



**Boston
Children's
Hospital**

Until every child is well

Plastic and
Oral Surgery

Robin Sequence

A Guide for Families





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WELCOME!

Welcome to the Craniofacial Program at Boston Children's Hospital.

This guide is meant to give you an overview of the facial differences known as Robin sequence (or Pierre Robin sequence) and some related issues your family may face. It also includes helpful suggestions for locating the many resources available to you and your family.

In addition to providing the best possible medical and surgical care, our program is committed to supporting you throughout the treatment process. We are always here to address any questions and concerns.

Thank you for entrusting us with your child's care. We hope the following pages will inform and reassure you.

To schedule an appointment or speak to a member of our team, please call our program coordinator at **617-355-6309**.

Meet Our Team | Surgeons



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An Overview of Robin Sequence

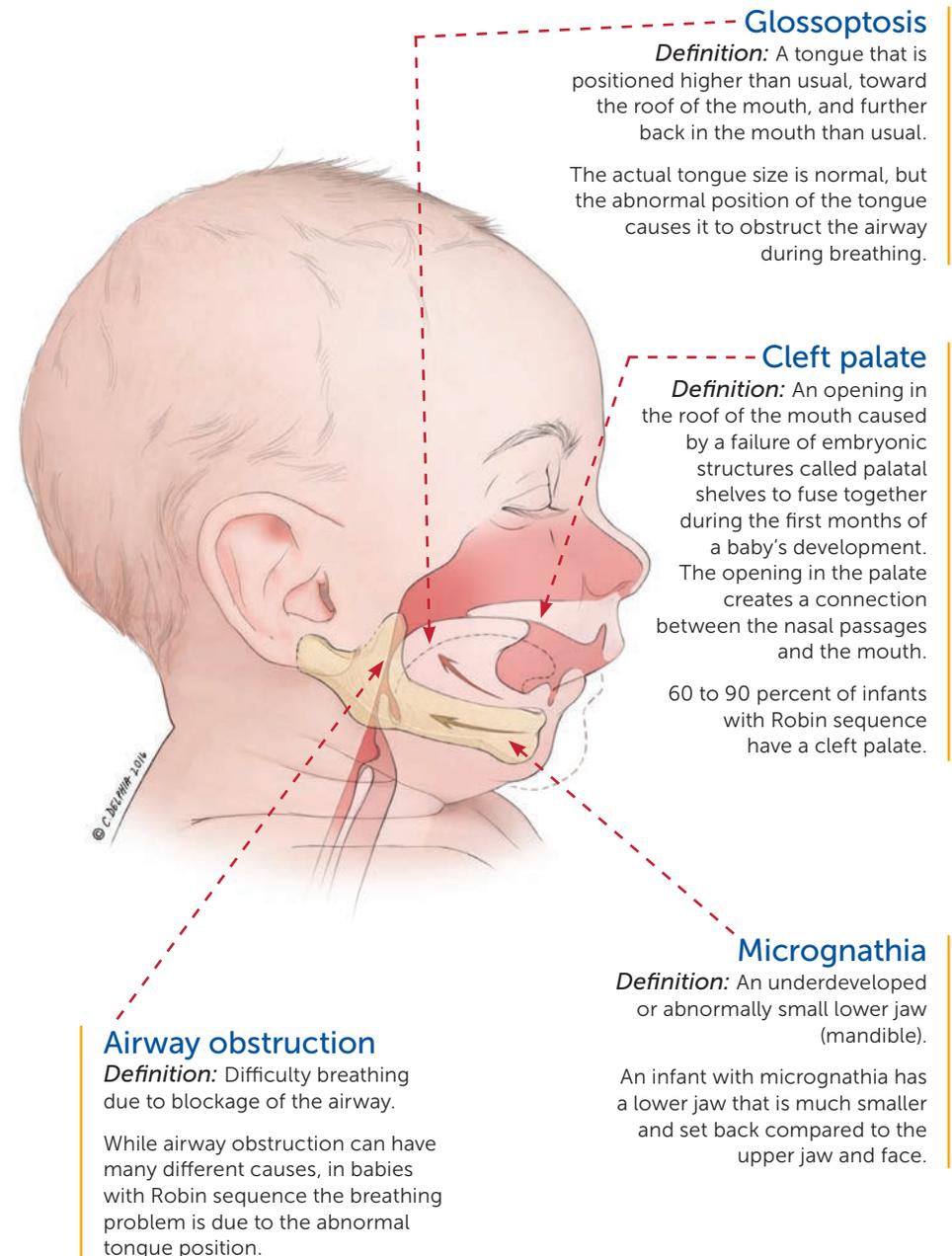
Robin sequence, also called “Pierre Robin sequence,” is a combination of facial differences that are present when a baby is born. It is described as a “sequence” because the initiating feature, a small, underdeveloped lower jaw (micrognathia), is thought to be the cause of the other key features: glossoptosis (abnormal positioning of the tongue in the mouth) and breathing difficulty due to airway obstruction (blockage of the airway). A cleft palate is also present in 60 to 90 percent of babies with Robin sequence.

