

Cleft lip and palate

A guide for families



Boston Children's Hospital
Plastic and Oral Surgery

bostonchildrens.org/plastics

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Welcome

Welcome to the Boston Children’s Hospital Cleft Lip and Palate Program. This guide is meant to provide you with important information about cleft lip and cleft palate, as well as related issues that children and families often face. It also includes helpful suggestions for handling insurance questions and locating the many resources available to you and your family.

In addition to providing the highest quality medical and surgical care for all patients, our program is committed to supporting families throughout the treatment process. We are always here to address your questions and concerns. Please do not hesitate to contact us at any time by calling **617-355-6309**.

Thank you for entrusting us with your child’s care. We hope the information on the following pages reaffirms the reasons you chose Boston Children’s.



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Introduction to our program

The Cleft Lip and Palate Program at Boston Children’s Hospital is one of the largest and most experienced treatment centers in the country, caring for more than 575 babies, children and adolescents every year. Our combination of specialized training, innovative treatments and compassionate, family-centered care has made us a national leader in the field.

Because a cleft can involve the lip, nose and palate (the roof of the mouth), a child’s hearing, feeding, speech and dental functions may all be affected. A dedicated and knowledgeable team of specialists from several disciplines is necessary to give these patients—and their families—the care and long-term support they need. Such a team is the heart of our program.

About insurance

Figuring out the details of insurance coverage can be a confusing process. Here are some tips:

- **Ask your child’s doctor which dental and medical procedures are likely to be necessary** We will define the different types of procedures later in the booklet.
- **Be sure to enroll your child in your dental insurance plan as well as your medical plan.** Dental procedures are usually not covered by medical insurance.
- Check with your insurance provider to make sure **all of the procedures are covered by your plan.**
- Also **ask your doctor whether there are additional referrals** you may need down the line.

If you have other questions, please contact your insurance provider by calling the phone number on the back of your insurance card.

Timeline of care

Here is a general guide to the different steps that may be required during your child’s cleft care.

Timeline for the care of cleft lip and cleft palate			
Age	Cleft lip	Cleft palate	Cleft lip and 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: Otolaryngology

Overview of cleft lip

What is a cleft lip?

A cleft lip is a separation in the skin and muscle between a baby's nose and lip. Cleft lips are congenital, which means they are always present at birth.

Do all children with cleft lips also have a cleft palate?

No. A child can be born with just a cleft lip, just a cleft palate or a cleft lip and cleft palate together.

What causes a cleft lip?

Between the fourth and sixth weeks of pregnancy, a developing baby's lip begins to form. During this time, skin and muscle normally grow in from both sides of the baby's face to join with the skin that grows down from the tip of the nose. When this tissue and muscle fail to connect all the way, a cleft lip occurs.

No one knows exactly why this happens, but experts believe that cleft lip may be a genetic condition—meaning it is caused by an error in the genes. Cleft lip (as well as cleft palate) can run in families, but many children with a cleft have no known family history of the condition.

Our researchers are continuing to work toward a better understanding of how and why cleft lip occurs.

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

This depends on many factors, including whether other members of your family were also born with a cleft. Our program team, including our geneticist, will provide detailed recommendations based on your family's particular situation.

Are all cleft lips the same?

No. There are different types of cleft lip that can also involve the nose and the palate. The major types are:

- **Unilateral** (affecting only one side of the face)
- **Bilateral** (affecting both sides)

And within those two categories, there are three subtypes:

- **Incomplete** (the two sides of the lip are partially fused)
- **Complete** (the sides of the lip do not connect at all)
- **Asymmetrical** (the cleft is complete on one side and incomplete on the other)



Unilateral incomplete cleft lip

Unilateral complete cleft lip/palate

Bilateral complete cleft lip/palate

How is a cleft lip diagnosed?

An ultrasound can detect a cleft lip as early as the 16th week of pregnancy. The diagnosis will then be confirmed with a detailed exam when the baby is born.

Cleft lip repair

How is a cleft lip treated?

