

The plastic surgeon brings together the separated muscles and tissue from the two halves of the palate to close the opening.



Cleft palate before (left) and after closure (right)

Closure of a cleft palate is performed in the operating room under general anesthesia. Your child will be in the hospital for 1-to-3 nights after the operation. We encourage one parent to stay with the child in the hospital. There is information in this booklet about feeding your child before and after cleft palate repair.

As my child gets older, will another operation on the palate be necessary?

About 5% to 15% of children who have had a cleft palate repair will need an operation to correct persistent nasal speech. The most common operation is called a "pharyngeal flap." This operation is usually done when the child is around five years of age or older.

How do I feed my baby before the operation?

Your baby's ability to feed, whether by breast or bottle, is determined by the extent or severity of the cleft lip/palate. At birth, the team nurse will determine the type of feeding method that's best for your baby.

- A cleft palate only may require some adaptations for feeding. If there is soft
 palate cleft only, sometimes a "nipple" shield can be used to assist in breast
 feeding. If the infant is working too hard or the hard palate is involved, then a
 VentAire® feeder may be effective with a small cross cut in the silicone nipple.
 Other devices can be used, like the Haberman® feeder, pigeon nipple or Ross®
 nipple.
- Cleft of the lip and palate usually requires a special feeding device. Usually a
 Haberman® feeder is recommended. The reason for this is the milk can be
 pumped in synchrony with the suck-swallow sequence. The other helpful
 device is a Ross® nipple to help deliver the flow.



Feeders

The cleft team nurse will also show you how best to feed your infant.

Can I breastfeed?

If your infant has a cleft lip only, attachment to the breast is fine with some adaptation to cover the cleft. If breast feeding is not an option, then any type of silicone nipple and bottle may be used.

Feedings should last no more than 30 minutes. Prolonged feeding can exhaust you and the baby. Infants spend calories very quickly. The nurse will determine the total amount of milk for your infant over a 24-hour period. Feeding should occur every three to four hours. Never let your infant go more than four hours without a feeding. The exception to this rule is if the infant is close to making the volume quota for the 24-hour period. Establishing a "rhythm" with your infant is paramount. Watch for your infant's "hunger cues" and do not interrupt a sucking

pattern. When the sucking stops, burp your infant while holding the child upright and supporting the lower jaw.

Is there a positioning-technique for bottle feeding?

Wrap your infant, enclosing the hands, in a blanket. This is called "swaddling." Sit in a comfortable chair, like a rocking chair or a "glider" chair, with a footstool. Hold the baby upright in your arm or hold the head from behind. Relax both arms, and place the nipple gently into the baby's mouth.

Lay the nipple on top of the tongue. Rotate your arm so that the underside of your hand is holding the bottle. Put your ring finger under the baby's chin. With firm pressure, keep your ring finger in place so as the baby suckles you feel pressure against your finger. Your infant should feel comfortable while suckling (i.e., no straining or squirming to access the nipple and to swallow). If your infant has a cleft of the lip and palate, position the nipple so that the upper and lower gums connect with it.

With gentle pressure under the chin, push up to start the baby's sucking. Maintain this pressure. If, after a minute of sucking, there is little flow of milk, rotate the nipple to a longer line or compress the nipple with gentle pressure in synchrony with the suck-swallow reflex.

Watch for cues that the infant is either satiated or needs burping ("bubbling").

What should my baby and I do right after feeding?

Keep the baby upright for about 20 minutes, either by holding or placing the infant in a seat. When placed in a bed, slightly turn the infant's body to the side with a wedge. Elevate the bed by 20 degrees: the chest should be higher than the stomach. An infant with a cleft palate may exhibit some esophageal and nasopharyngeal reflux, as evidenced by milk coming out through the nose, or regurgitating shortly after feeding has ended. You should always keep a suction bulb handy.

Record the time, length and amount of feeding.

Weigh your infant once per week. If your infant is not gaining more than one ounce per day, you should talk to your pediatrician about increasing the caloric content of the milk.

How can I adjust my feeding if my baby is not gaining sufficient weight?

If your baby is gaining less than one ounce per day, calories in the formula or breast milk need to be increased. This can be accomplished easily by concentrating the formula or adding powdered milk to your breast milk. This should be discussed with your pediatrician or cleft nurse.

