

CLEFT LIP AND PALATE

Description

Clefts, the most common birth defects affecting the face, are separations occurring in the lip or the palate (roof of the mouth), or both. The cleft can be situated on one side of the lip (unilateral) or both (bilateral).

Approximately 80% of cleft cases are unilateral. Cleft palate can occur along with a unilateral or bilateral cleft lip or with a normal lip.

A cleft lip may simply be a notch in the upper lip (incomplete) or more extensively when the gap extends upward into the nostril (complete). Similarly, a cleft palate can either be complete or incomplete. Complete cleft palates have the opening or split extending from the (front) hard palate to the (back) soft palate. This means there is an opening between the mouth and the nasal cavities. An incomplete cleft palate is denoted by a gap in the front of the roof of the mouth most often extending from the upper gums to the area just to the rear.

Children with cleft lip and palate often have a flattened nose and spread lips. Dental abnormalities frequently accompany clefts with teeth being either deformed, absent or extra. Mild ocular hypertelorism (abnormally wide space between the eyes) and abnormal cartilage growth surrounding the nose is also noted.

Prevalence/Causes

The most common congenital deformity of the face, clefts occur in 1:600 births. In children of Asian descent, the frequency rises to 1:500 live births and in African-American populations it falls to 1:2000.

Cleft lip appears twice as often amongst males than females while females are more likely to be born with cleft palates, though the actual cause is unknown.

What *is* widely documented is that the risk increases to 1:20 for a subsequent child being born with a cleft if an older sibling has the condition.

It is recommended that given the dramatically increased risk for subsequent children being born with clefts, a geneticist be consulted.

Treatment

Feeding: Because of the air space between the child's mouth and nose through the cleft in the palate, the infant cannot suck effectively making breast feeding very difficult. If the cleft is of the lip only, breast feeding is usually successful.

The use of a collapsible bottle with a longer nipple and a large crosscut opening, which allows parents to control, the flow of milk can help.

Prior to Surgery: A custom-made device, similar to a retainer or dentures, is placed in the baby's mouth. Over the course of the next few weeks, this device gradually pulls the edges of the cleft closer so that an optimal lip repair can be achieved. The closer the sides of the cleft, the less tension there will be on the surgical repair and the better the ultimate cosmetic result. This device facilitates feeding as it seals off the cleft in the roof of the mouth curtailing the flow of liquids into the nose.

First Surgery: Cleft lip (uni- or bilateral) is closed during the first surgery. At the same time, the wide nostril, elevation of the flattened nasal tip and gum gap are corrected. A bilateral cleft lip may require a second procedure that lengthens the skin on the bottom of the nose between the nostrils (columella).

Cleft Palate Repair: In order to maximize facial growth and minimize the cleft's impact on speech, palate repair is usually done between 12-14 months of age. A speech pathologist monitors verbal development at about 10 months and recommends repair when specific elements of speech are noticed.

Cleft repair involves sewing the palate edges together and reconstructing the muscle in back of the palate so that speech is maximally effective.

Speech Therapy: Because the nose is not sealed off in a child with cleft palate, speech can be typified by hard to understand hypernasal sounds. If a child learns to talk *before* palate repair, abnormal speech mechanisms are developed to compensate for the air leaking out of the nose. Although most

children develop normal speech if surgery is performed at the appropriate time, some may need speech therapy to help them unlearn compensatory speech behaviors. In some cases, children, despite surgery, cannot seal off their nose when speaking; it is then that another surgical procedure may be indicated. A “pharyngeal flap” (strip of tissue from the back of the throat) is used to partially block communication between the mouth and nose.

Dental: Almost every child with a cleft palate will need orthodontic therapy as the teeth closest to the cleft site often come in incorrectly. Orthodontic work can begin when the child is as young as 4 years old if he/she is cooperative. In some cases, bone grafting at around age 8 is required to allow adult teeth to come in properly. In this procedure, small pieces of bone are taken from the hip or outer surface of the skull and placed in the gap between bone edges so they can adhere to solid bone.

Later Surgery: During teenage years, some patients choose to have their noses touched up. At the same time, most children with a unilateral cleft lip and palate have a deviated septum which makes nasal breathing more difficult. This can also be corrected during adolescence.

When earlier surgical repair of the palate restricts growth of the upper jaw and orthodontic therapy does not work because the lower jaw is actually in front of the upper, another surgical procedure, maxillary advancement, may be required.

Associated Conditions

- Increased frequency of colds
- Fluid in the ears often leading to an increase in ear infections
- Tonsil and adenoid problems
- Sore throats
- Speech difficulties arising from open connection between nose and mouth
- Teeth located in the area of the cleft may be absent or improperly placed affecting appearance and chewing.

Psychosocial

It is important to remember that children perceive surgical outcomes differently from adults and this difference extends well into the teen years. Therefore, despite the fact that there may be a good cosmetic repair, the child or teen with the cleft may feel that he or she still looks significantly different. Parents need to be supportive of the right of the child or teen to have these feelings while offering reality testing so that perceptions may become more realistic.

As children age into teenagers, the desire for touch up cosmetic surgery may arise as the importance of good physical appearance becomes dominant. These issues should be discussed openly within the family and with the physician.