All cleft lips require some type of surgical treatment. There are three different operations that may be used for a cleft lip:

- Before they have a repair operation, some children first need a procedure called a lip-nasal adhesion. This procedure is performed around the age of 3 months, and involves three steps:
 - Closing the hole in the lip
 - Taking the first step to correct the positioning of the nose
 - If possible, closing the hole in the gum (this step is also referred to as a gingivoperiosteoplasty)
- An incomplete cleft lip (either unilateral or bilateral) is usually repaired in a single operation when the child is between 3 and 5 months of age. During the operation, a plastic surgeon uses the existing muscle and tissues of the child's lip and nose to close the cleft and corrects the baby's nasal asymmetry (unevenness in the shape of the nose).
- Children with a complete cleft lip usually need a second repair operation to make additional corrections to the cleft lip and nose. This procedure usually takes place at 5 to 6 months of age.

Will my child need to be admitted to the hospital for the repair?

Yes. Because all cleft lip repair procedures are performed in the operating room under general anesthesia, your child will need to stay in the hospital for one or two nights to recover.

You (or your child's other guardian) will be able to stay at the bedside, and our nursing team will be here to guide you through the process of feeding and caring for your baby during recovery. *Visit page 12 of this booklet to learn more about feeding.*

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

Your child's lip, nose and face will be swollen for a few days right after the operation. The area around the incision may be red for several weeks, and the resulting scar may take as long as a year to soften and fade. Although the scar will never completely disappear, it will become almost invisible over time.

Once the healing process is complete, your child's lip and nose will be nearly normal in appearance.

As my child gets older, will another operation of the lip or nose be necessary?

Most children won't need an additional operation. However, some children with a cleft lip that involves the alveolus (gum line) will need another procedure when they are between 8 and 10 years old. In these cases, an operation called an alveolar bone graft is needed to help the permanent teeth erupt and make it possible for the child to wear orthodontics (braces).

You can read more about orthodontics and the alveolar bone graft operation on page 21.



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

Overview of cleft palate

What is a cleft palate?

A cleft palate is a separation in the palate (the roof of the mouth).

What causes a cleft palate?

A developing baby's palate forms between the 8th and 12th week 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 bottom 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.

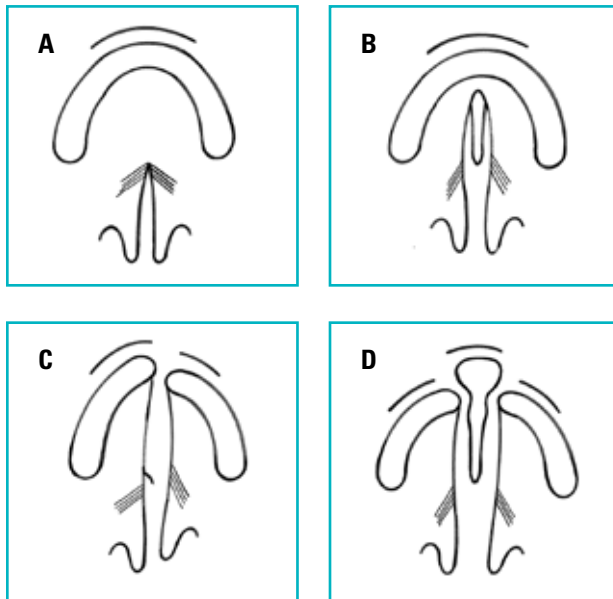
It is believed that, like cleft lip, cleft palate may be a genetic condition. Some children with a cleft palate have other family members with the defect, but many others do not. Experts are still working to determine the exact causes of cleft palate.

Are all cleft palates the same?

No. The palate is made of two parts, a muscular part (called the soft palate) and a bony part (the hard palate). A cleft can involve one or both parts of the palate.

How is a cleft palate diagnosed?

A prenatal ultrasound can detect a cleft palate as early as the 16th week of pregnancy. The diagnosis will then be confirmed with an exam when the baby is born.