If the feeding device is not working for you or the infant, the cleft team nurse will give you alternative feeders to try. If weight gain is insufficient even after increasing the calories per volume, your child may need consultation with a gastrointestinal/nutrition specialist.

Are there ways to supplement or replace oral feeding?

When oral feedings are not sufficient for your baby, there are other methods that can be used to deliver the needed calories. Sometimes a naso-gastric tube is passed through the nostril into the stomach. The tube is connected to a syringe. Typically, use of a naso-gastric tube is a temporary solution until the infant is able to take all the necessary calories by mouth. A gastrostomy tube is another method of feeding. Insertion of this tube directly into the stomach from the outside of the belly requires anesthesia. A gastrostomy is usually used in situations where the infant has difficulty in both breathing and eating. Also, it is used if there is danger of aspiration (the entry of food, liquids or foreign material into the trachea and lungs). The food source is again delivered through a syringe or mechanical device. This is usually not a permanent method of feeding. As infants grow, they learn to eat by mouth.

How do I feed my child in preparation for cleft palate repair?

In some, but not all babies, cup feeding may be introduced prior to the cleft palate repair. It will take several months for your child to become used to cup feedings. You should begin by introducing the cup at six months of age. Start by using the cup to replace one feeding a day and gradually increase the number of cup feedings. Over several weeks, you should be able to completely wean your child from the "cleft feeder." If you are not able to completely wean off the cleft feeder to a cup, you may continue using it. Begin spoon feedings when your child is six months old, using a soft-tipped spoon. Give your child baby cereal, fruits, vegetables and other foods as directed by your child's pediatrician. You may also give liquids with a spoon.

How do I feed my child after cleft palate repair?

One of our cleft nurses will review feeding instructions around the time of the cleft palate repair. In some instances your child may return to using a cleft feeder. If the cleft feeder is not appropriate for your child, a "sippy cup" will be introduced prior to the palatal cleft repair. Your child should use a cup without a spout or with a very short spout. Your nurse will show you which type of cup is right for your child. You will need to use a cup for all feedings during the first 10 to 14 days after palatal repair. If your child is having difficulty getting enough fluid with the cup, it is permissible to use a silicone nipple with a large cross cut. This can be accomplished by using the Haberman® bottle or a standard soft silicone nipple. It may be necessary to feed your child with the Ross® nipple. After ten days, give soft foods using the side of a soft-tipped baby spoon. After each feeding, rinse your child's mouth with a sip of water from a cup. Rinsing is very important, especially for the first 10 to 14 days after the palatal closure. Ask your nurse about using a special syringe for rinsing. Your child may regurgitate some food and liquid through the nose for up to three months after the operation. This is normal. It takes time for swelling to diminish and for the palatal muscles to begin working properly. You'll be given further post-operative instruction after discharge.

Will arm restraints be necessary for my child after surgery?

Please discuss this with your surgeon. There are some infants for whom restraints are unnecessary.

Will my baby be nurtured as well as nourished?

Your infant is a normal baby with an anatomic defect that can be surgically corrected. Once you feel comfortable with the feeding method, you will be more at ease with nurturing. You will see your baby thriving, smiling and responding to your touch. It is very important that at least three people supporting you feel comfortable with feedings. Your infant will sense when someone is confident with feedings and will be more relaxed.

Do support groups exist?

You are not alone in caring for your infant. The cleft team will do everything to help you through this time. Also, there are several parent support groups available who are willing to help. This information is available in your packet. You may also visit the Boston Cleft Lip and Palate Page on Facebook at www.facebook.com/bostoncleft.

Will my child have difficulty hearing?

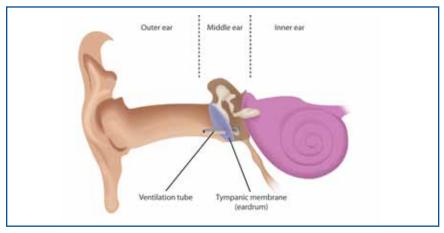
Children born with a cleft palate often have temporary hearing loss because of fluid in the middle ear and recurrent infections. The hearing loss may last for a short time or a number of months. Speech and language development is influenced by a child's ability to hear well. Your child will have their first hearing test early in life, and again prior to the operation to repair the cleft palate. The child's age and developmental level determine which hearing test method will be used.