At 7 to 10 weeks into a pregnancy, a baby’s lower jaw typically grows rapidly. This allows the tongue to descend from between the two halves of the palate, or roof of the mouth. If the lower jaw does not grow properly, the space is too small and the tongue is forced to the back of the mouth. This abnormal position of the tongue can block the airway, making breathing difficult, and can also prevent the palate from closing, resulting in a cleft palate.

Robin sequence affects 1 in 8,500 to 14,000 births, making it one of the most common facial differences. About 60 percent of babies born with Robin sequence develop the condition as a result of a malformation syndrome that may also affect other organs and tissues in the body; this is known as syndromic Robin sequence. The other 40 percent of babies have non-syndromic Robin sequence, meaning they have no other associated findings.

Robin sequence causes problems with breathing and feeding early in life. As a result, some affected babies have difficulty gaining weight at the expected rate. While many babies with Robin sequence will improve with time and can be cared for without surgery, some babies will need an operation early in life.

Features of Robin Sequence



Diagnosis and Early Care

Features of Robin sequence are sometimes identified before birth by prenatal ultrasound or MRI. While these tests provide information to help make a baby's delivery as safe as possible, the diagnosis of Robin sequence is not made until after birth when the baby can be examined by a clinician. Evaluation and care will then involve a coordinated team approach.

Breathing and feeding

Because the tongue blocks the airway, an infant with Robin sequence may have difficulty maintaining proper oxygen levels while lying on his or her back. Airway obstruction can also cause difficulty with breast and bottle feeding because infants cannot coordinate their labored breathing with the need to suck and swallow. Low oxygen levels may become apparent during the first several feedings, and poor coordination of

feeding and breathing can cause the infant to choke and sputter while feeding. Many babies with Robin sequence will therefore need feeding tubes to consume enough calories for weight gain. It is common to admit a baby to the Neonatal Intensive Care Unit (NICU) for oxygen monitoring and assistance with feeding.

Airway evaluation

Infants with Robin sequence will often have a polysomnogram, a study in which the baby is monitored during sleep to determine the frequency and severity of the airway obstruction ("sleep study"). The airway anatomy will also be evaluated with a video-scope or direct visualization to determine the site of obstruction and to rule out other contributing airway anomalies.

Treatments for Robin Sequence

Non-operative interventions

The first priority in caring for babies with Robin sequence is relief of their airway obstruction. Many babies can get relief with non-surgical therapies.

Positioning

Placing the baby in a side-lying position or prone position (on the belly) helps to pull the tongue forward. This can sometimes alleviate airway obstruction. Infants should be continuously monitored with an oxygen saturation monitor or apnea monitor while in these positions, both in the hospital and at home.

Nasopharyngeal airway

Some babies will benefit from a small tube inserted into the nose and down the pharynx (or throat). The tube prevents the tongue from falling back and blocking the airway.

Positive pressure ventilation

Continuous positive airway pressure (CPAP) can be delivered via a nasal or facial mask. Generating a low continuous air pressure helps the collapsible airway stay open.

Intubation

Intubation, or placement of a tube into the airway to assist with breathing, is sometimes necessary after birth. An operation may later be necessary to allow this breathing tube to be removed.



Patients with a tracheostomy

Treatments for Robin Sequence

Operative interventions

Tongue-lip adhesion (TLA)

In this operation, the tongue is pulled forward and stitched to the lower lip. This helps to keep the tongue out of the airway until “catch up” growth of the lower jaw helps to permanently open the airway. The TLA is usually reversed when a baby is between 8 to 12 months of age, and this can be combined with cleft palate repair when indicated. Results of TLA are mixed, with an overall success rate of about 50 percent. For that reason, we recommend this procedure for only a small number of patients.