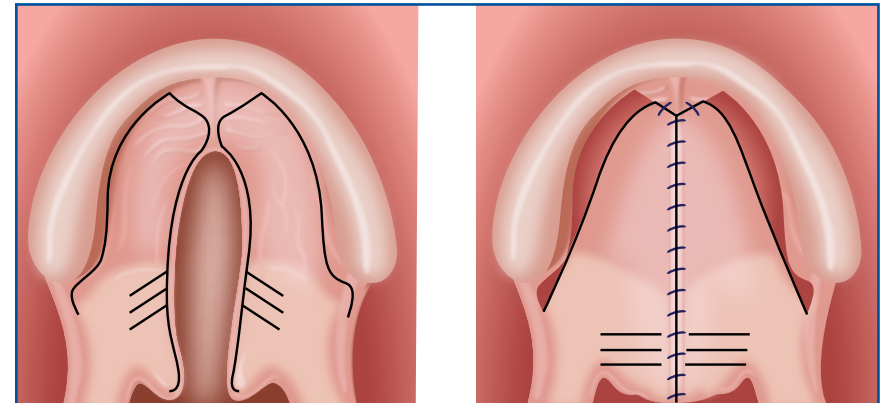


Types of cleft palate - (A) soft palate; (B) bilateral complete cleft palate; (C) unilateral complete cleft lip and palate; (D) bilateral complete cleft lip and palate. Lines indicate the abnormal direction of palatal muscle before repair.

Cleft palate repair

How is a cleft palate repaired?

A cleft palate needs to be repaired surgically. During the operation, a plastic surgeon brings together the separated muscles and tissue from the two halves of the palate, closing the opening.



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

When will my child's cleft palate be repaired?

The repair operation usually takes place between the ages of 8 and 11 months (before a baby's first attempts at speech).

Will my child need to be admitted to the hospital for the repair?

Yes. All cleft palate repair procedures are performed in the operating room and require general anesthesia. Your child will need to stay overnight for one to three nights to recover.

You (or your child's other guardian) will be able to stay at the bedside, and our nursing team will help you learn the best ways to feed and care for your baby during recovery. Visit the next section of this booklet to learn more.

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

Your baby's doctor will give you specific recommendations, but here are some general guidelines:

- If the doctor says it is ok for your baby to begin cup feeding before the cleft repair, you should start by introducing the cup around the age of 6 months.
- The cup can be used for one feeding a day, and then gradually used more often until your child is completely weaned from his or her cleft feeder.
- Begin spoon feedings around the age of 6 months. Use a soft-tipped spoon to give your baby liquids, as well as cereal, fruits, vegetables and other foods as directed by your pediatrician.

Feeding issues

How should I feed my child after cleft palate repair?

Our cleft nurses will review detailed feeding instructions, customized to meet your baby's needs, at the time of the cleft palate repair. Here are some of the basics:

- Your child should use a cup that either does not have a spout, or has a very short one. (Your nurses will advise you which type is best).
- You will need to use a cup for all feedings during the first 10 to 14 days after the repair procedure.
- If your baby is having difficulty getting enough fluid from the cup, you can use a silicone nipple with a large cross cut (for example, using the Haberman® bottle or a standard soft silicone nipple). It may be necessary to use a Ross® nipple if your baby continues to have problems with fluid intake.
- After 10 to 14 days, you can start giving your baby soft foods—using the side of a soft-tipped baby spoon.
- After each feeding, rinse your baby's mouth with a sip of water from a cup. *Rinsing is very important, especially while your baby's mouth heals.*
- Ask your nursing team whether you should try a special syringe for rinsing.
- Your baby may regurgitate some food and liquid through the nose for as long as three months after the operation. This is normal while the swelling goes down and your baby's palate starts to work properly.

Will my child look normal after the cleft palate repair?

Your child's lip, nose and face will be swollen for a few days right after the operation. The area around the incision may be red for several weeks, and the resulting scar may take as long as a year to soften and fade. Although the scar will never completely disappear, it will become almost invisible over time.

Once the healing process is complete, your child's lip and nose will be nearly normal in appearance.

As my child gets older, will he need another operation on the palate?

Approximately 5 to 15 percent of all children who have undergone a cleft palate repair will need a second operation to correct "nasal"-sounding speech. The most common procedure used to fix this problem is by using a pharyngeal flap, when the child is about 5 years old. You can read more about this surgery on page 19.