- For the very young infant (newborn to six months), a special hearing evaluation called an Auditory Brainstem Response (ABR) is performed while the child is asleep.
- The older child (who is at a developmental level of six months or more) can undergo a hearing evaluation by behavioral audiometric test methods. The test is conducted while the child is awake and able to participate.
- The test that is typically conducted on a child between seven months and twoand-a-half years old is the Visual Reinforcement Audiometric (VRA).
- A child who is between two-and-a-half and five years old should have a hearing test by conventional audiometric testing methods (hand-raise response).

Tympanometric testing is often performed at the time of hearing evaluation. This is a test of middle ear function. It can also be used to check the function of ear tubes or the presence of a perforation of the eardrum. The audiologist on the team will monitor your child's hearing every 6 to 12 months. The audiologist and otorhinolaryngologist (ORL) work together closely. The ORL surgeon's specialty is the care of ear, nose and throat problems. Before the cleft palate repair, your child will have an appointment with an ORL specialist in order to discuss the ear and care of the child with cleft palate.

Is there treatment for persistent middle ear fluid and associated hearing loss?

The tube that connects the middle ear to the throat (Eustachian tube) does not drain normally in an infant with a cleft palate and thus fluid collects in the middle ear space. Fluid in the middle ear space (effusion) is present in virtually all infants under one year with an unrepaired cleft palate. This middle ear fluid is associated with hearing loss that can cause problems with the development of speech, language and cognitive function. Persistent middle ear fluid is also associated with an infection called "otitis media." Infants with cleft palate, middle ear effusion and hearing loss will require an operation to remove the fluid and to insert a ventilation tube. This operation is performed under general anesthesia, generally at the same time as the cleft palate repair. The ventilation tubes usually stay in place for 9 to 12 months. The ORL specialist evaluates the tubes every 6 months. Ventilation tubes normally fall out spontaneously; they do not require an operation for removal. Unfortunately, 50% of infants with cleft palate repair will require the repeat insertion of ventilation tubes. Two problems associated with the insertion of ventilation tubes are scarring of the eardrum and a hole (perforation) in the eardrum. These problems can also be caused by persistent fluid in the middle ear or by repeated ear infection. Ventilation tubes are necessary for normal long-term hearing, and also help in development of normal speech.



Basic anatomy of the right ear with ventilation tube

How does the palate affect speech?

The hard and soft palate separate the mouth from the nose. When we breathe, the air flows in and out of our lungs through the nose and throat. When we talk, the muscles in the soft palate move the palate to the back of the throat to seal off the nose (called the velopharyngeal valve). This allows the air to flow through the mouth alone when we speak. There are only three English speech sounds (m, n, and ng) that are made through the nose.

Cleft lip alone does not cause speech problems. A child born with a cleft palate will be unable to make normal speech sounds other than the nasal (m, n, and ng) until after the palate is closed. Your child will be evaluated by the speech pathologist on the team shortly after the palate is repaired. Immediately following palatal closure, vocalizations may decrease in frequency and variety. It may take up to six weeks for children to resume their typical level of vocalization. The speech pathologist will give you suggestions for home activities that are aimed at developing normal speech and language skills. For example, your baby should be encouraged to make lip sounds (p and b) and front-of-the-tongue sounds (t and d). You can do this with playful lip-popping games (mimicking a fish) and tongue clicking games (mimicking a horse trot). Sounds made in the throat such as "uh oh" or animal roaring sounds should be discouraged as they can cause poor speech habits in children with a repaired cleft palate.

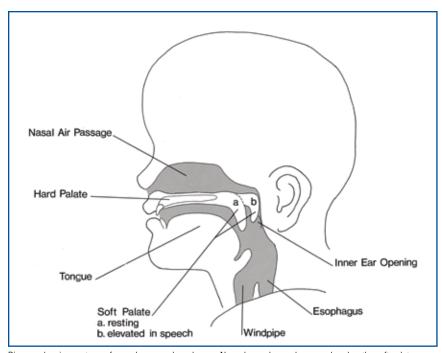


Diagram showing anatomy of nasopharynx and oropharynx. Normal speech sounds are produced as the soft palate elevates from ${\bf a}$ to ${\bf b}$.

