

## PIERRE ROBIN SEQUENCE

### What is Pierre Robin Sequence?

Pierre Robin Sequence (pronounced Ro-BEN) is a birth condition that involves the jaw, the tongue, and the roof of the mouth.

At about 7-10 weeks of pregnancy, the lower jaw usually grows quickly, leaving a space at the bottom of the mouth for the tongue. Pierre Robin Sequence (PRS) starts when the lower jaw doesn't grow enough, pushing the tongue to the roof of the mouth and back toward the throat. In this position, the tongue often blocks the roof of the mouth (the palate) from forming properly, leaving a hole called a cleft palate (Pierre Robin Sequence is not associated with cleft lip). This "sequencing" of events during pregnancy is why the condition has its name.

### Could a child with PRS have other health problems?

Most babies born with Pierre Robin Sequence have no other health problems. This form is called isolated. Some babies have health problems beyond Pierre Robin Sequence such as a genetic syndrome. Two common genetic syndromes associated with PRS are *Stickler Syndrome* and *22q Deletion Syndrome*. It is important that babies born with PRS be evaluated by a team of professionals to identify any other issues (For more information on team care, see below.)

### How common is this condition?

Pierre Robin Sequence happens once in 5,000 births in isolated form and once in 30,000 births as part of a syndrome. In comparison, cleft lip and/or palate occurs about once in every 700 births.

### What health issues can be expected?

Pierre Robin Sequence varies from child to child. One person may have severe problems related to the jaw, tongue, and palate. Another may experience some, but not all of those issues. Also, it is possible, but uncommon, to have Pierre Robin Sequence without a cleft palate.

There are a few health issues commonly related to PRS. The small jaw associated with Pierre Robin Sequence can cause problems with breathing. Because the tongue rests at the back of the mouth, close to the throat, it can fall back and block the airway.

Pierre Robin Sequence can also affect eating. A baby with a cleft palate is unable to seal off the mouth from the nose. Because air and milk leak out of the hole, the baby is unable to breastfeed or drink from a regular baby bottle.

A child born with Pierre Robin Sequence can also have problems with hearing. If a child has a cleft palate, fluid may build up behind the eardrum, causing short-term hearing loss (if a child has a syndrome or another medical condition, hearing loss may be permanent). As a baby grows, any hearing loss can affect speech and language. A cleft palate can affect speech in other ways, too, even after it is repaired during infancy.

### Who treats Pierre Robin Sequence?

The issues linked with Pierre Robin Sequence are complicated but they can be treated. A child needs to have care from many types of professionals. It is important to find a *cleft palate* or *craniofacial team*, a group of professionals who work together to evaluate a child's needs and plan his or her care. A cleft palate team includes a surgeon, a speech-language pathologist, an orthodontist, and access to other specialists. A craniofacial team treats patients with conditions beyond cleft lip and palate; it includes a craniofacial surgeon and access to a psychologist and other specialists. Both types of teams treat patients with Pierre Robin Sequence.

### How is the condition treated?

The treatment for Pierre Robin Sequence varies from child to child, depending on symptoms and growth. The most important issues to treat during infancy relate to the airway (breathing) and to feeding. The airway can be managed by physical positioning, with *nasal airways* (soft tubes placed in the nostrils), or with observation and waiting for growth. In some cases, a baby may need surgery. Feeding a baby with PRS usually requires a special bottle and help from a feeding specialist. (Treatment for each issue is described in further detail, below.)

Late in infancy, a child born with a cleft palate will need to have surgery to close it. At around that time, the child's team will also recommend an evaluation for hearing and possible placement of hearing tubes in the eardrum. As a child grows, he or she will need to be evaluated by a speech-language pathologist and possibly receive speech therapy.

Treatment usually begins after birth and can last through childhood and into the teen years. It is critical that a child with Pierre Robin Sequence be treated by a cleft palate or craniofacial team.

### How are jaw and breathing issues treated?

During infancy and early childhood, most children with Pierre Robin Sequence have some trouble breathing. Early on, a baby needs a full exam. Members of the child's team may do x-rays, CT scans, a sleep study, and a *scope* of the airway, an exam performed with a small camera to check for specific problems. The team may also examine how a child swallows.

Many babies do not need surgery to help with breathing. Instead, team members usually ask parents to take special precautions at home, particularly during a baby's sleep time. A baby with Pierre Robin Sequence needs to sleep on his or her stomach (*prone*). Prone positioning lets the tongue fall forward and open the airway. An adult needs to monitor the baby to make sure the airway stays open. Babies with Pierre Robin Sequence should NOT sleep on their backs.

If breathing issues are severe, doctors may recommend surgery. About one in three babies born with Pierre Robin Sequence needs surgery during the first year. The goal of surgery is usually to open the airway by pulling the tongue forward in the mouth. A surgeon either stitches the bottom side of the tongue to the lower lip (called a *tongue-lip adhesion*) or lengthens the lower jaw (called *mandibular distraction*). Beforehand, a baby may need a special breathing tube.

For some newborns with other medical problems, tongue-lip adhesion or mandibular distraction does not do enough to help with breathing. In these cases, a child's team may suggest a *tracheostomy*, a procedure to place a breathing tube directly into the windpipe through the neck.

For some children with PRS, jaw issues self-correct with time. A small lower jaw may grow quickly during early childhood. By about age six, the jaw may reach a normal size. Some children do not experience this "catch-up" growth, however. These children may need surgery to increase the size of the jaw later in childhood or in the teen years.