Can I breastfeed my child?

Your baby's ability to feed, whether by breast or bottle, depends on how large the cleft is. Right after birth, your care team will help you determine the type of feeding method that's best for you and your baby.

- If your baby has a cleft lip only, breastfeeding is usually fine as long as the cleft is covered during feeding.
- If your baby has a cleft palate that only involves the soft palate, a nipple shield might be helpful in assisting with breastfeeding.
- If your baby is working too hard to suckle, or if the cleft palate involves the hard palate, a device such as a VentAire® feeder with a small cross cut in the silicone nipple may be needed.
- If your baby has both a cleft lip and a cleft palate, a special feeding device will most likely be necessary. A Haberman® feeder is usually recommended, because it allows milk to be pumped in time with the baby's suck-swallow sequence. A Ross® nipple might be added to help deliver the flow of milk.



How long should my baby's feeding sessions be, and how much milk should be consumed in a session?

- Feedings should last no more than 30 minutes at a time, every three to four hours. Prolonged feeding can exhaust both you and the baby.
- Infants burn calories very quickly. Your nursing team will determine the total amount of milk your child needs over a 24-hour period.
- Never let your baby go more than four hours without a feeding—unless you are close to exceeding the amount of milk your nursing team has recommended for a 24-hour period.
- Establishing a rhythm with your baby is key. Watch for any hunger cues and do not interrupt a sucking pattern. A Ross® nipple might be added to help deliver the flow of milk.
- When the sucking stops, burp the baby while holding him or her upright and supporting the lower jaw.

Is there a positioning technique I should use when bottle feeding?

Yes. We have found this approach to be very beneficial:

- 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 his or her head from behind.
- Relax both arms and place the nipple gently into the baby's mouth.
- Lay the nipple on top of your baby's tongue.
 - If your infant has both a cleft lip and a cleft palate, position the nipple so that his or her upper and lower gums connect with it.
- 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 that, as the baby suckles, you feel pressure against your finger.
- With gentle pressure under the chin, push up to start the baby's sucking.
- Your baby should feel comfortable while suckling (meaning no straining or squirming to access the nipple and to swallow).
- Maintain 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 to match your baby's suck-swallow reflex.
- Watch for cues that your baby is either full or needs burping.

What should my baby and I do right after feeding?

- Keep your baby upright for about 20 minutes after you are finished with feeding. This can be accomplished either by holding the baby or placing him or her in a seat.
 - If you place your baby in bed, slightly turn him or her to the side with some type of "wedge."
 - Elevate the bed by 20 degrees, so that the baby's head is higher than his or her stomach.
- If your baby has a cleft palate, he or she may exhibit what is called esophageal or nasopharyngeal reflux—meaning milk either comes out of the nose or is regurgitated shortly after feeding has ended. Always keep a suction bulb handy in case this happens.
- Record the time, length and amount of milk consumed in the feeding.

How can I ensure my baby is gaining sufficient weight?

- Weigh your baby once a week. If he or she is not gaining more than one ounce per day, you should talk to your pediatrician about increasing the calories in the milk.
- Calories can easily be added by concentrating formula or adding powdered milk to breast milk. Your pediatrician or cleft nursing team can advise you on the right approach.
- If your baby has still not gained enough weight even after increasing calories, your pediatrician may recommend an appointment with a gastrointestinal or nutritional specialist.

Are there ways to supplement or replace oral feeding?

Yes. When oral feedings are not enough to meet your baby's nutritional needs, there are other methods that can be used to deliver the needed calories:

- Sometimes, a device called a naso-gastric tube—a tube connected to a special syringe—is inserted through the child's nostril down into the stomach. This is a temporary solution to help the baby absorb enough calories until he or she can do so by mouth.
- Another type of tube used in feeding is a gastrostomy tube, which is inserted directly into the stomach from the outside of the belly. This procedure requires anesthesia, and is used in more serious situations when a child has trouble with breathing as well as eating or is at risk of aspiration (inhaling food, liquids or other materials into the lungs and trachea). As a child grows and learns to eat by mouth, the gastrostomy tube can be removed.