Will my child experience difficulty speaking?

Some children with a cleft palate can have speech and language delays. These may be related to the temporary hearing loss associated with the cleft palate and middle ear fluid. Some children may exhibit difficulty with speech if their palate is not effectively closing off the nose from the mouth while they are speaking. Many children will acquire speech and language skills at a normal pace after the palate is closed and middle ear tubes are placed for drainage.

Will my child require speech therapy?

Speech therapy helps children to make sounds in a normal manner. The speech pathologist on the team will determine your child's need for speech therapy. Recommendations for speech therapy, when needed, will be made to the speech pathologist in the early intervention program or school system in your community.

Are additional operations needed?

The adenoids naturally assist children in sealing off the nose during speech; they should only be removed after consultation with the Cleft Lip and Palate team. The tonsils do not help in sealing off the nose from the mouth and can be removed if there is a medical need to do so. About 5% to 15% of children will not be able to adequately seal off the nose during speech after the palatal repair. This results in hypernasal speech (too much nasal airflow) called "velopharyngeal insufficiency" (VPI). This requires another operation, called a "pharyngeal flap," to correct the problem. When needed, this option is performed when the child is approximately 5 years old following a complete speech evaluation with a speech pathologist, otolaryngologist and plastic surgeon.

Your child's speech should be evaluated every 6 months until the age of 3 and on a yearly basis thereafter. You are encouraged to call the team's speech pathologist at any time if you have questions or concerns.

Will my child's cleft lip/palate affect his/her teeth?

Your child's first tooth (usually the lower incisor) may appear between 4 and 14 months of age. By age 3, children usually have their primary (baby) teeth. The child with cleft lip and/or cleft palate may have poorly formed enamel (outer tooth layer) on some of the teeth, especially those near the cleft. Teeth in this region may also be out of alignment, partially erupted, and, therefore, difficult to clean. All of these factors make the teeth more susceptible to the development of cavities.

How do I clean my child's teeth?

Brush your child's teeth at least twice a day to minimize cavities. Avoid foods with a lot of sugars and starches. Frequent snacking is especially harmful to the teeth since the bacteria in dental plaque produce cavity-causing acids each time food is placed in the mouth. Fluoride, whether through the water supply or through prescribed supplements, clearly reduces the amount of decay in the baby teeth and permanent teeth. The greatest benefits from fluoride occur between 6 months and 8 years of age. Therefore, the child with a cleft lip/palate should be placed on the optimal fluoride dosage early in life.

When should my child see the dentist for the first time?

If your child had a complete cleft lip/palate and required a dental appliance, you probably have already met our dental team. Your child should visit a pediatric dentist between 12 and 18 months old, or earlier if you have any questions or concerns. Your dentist will instruct you in the proper techniques for brushing your child's teeth. Fluoride toothpaste may be used, but only in very small amounts.

What dental issues should I expect as my child gets older?

As your child grows and the teeth and bite develop, the pediatric dentist and orthodontist will periodically evaluate the need for treatment. Common problems include missing, malformed or extra teeth in the region of the cleft. Absent teeth may need to be replaced artificially or by moving teeth into the space with orthodontics. A dental implant can be inserted if a tooth is missing. This may be an option for your child once growth is complete.

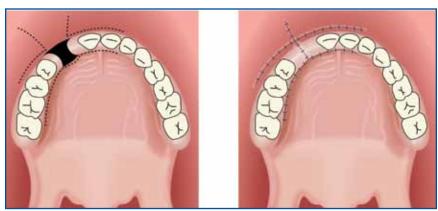
Will orthodontic therapy be required?

The bite is almost always affected in some way, and most children with a cleft require one or more phases of orthodontic treatment (braces). The decision to treat the teeth and/or bite should be made by a pediatric dentist and/or orthodontist with experience in treating children born with a cleft lip/palate.

What happens if my child's cleft includes the gum?

Although the gum cleft is usually repaired at the time of lip repair, children whose cleft lip involves the "alveolus" (gum line) usually need an initial phase of orthodontics (braces) and secondary closure of the alveolar cleft to allow the permanent teeth to erupt. **Phase I** orthodontics involve an appliance used in the maxilla to widen the palate prior to the bone grafting procedure. Once the orthodontic movements have been completed the patient is prepared for a procedure known as the "alveolar bone graft."