Tracheostomy

A tracheostomy is the placement of a tube into the trachea (windpipe) to allow the flow of air to bypass the upper part of the airway. This is very effective in relieving breathing obstruction in babies with Robin sequence as it eliminates the need to breathe through the obstructed area. In some patients with additional medical problems and breathing problems that are not primarily caused by obstruction at the tongue base, tracheostomy may be the best option.

Q: *How can I feed my baby after the tongue-lip adhesion?*

The priority in the first week is to let your baby rest and to let the incision sites heal. To help with feeding, your baby will either have a small tube placed through the nose down to the stomach (an NG-tube) or surgery to place a tube through the belly wall into the stomach (a G-tube). Your baby will not be able to feed by mouth for seven days after the TLA.

On day 7 after the operation, you can begin to use a syringe and offer small amounts of milk by mouth at each feeding. This way, you can slowly build up the volume your baby can take by mouth. The majority of nutrition will still be given through the tube at this point.

About three weeks after the operation, if your baby is breathing well, you will have an appointment with the cleft/craniofacial feeding nurse to reintroduce bottle feedings. It can take several weeks to months for your baby to feed entirely by mouth. The cleft/craniofacial nurse will stay in close communication to support you and get updates on how your baby is doing. The feeding tube will remain in place until your baby shows all feeds can be taken by mouth. If your baby has a cleft palate, the tube will typically remain until after the palate repair.

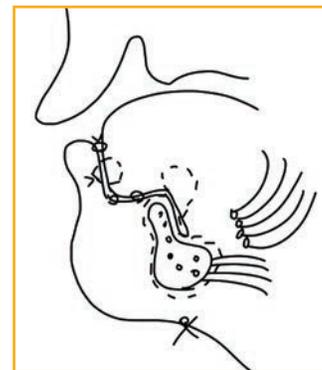


Illustration of tongue-lip adhesion



From pre-operative to 10 days post-operative over the course of Mandibular Distraction Osteogenesis

Mandibular distraction osteogenesis (MDO)

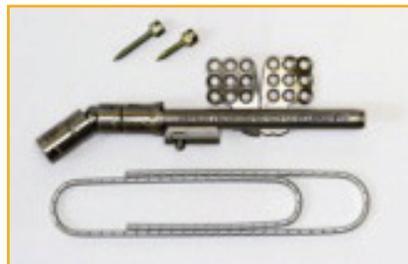
This technique uses the body's natural healing potential to elongate bones. MDO involves surgically separating the jaw in two places (one bone cut on each side) and applying corkscrew-like devices—one on each side—to gradually move the bone edges apart over a period of 7–14 days. This lengthens the jaw in a forward direction, bringing the tongue base forward and relieving obstruction of the airway.

The corkscrew devices are buried under the skin during the operation and are accessible by a turning pin under each ear. A special screwdriver is then used to turn the device a little bit each day until the jaw has been advanced as much as necessary. Babies usually remain in the hospital during this active turning process. The turning pins, also known as activation arms, are then removed easily at the bedside, and babies often can go home if they are breathing well. The corkscrew devices remain in place for several months to allow the newly formed bone to heal.

Compared to TLA, MDO has been shown to be more predictable in improving breathing and avoiding the need for tracheostomy in patients with Robin sequence. Oral feeding also may improve after MDO, and fewer babies need a G-tube after MDO compared to TLA.



Patient after mandibular distraction device is surgically inserted



The implanted distraction devices are the size of a paper clip and are secured with screws that are 1 mm in diameter

Q: How do I decide if MDO is the best operation for my baby?

This can be a difficult decision and all clinical parameters and options must be carefully weighed with your care team. We would only consider MDO after exploring non-surgical options.

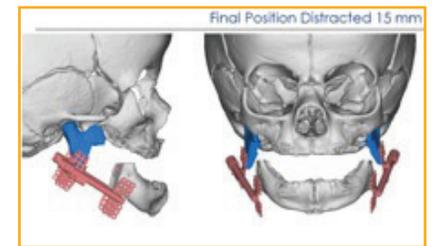
Q: How likely is MDO to work?

The success rate for relieving airway obstruction with MDO is about 90 percent in children who do not have a syndrome or other medical problems. Success rates are lower for babies who have Robin sequence as part of a genetic syndrome.

Q: What are the risks of MDO?

While MDO is generally a safe operation, every procedure has some risks. The short-term risks include post-operative pain, facial swelling and bruising of the skin. The most common risk is a low-grade infection around the turning pins, which can typically be managed with wound care and antibiotics. There is a risk of damage to nearby anatomic structures, including developing tooth buds, the nerves that move the corners of the lower lip and the nerves that provide sensation to the lower lip and chin.