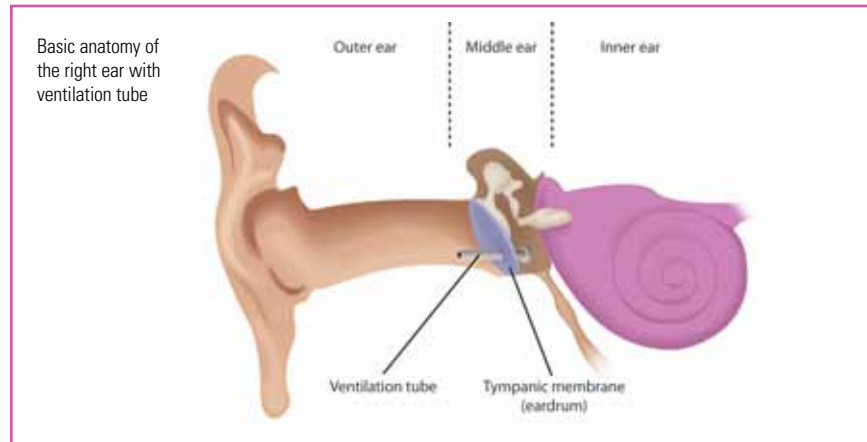
How can I ensure that my baby is properly nurtured, as well as nourished?

- It's essential to remember that your child is a normal baby who just happens to have a birth defect that can be surgically corrected.
- It is also very important that at least three people supporting you feel comfortable with feeding your child and can assist you as needed, because your infant will sense when someone is confident with feedings and will be more relaxed as a result.
- Once you feel comfortable with the feeding method recommended by your treatment team, you will find it easier to nurture your baby. You'll see him or her thriving, smiling and responding to your touch.

Hearing issues

Will my child have difficulty hearing because of the cleft?

A cleft lip does not affect hearing, but children born with a cleft palate often have temporary hearing loss because the tube that connects the middle ear to the throat (called the Eustachian tube) does not drain normally. Fluid in the middle ear space (also known as effusion) affects almost all babies under the age of 1 year with an unrepaired cleft palate, and can cause frequent infections—as well as hearing loss that can interfere with speech and language development.



How and when will my child's hearing be evaluated?

Your child's age and other health circumstances will determine which hearing tests are used, and when. In general:

- Very young babies—from newborns to 6-month-olds—usually have a special hearing evaluation called an auditory brainstem response (ABR). This test measures how well sound travels to the baby's brainstem.
- After the age of 6 months, babies may go through what is known as behavioral audiometric testing, which prompts them to turn their heads in response to certain sounds. Children between 7 months and 2½ years usually have a visual reinforcement audiometric (VRA) test, which uses special light-up toys and particular sounds to gauge how well they hear. A child who is between 2½ and 5 years old is likely to have an audiometric test, in which he or she will be asked to raise a hand in response to specific sounds.

- In addition to all of these tests, your child may also undergo tympanometric testing—an evaluation of middle ear function that can also check how well the ear tubes are functioning and can detect any signs of a hole in the eardrum.

Here at Boston Children's, one of our audiologists (hearing specialists) will evaluate your child's hearing every 6 to 12 months and will work closely with an otolaryngologist (ear, nose and throat specialist/ORL) to assist with any hearing issues related to the cleft.

What treatment is available for persistent fluid in the middle ear and associated hearing loss?

Yes. Babies with middle ear fluid and related hearing loss need an operation to drain the fluid and insert ventilation tubes—which are very important in protecting long-term hearing and allowing children to master normal speech. This operation is performed under general anesthesia and can usually be done at the same time as the cleft palate repair.

If your child has this procedure, the ventilation tubes will likely need to stay in place for nine to 12 months, with checkups by the ORL specialist every six months. The tubes usually fall out on their own, and so do not require another operation to remove them. There is a 50 percent chance that your child will need to have ventilation tubes re-inserted at some point.

Speech issues

Does a cleft lip affect speech?