### How are sleep issues treated?

Some children with Pierre Robin Sequence have *sleep apnea* during childhood or the teen years. Sleep apnea is a disorder in which breathing stops and starts repeatedly during sleep. The ear, nose and throat surgeon (*otolaryngologist* or *ENT*) on the team may ask parents to look and listen for signs, such as snoring and long spaces between breaths. Sleep apnea can be treated with special breathing equipment called CPAP, or in some cases, with surgery to enlarge the jaw.

### How do I feed my child?

Babies born with a cleft palate are able to have skin-to-skin time with a caregiver but are unable to form the suction necessary to breastfeed or drink from a regular

baby bottle. Babies born with cleft palate should drink formula or pumped breast milk from a special bottle.

Parents often need help feeding a baby with PRS. Right after birth, a feeding specialist on a child's team meets with families. The specialist shows parents how to use a special bottle, how to hold the baby while feeding, and how to read the baby's signals. Most babies born with PRS feed successfully with one of these bottles. They usually need to be held upright for feeding or in a side-lying position. For more information on feeding a baby with a cleft palate, visit the ACPA Family Services website.

In rare cases, a baby can't get the nutrition he or she needs, even from a special bottle. In these cases, a member of the team may insert a feeding tube into the baby's nose or stomach. Infants with a feeding tube should continue to see the feeding specialist for as long as the feeding tube is in place. In certain cases, such as for children with tracheostomies, a feeding plan may be more involved.

Sometimes, a baby with Pierre Robin Sequence will stop wanting to eat or drink. This is called *oral aversion*. It can happen after a baby has had a lot of medical interventions or is tube-fed. It is important for tube-fed babies to suck on a pacifier during their tube feeds so that they can learn that they need to suck to eat and so they can practice sucking skills. The pacifier (or another kind of non-nutritive sucking) has other benefits. It can calm a baby. It also keeps the tongue and jaw in a good position for breathing.

While some of the interventions mentioned above may be uncomfortable for a baby, they are needed and should not be stopped.

Some babies are born with a *tongue-tie*, a short band of tissue that connects the tongue to the bottom of the mouth (any baby can be born with a tongue-tie; it is not related to PRS). While doctors often recommend cutting a tongue-tie for a baby without PRS in order to help with feeding, surgeons usually recommend keeping it in place for a baby with PRS. Cutting a tongue-tie in an infant with Pierre Robin Sequence usually does not help with feeding. In fact, the tie may help keep the tongue in an optimal position. Without the tie, the tongue will be more likely to move into the airway. Eventually, a tongue-tie can be cut, but the judgment should be made by a surgeon who regularly treats children with this condition.

Feeding a child with Pierre Robin Sequence can be challenging. In most cases, though, feeding gets better with time and special care. A specialist can help families with bottle feeding, and later on, with solid foods.

### How is a cleft palate treated?

*Palate repair surgery* is a surgical procedure to close a cleft palate in order to help a child with eating and speech. As a baby grows, the team will decide on the best time for this surgery. In many cases, the team recommends performing this procedure somewhere between ages 9 and 18 months.

If a baby is having problems with hearing and/or frequent ear infections, the ENT on the team may insert tubes into the ear-drum at the time of palate-repair. Tubes can treat the temporary hearing loss until the palate begins to function properly.

### How are hearing and speech issues treated?

All babies born in the U.S. have a hearing test at birth. When a baby is born with Pierre Robin Sequence, he or she will need follow-up care by members of a cleft palate or craniofacial team. An *audiologist* will perform further hearing tests as a baby grows. A pediatrician and an ENT surgeon will check for ear problems. As stated above, the ENT treats temporary hearing loss.

Because children with Pierre Robin Sequence can have problems with speech, they need to be seen by a *speech-language pathologist*, an expert who assesses, diagnoses, treats, and helps prevent problems with communication and swallowing. The speech pathologist may suggest speech therapy or other treatment.

### Will future children be affected?

Many parents wonder if Pierre Robin Sequence will happen in future pregnancies. A person with the condition may wonder, too, whether it could happen for his or her future children.

The chances of Pierre Robin Sequence happening again depend in part on how it was caused. PRS can be caused by genes, by an event during pregnancy, or both.

A *geneticist* is an expert on how characteristics are passed down in families (*inherited*). The geneticist on a child's team can do blood tests to learn about a person's genes. Sometimes, those tests show a *chromosomal change* or *genetic mutation* in a patient with Pierre Robin Sequence, which may indicate the presence of a genetic syndrome in that person (some examples of related syndromes include Stickler syndrome, 22q11 deletion syndrome, and Treacher Collins Syndrome). Knowing about a syndrome affects a person's treatment. It also tells the geneticist and family about the chances of Pierre Robin Sequence happening again.

Parents who have had a child with isolated Pierre Robin Sequence (without a syndrome) probably have a higher risk over the general population of having another child with this condition. This is because they may have genetic characteristics related to PRS that they could pass on to another child.

While Pierre Robin Sequence can be passed on from a parent to a child (inherited), it can also occur with no family history. Scientists have not done enough studies to know the chances of this happening.

Sometimes, a child has Pierre Robin Sequence because of an event during pregnancy. This manifestation is called *environ-mentally induced*. Knowing certain information about that pregnancy can help the geneticist predict the chances of Pierre Robin Sequence happening again.

It is very important for people with Pierre Robin Sequence and their relatives to see a geneticist. A geneticist can help families learn about future pregnancies. He or she can also find out if a person has a syndrome that needs medical care now.

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### For more information, or for a referral to a cleft or craniofacial team, please contact:

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