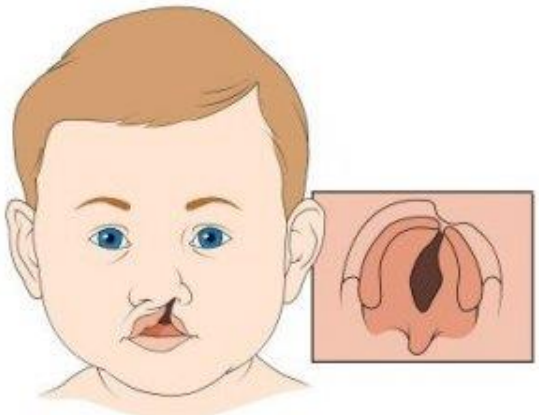
Cleft lip and palate (orofacial cleft)

Orofacial clefts are birth defects where the mouth or roof of the mouth (palate) do not grow together properly during development. Oral clefts occur in 1 in 700 live births.



A separation or gap in the lip is called a cleft lip. The separation may be on only one side of the lip (unilateral) or on both sides of the lip (bilateral). The cleft is called incomplete if the cleft does not extend into the nose, or complete if the cleft extends into the nose. Unilateral cleft lip is more common and usually affects the left side. Cleft lip occurs more commonly in boys.

An opening in the roof of the mouth is called a cleft palate. Although a cleft palate may occur alone or along with with a cleft lip a cleft lip occurs with a cleft palate in more than three-fourths of cases.

The incidence of cleft lip with or without a cleft palate CL[P] varies by racial and ethnic group. CL[P] occurs in about 1 in 500 births in people of Native American and Southeast Asian origin. Whites and Hispanics have lower rates of about 1 in 1000 births. African Americans have the lowest rate of about 1 in 2000 births.

Cleft palate occurs in about 1 in 2000 live births and does not appear to be as influenced by racial background and ethnicity as does cleft lip with or without cleft palate.

What Causes Cleft Lip And Cleft Palate?

Although the cause of most cases of cleft lip and cleft palate are unknown some cases may be caused by errors in inherited factors (genes) passed on to the affected child from their parents.

Environmental factors may also play a role. Exposure during pregnancy to phenytoin (a medication used to control seizures), corticosteroids , smoking , and binge drinking (5 or more drinks per occasion-weekly) are believed to increase the chances that a baby will be born with a cleft. Mothers taking multivitamins containing folic acid seem to be less likely to have babies with oral clefts.

In some cases, a cleft lip and/or palate may be associated with a collection of other findings that suggest a specific disorder (syndrome). When a cleft lip or palate are found on a prenatal ultrasound examination a search for skeletal defects and heart defects will usually be performed to evaluate the fetus for a possible syndrome. An amniocentesis may be recommended for further evaluation, since additional findings in some syndromes may otherwise go undetected prenatally..

Hundreds of syndromes have been associated with a cleft lip and /or palate. Some of the more common conditions include van der Woude syndrome, 22q11.2 deletion syndrome, Pierre Robin Sequence, Stickler syndrome, trisomy 13, and trisomy18.

Will it happen again?

The likelihood that cleft lip and/or palate will recur in a future child can be as high as 50% if the cleft is part of an autosomal dominant syndrome.

If there is no identifiable cause the risk of an isolated cleft lip with or without cleft palate recurring in a future child is 3 % if one child and neither parent has it, 3% if no child but one parent has it, 8 % if two children and neither parent has it, 11% if one child and one parent has it , 19% if two children and one parent has it, and 34 % if no children have it but both parents have it..

What sort of problems may I expect?

Feeding Problems

The first problem you may encounter may be with the feeding of your baby. A cleft lip or palate makes it difficult for the baby to get good suction during breast or bottle-feeding. Gently squeezing the infant's cheeks together around the nipple may improve oral suction. In addition the baby's sucking and swallowing reflexes may not work correctly, and the baby may swallow a lot more air during feeding. Holding the baby in an upright position can make feeding easier. Burping the baby frequently, after every ounce of formula is also helpful. Infants with only a cleft lip or a narrow cleft palate can usually breast-feed. Infants with both cleft lip and cleft palate will usually need to be bottle fed using a crosscut nipple. Specialized squeezable bottles designed for babies with cleft palate and other special feeding problems are available.

The Mead-Johnson Cleft Palate Nurser shown at right includes a long soft nipple with a crosscut hole attached to a flexible plastic bottle that can be squeezed to increase the flow of liquid during the baby's sucking.

The Haberman Feeder shown below is designed for babies with cleft palate and other special feeding problems. The Haberman nipple is larger and longer than most nipples. The nipple can be squeezed to help the baby extract the liquid. Air is squeezed out of the nipple before the feeding begins.

Moms should stay alert for signs of dehydration in the baby. If the baby becomes very sleepy, has no tears when crying, dry lips, dark urine, or no wet diapers for more than 6 hours moms should call their baby's doctor for advice. If the baby shows signs of severe dehydration such as sunken eyes the baby may need to be taken to the emergency room.

Dental Problems

Children born with with clefts may have special problems related to missing, malformed or malpositioned teeth in the area of the cleft. Evaluation by a dentist who is familiar with the needs of children with a cleft is recommended.

Ear Infections

Children with cleft palate are more susceptible to ear infections because of abnormal development of the muscles that control the opening and closing of the Eustachian tubes (or auditory tubes) The Eustachian tubes connect the middle ear to the throat above the palate. The Eustachian tubes allow secretions to drain from the middle ear and function to equalize the air pressure on both sides of the eardrum.

Repeated ear infections can lead to permanent hearing damage which can effect the normal development of speech. Ear tubes (also called myringotomy or tympanostomy tubes) may be placed in order to improve drainage from the middle ear.

Psychological

Children with clefts have an increased risk for developing problems especially those relating to self concept, peer relationships, and appearance. Family therapy, support groups, and group discussions about cleft lip and palate with the child's teachers and classmates are often helpful.

Speech

Cleft palate is associated with the abnormal development of the muscles that control the soft palate (velum) . When the soft palate does not successfully close off the nose for the oral sounds a hypernasal quality of speech occurs.

Even after palate repair proper closing of the soft palate muscle during speech may not return entirely to normal. Children may have difficulty producing the sounds "p, b, d, t, h, k, g, s, sh, and ch", and speech therapy is often helpful.

When Can The Cleft Be Fixed?

Cleft lip is usually repaired at 3 months of age. Cleft palate is usually repaired at 9 to 12 months of age.

Revision is sometimes needed at 4 or 5 years of age. Some children with very large gaps in the hard palate may also need to have the gap filled with bone when they are 4 to 11 years of age. Additional corrective nasal surgery may be required when the child's nasal growth is complete.

The many problems encountered during the care of children with orofacial clefts are typically managed by a Cleft Team that usually includes a pediatrician, a surgeon, a dentist, a speech therapist, an ear-nose-throat specialist, a psychologist, a social worker, and a genetic counselor.

Online Resources