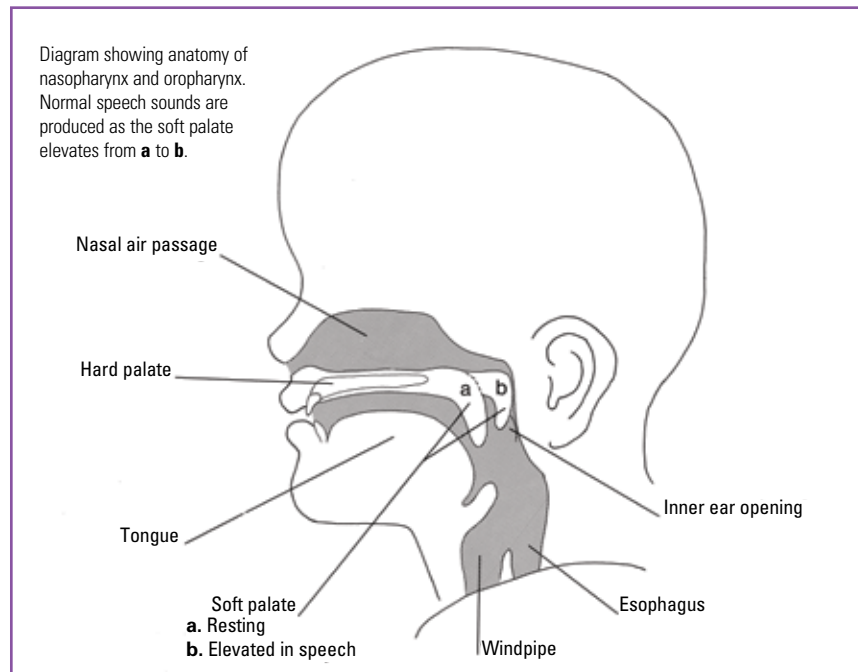
A cleft lip alone does not cause speech problems.

What about cleft palate?

The hard and soft palate separate the mouth from the nose. During normal breathing, the air flows in and out of a child's lungs through the nose and throat. During normal speech, the muscles in the soft palate move the palate to the back of the throat to seal off the nose (this process is called the velopharyngeal valve). Such movement allows the air to flow through the mouth, and only the mouth, during speech. By contrast, there are only three English speech sounds ("m," "n" and "ng") that are made through the nose.

A child born with a cleft palate will be unable to make normal speech sounds other than "m," "n" and "ng" until after the palate is closed.

The good news is that many of these patients acquire speech and language skills at a normal pace once the palate is closed and ear tubes are placed for drainage.



Will my child need speech therapy? What will this involve?

Here at Boston Children's, your child will be evaluated by one of our speech therapists shortly after cleft palate repair to determine whether further treatment is needed. If speech therapy is recommended, we will help you connect with services in your community.

Right after your child's palate is closed, you may notice that he or she vocalizes less often and in fewer ways. It may take up to six weeks to resume normal vocalization.

Your speech therapist will give you suggestions for home activities to help develop 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 (like mimicking a fish) and tongue-clicking games (mimicking a trotting horse). Sounds made in the throat, such as "uh oh," or animal roaring sounds should be discouraged; they can cause poor speech habits in children with a repaired cleft palate.

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

Will my child need other operations, like an adenoidectomy or tonsillectomy?

The adenoids naturally assist children in sealing off the nose during speech; they should only be removed after consultation with our Cleft Lip and Palate Program 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. Approximately 5 to 15 percent of children will not be able to adequately seal off their nose during speech, even after the palate is repaired. This results in hypernasal speech (too much nasal airflow), a problem called velopharyngeal insufficiency (VPI).

VPI will require another operation, known as a pharyngeal flap. When needed, this procedure is performed when the child is approximately 5 years old, following a complete speech evaluation by a speech therapist, otolaryngologist and plastic surgeon.

Dental issues

Will my child's teeth be affected by a cleft lip or palate?

Your child's first tooth (usually the lower incisor) will probably appear between 4 and 14 months of age. At 3 years, he or she will likely have all the primary (baby) teeth.