There is a small chance of device failure or breakage, possibly requiring additional operations. In the long term, there is a possibility that the operation will impair future growth of the jaw and that future procedures will be required to further advance the lower jaw.



Virtual surgery is first performed using a CT scan of your baby in order to reduce risk and anesthetic time in the operating room

Q: If we choose to proceed with MDO, what do we have to do to prepare?

To plan the MDO procedure, we will obtain a CT scan of your baby's face. This may require sedation with or without placement of a breathing tube. Information from the CT scan will then be used to create a 3-D model of your baby's face, allowing us to simulate the operation. This pre-operative planning improves accuracy, minimizes operating time and helps avoid damage to important structures such as developing teeth and nerves.

Q: *Once we decide to proceed with MDO, when is the best time to do it?*

The timing for MDO is individualized to your baby's needs. Some babies undergo MDO within the first few weeks of life; others when they are much older. Your baby's breathing pattern will change naturally over the first few days and weeks of life, so we first make sure that your baby truly needs this operation. Once arrangements have been made to proceed, it typically takes two to three weeks to plan and schedule the operation.

Q: *Will my baby need a breathing tube for the operation and how long will that tube stay in?*

Yes, a breathing tube will be inserted at the beginning of the operation if one is not already in place. The breathing tube will stay in for some period of time after the operation. How long the tube remains depends on your child's progress after the operation, and ranges from three days to the entire length of the active distraction (7 to 14 days). While the breathing tube is in place, your baby will be kept comfortable in the intensive care unit.

Q: *Will turning the device hurt my child?*

Advancing the distraction devices is simple and most babies tolerate it very well. In fact, the average baby is bothered more by the turning of the head necessary to access the activation (turning) pins than by the activation itself.

Q: *Who will be responsible for turning the devices?*

Your surgeon will discuss options for turning the device with you. Typically, to involve family caregivers as much as possible, we suggest that you take responsibility for turning the device or designate someone to do so.

Q: *Does my baby have to go back to the operating room to have the turning pins removed at the end of the turning process?*

No, your surgeon will remove the pins very simply at the bedside. This process is not painful and does not require anesthesia.

Q: *What is the process for removing the devices?*

The distraction devices must be removed two to three months after they are placed. This is done in the operating room under general anesthesia. A breathing tube is required for this operation, but is typically removed at the end of the procedure. Most babies stay in the hospital the night of the operation and go home the next day.



Feeding a Baby with Robin Sequence

Your baby's ability to feed by breast or bottle depends on the severity of the breathing problem and whether or not there is a cleft palate. Your care team and our specially trained craniofacial nurses will help you determine whether it's safe to attempt feeding by mouth, and if so, what type of feeding is best for your baby. In some cases, a temporary feeding tube will be placed through your baby's nose to safely provide nutrition and calories. This may be done either instead of or in addition to feeding by mouth.

If your baby has a cleft palate, he or she will not be able to generate suction well, and will usually need a specialty bottle for feeding. The two specialty feeders we find

most successful are the Medela SpecialNeeds® feeder (also called the Haberman bottle) and Dr. Brown's® Specialty Feeding System. These bottles are designed to help your baby feed easily through compression of the nipple and do not require suction to express milk. They may also be useful for babies with Robin sequence without cleft palate who have feeding difficulty.



Q: Can I breastfeed my child?

Most babies with Robin sequence will not be able to breastfeed before their breathing problems are corrected. After breathing problems have been improved, the presence of a cleft palate may still make it difficult for your baby to create the suction necessary to express milk directly from your breast. You can, however, provide breast milk to your baby by pumping and giving it by bottle.

Q: 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, 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 the suck/swallow/breathe pattern. When the baby takes a long pause, use that time to burp the baby.

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

- Wrap your infant in a blanket, enclosing the hands. This is called swaddling.
- Sit in a comfortable chair, like a rocking chair or a glider chair, with a footstool.
- Hold your baby upright in your arm or hold his or her head from behind.
- Relax both arms and place the nipple gently into your baby's mouth.
- Lay the nipple on top of your baby's tongue.
- Put your ring finger or small finger under your baby's chin.
- With firm pressure, keep your finger in place so that, as your baby suckles, you feel pressure against your finger.
- With gentle pressure under the chin, push up to start your 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 if using the SpecialNeeds/Haberman Feeder, 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.