This operation is usually performed when the child is 8 to 10 years old. Minor revisions of the lip and nose also can be done during the bone graft operation. The bone is harvested from the posterior hip region; it is inserted in order to join together the soft tissue and bony gap in the upper jaw. The bone graft heals and solidifies to provide support for the surrounding teeth and/or a dental implant.



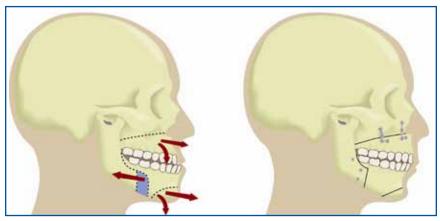
Grafting alveolar cleft (left) and closure of fistula (right)

A second phase (Phase II) of braces may be required and allows for proper alignment of the permanent teeth and usually takes place in the teenage years. This may include dealing with canine eruption, further alignment of teeth, resolution of crowding and use of reverse-pull headgear to address the abnormal relationship between the upper and lower jaw (called an "under-bite").

Children with a repaired incomplete or complete cleft lip, but intact palate, usually have normal facial growth. Children with a complete cleft lip/palate often do not have normal forward and downward growth of the upper jaw. This results in an under-bite. Some patients can be managed with **Phase III orthodontics** between 13 and 15 years old for girls and 16 and 18 years old for boys.

Orthognathic Surgery

For some patients, orthodontic therapy is sufficient; for other patients an operation is necessary to properly align their jaws and bite after growth is complete. The operation, called Le Fort I maxillary advancement, is needed just as facial growth has been completed in the late teen years. Orthognathic surgery refers to the repositioning of the maxilla, mandible and the alveolar bones to give facial symmetry. One or more segments of the jaw can be concurrently repositioned to treat various types of malocclusions and jaw deformities. These procedures correct both functional and esthetic problems that are due to underlying skeletal malformation.



Repositioning of upper and lower jaws to correct the bite and provide facial balance

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Age	Cleft Lip	Cleft Palate	Cleft Lip & Palate
6 weeks to 3 months		Consider ABR test	Dentofacial orthopedics for complete cleft lip and palate
2 to 5 months	Repair cleft lip and nasal deformity		Repair cleft lip, nasal deformity, and cleft gum
8 to 10 months		Hearing test and ear exam	Hearing test and ear exam
10 months		Repair cleft palate; place ear tubes if needed	Repair cleft palate; insert ear tubes if needed
10 to 15 months		Speech/language evaluation	Speech/language evaluation
1 to 5 years		Hearing test every 6 to 12 months	Hearing test every 6 to 12 months
18 to 36 months	First dental evaluation	Dental evaluation	Dental evaluation
4 to 5 years	Consider revision of lip and nose	Consider pharyngeal flap (VPI)	Consider pharyngeal flap or revision of lip/nose
7 to 10 years			Phase I orthodontics; premaxillary expansion, removal of retained baby teeth; alveolar bone graft to close gum cleft and/or premaxillary osteotomy
12 to 14 years	Consider revision of nasal tip		Consider revision of nasal tip; Phase II orthodontics; full orthodontic treatment to coordinate bite and relieve crowding rotations
15 to 20 years	Final correction of external nose and septum		Final correction of external nose and septum; Phase III orthodontics; orthodontic treatment with orthognathic correction

^{*}ABR: auditory brainstem response hearing evaluation

^{*}VPI: velopharyngeal insufficiency; persistent nasal speech

^{*}ORL: The Department of Otolaryngology (ORL) and Communication Enhancement

Resources

- John B. Mulliken, MD 617-355-7686
- John G. Meara, MD, DMD 617-355-4401
- Contact a Nurse 617-355-4513
- Cleft Lip and Palate Program childrenshospital.org/cleftlip 617-355-6309
- Department of Plastic & Oral Surgery childrenshospital.org/plastic 617-355-7252

- facebook.com/bostoncleft
- bit.ly/bostoncleftapp (iPhone® app)
- Foundation for Faces of Children facesofchildren.org

Notes			



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