Children with a cleft lip and/or cleft palate often have poorly formed enamel (the outer tooth layer) on some of their teeth, especially those that are closest to the cleft. The teeth in this region may also be out of alignment and only partially erupted—and, therefore, difficult to clean. All of these factors will make your child's teeth more susceptible to cavities, so good dental hygiene is very important.

How should I keep my child's teeth clean and protected?

To minimize the risk of cavities:

- Brush your child's teeth at least twice a day. (Fluoride toothpaste should be used, but only in very small amounts.)
- Avoid foods with a lot of sugars and starches.
- Don't allow frequent snacking, which is especially harmful to the teeth since the bacteria in dental plaque produce cavity-causing acids each time food enters the mouth.
- Put your child on the fluoride dosage recommended by your dentist early in life. Fluoride, whether through the water supply or through prescribed supplements, has been proven to reduce the amount of decay in both baby teeth and permanent teeth. The greatest benefits from fluoride occur between 6 months and 8 years of age.

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

If your child has a complete cleft lip/palate and has needed any type of dental appliance, you probably have already met our dental team. Either way, your child should visit a pediatric dentist by the age of 12 months—even earlier if you have specific concerns.

What other dental issues should I expect as my child gets older, and how can these be treated?

Common problems can include missing, malformed or extra teeth in the area of the cleft. Absent teeth may need to be replaced by a dental implant once the child has finished growing, or by moving natural teeth into the space with orthodontics (braces).

As your child grows, his or her pediatric dentist and orthodontist (a dental specialist who corrects teeth and jaw alignment) will determine whether any additional treatment is needed.

Gum and jaw issues

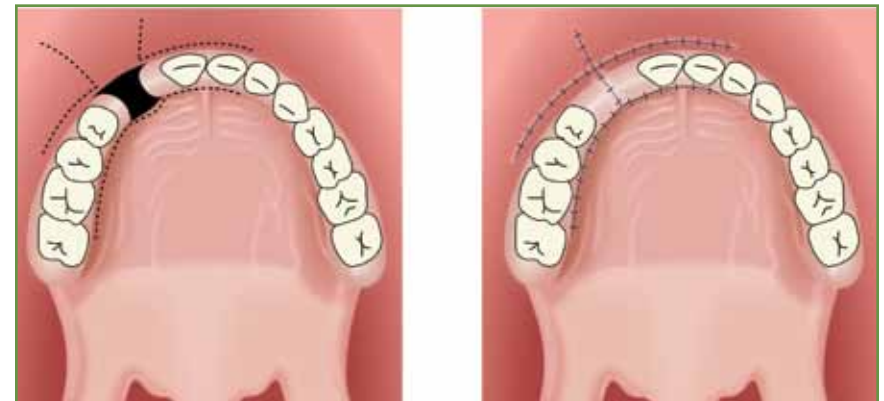
Will my child require orthodontic therapy (braces)?

Children with a cleft lip and/or palate almost always have some type of problem with their "bite" (teeth alignment), and most will need 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 who has special expertise working with children born with a cleft.

What will orthodontic therapy entail?

- Orthodontic therapy for children with a unilateral cleft lip/complete cleft palate is done in stages.
 - The first stage involves the insertion of a dental appliance called a Latham device. When your baby is between 3 and 6 weeks old, the pediatric dentist will take an impression of his or her gums and use the impression to make the device.
 - Between the ages of 6 and 12 weeks, your baby will have the appliance inserted by a pediatric dentist. This is done in the operating room under general anesthesia.
 - The appliance is then adjusted a little every day to slowly bring the gums closer together and make it easier to surgically repair the cleft lip.
 - After a period of 6 to 8 weeks, your baby's Latham device will be removed in the operating room shortly before the cleft lip is closed.
- Although the gum cleft is usually repaired at the time of lip repair, children whose cleft lip involves the alveolus (gum line) often need an initial phase of orthodontics before the cleft is closed.
 - For these children, the first phase of orthodontics involves an appliance placed in the maxilla (part of the upper jaw) to widen the palate before the alveolar bone graft procedure (see below).



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



Boston Children's Hospital
Until every child is wellSM