Q: What should my baby and I do right after feeding?

- Keep your baby upright for about 20 minutes after you are finished with feeding, either by holding 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 your baby’s head is higher than his or her stomach.
- A baby with a cleft palate may exhibit what is called gastroesophageal reflux (a tendency to have “heartburn” or to spit up after feeding) or pharyngonasal reflux (meaning milk or food comes out of the nose during feeding or when spitting up). Always keep a suction bulb handy in case this happens.
- Record the time, length and amount of milk consumed during the feeding.
- Keep a log to track your baby’s progress.

Q: How can I ensure my baby is gaining sufficient weight?

- Your baby should be weighed once a week, typically during a visit to your pediatrician. 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.

Q: Are there ways to supplement or replace oral feedings?

Yes. A device called a nasogastric tube (NG-tube)—a tube connected to a special syringe—can be inserted through the child’s nostril down into the stomach. This helps the baby absorb enough calories until he or she can do so by mouth.

Another type of feeding tube is a gastrostomy tube (G-tube), inserted directly into the stomach from the outside of the abdomen. This procedure requires anesthesia and is used when a child has trouble with breathing as well as eating or is at risk of aspiration (inhaling food, liquids or other materials). It can be removed as a child grows and eats by mouth.

After an operation to correct the airway obstruction, it often takes time to learn to feed orally. Therefore, most babies are discharged from the hospital with either a NG-tube or G-tube in place. Many babies will eventually have these tubes removed as their oral feeding improves. We will teach you how to use the devices before your baby is discharged from the hospital.

Q: How can I ensure that my baby is properly nurtured?

Once you feel comfortable with your baby’s feeding method, you will find it easier to nurture your baby. You will see him or her thriving, smiling and responding to your touch.

It is also very important to have at least two to three people supporting you who feel comfortable feeding your child. Your infant will sense their confidence with feedings and will be more relaxed as a result.

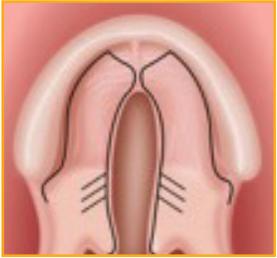


Patient with a NG-tube

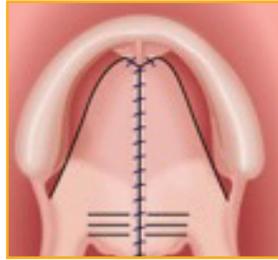
“I honestly cannot say enough great things about Dr. Resnick. He was the most responsive and caring doctor I’ve ever met. You can tell he truly cares about his patients.

Our friends tell us they couldn’t do what we’ve done. And I tell them, ‘Yes, you could, because you don’t have a choice. If your child needs you, you’ll be there.’”

~ words from a parent



Unrepaid palate



Repaired palate

Repairing a Cleft Palate

Q: How is a cleft palate repaired?

A cleft palate is typically repaired surgically to optimize speech development and correct pharyngonasal reflux. During the operation, the surgeon brings together the separated muscles and tissue from the two halves of the palate. This closes the opening and puts the muscles in a better position for speech.

Q: When will my child’s cleft palate be repaired?

Babies usually have the repair operation at around 10 months of age, but timing may range from 8 to 12 months. For babies with Robin sequence, the timing of cleft palate repair will depend on how well your child is breathing and gaining weight. It is ideal to repair the palate by 12 months of age, before a baby’s first attempts at speech, but only if it is safe to do so. Safety of your baby’s airway always takes first priority.

» Please refer to our [Cleft Lip and Palate brochure](#) for further information about cleft repair.

Further Resources and Support

Resources at Boston Children’s Hospital
bostonchildrens.org

Cleft and Craniofacial Center
617-355-6309

Contact a nurse
617-355-4513

Center for Families
617-355-6279

The Center for Families helps families locate information and resources. You are welcome to use the Center’s services at no extra cost.

Clinical social worker
617-919-6177

Outside Resources

pierreroobin.org

cleftline.org

facesofchildren.org

Insurance Information

Figuring out insurance coverage can be a confusing and sometimes difficult process to navigate. Ask your child’s insurance provider which procedures are and are not covered. It will also be beneficial for you to ask what referrals you might need. Most providers list a customer service phone number on the back of the insurance card.

Our administrative staff will be available to guide you through the insurance process once a plan of care is decided.



Patient one year post-op



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