

Cleft Lip & Palate in New Mexico

Information for Parents and Providers

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INTRODUCTION

Cleft lip & palate is a developmental condition affecting newborn babies. The exact cause is unknown. Most cases occur sporadically with no family history or other anomalies. The goal of treatment is to provide the child with as normal an appearance, speech and life as possible. To accomplish this, a multitude of specialists are involved at different stages of life from birth to infancy, childhood, adolescence and adulthood.

INCIDENCE OF CLEFT LIP AND PALATE IN NEW MEXICO

Cleft lip and palate is one of the most common birth defects. It occurs in approximately 2.6 per 1,000 births in New Mexico. For unknown reasons, the incidence is much higher in our state, possibly due to ethnic and genetic factors. Based on epidemiological studies, there are approximately 40 to 50 children born per year in this state with clefts. The majority of these undergo treatment with Dr. Cuadros at Presbyterian Hospital with the largest clinic in the state, having treated hundreds of children since 1989.

Of all clefts

- 25% will have associated syndromes
- 20% will be high risk
- 25% will involve cleft lip only
- 50% will involve cleft lip and palate
- 25% will involve cleft palate only

DIAGNOSIS AND CLASSIFICATION

When a baby is born with a cleft, it is important to establish a precise diagnosis. Children will appear to have missing tissue or often be mistaken to have a midline defect. Closer inspection will reveal the nature of the cleft. Tissue is misplaced, but not necessarily missing.

Questions to ask:

- Is the defect unilateral or bilateral?
- Does it involve the lip alone, palate alone, or both lip and palate?
- Is the defect complete or incomplete?
- Are there any associated syndromes?

GENETICS AND RISK OF CLEFT PALATE

- If this is **first child** with cleft,
 - The overall risk for another sibling or offspring = **4%**
- If **more than one** immediate family member is affected,
 - The overall risk for another sibling or offspring = **10-16%**

EMBRYOLOGY

Clefts occur during the first stages of embryological development as the face is forming by the fusion of various elements. Failure of fusion is the cause of clefts of both the lip and palate.

PRIORITIES in CLEFT MANAGEMENT

At birth:

- **Coping • Adapting • Feeding • Preparing for surgery •**

After birth:

- **Appearance • Speech • Hearing • Dental •**

FEEDING

Initially, feeding may be difficult due to the inability to maintain intra-oral negative pressure due to the open connection between the oral and nasal cavities. Breast feeding may not be possible when the palate is involved. Some mothers are successful but may need to supplement with breast pumping or formula. Feeding the baby may take a period of trial and error. Most mothers are successful with a soft nipple cross cut at the end or side attached to a Mead-Johnson squeeze bottle. Other products include Pigeon Nipple, NUK nipple, Haberman feeder. These feeders allow the baby to gum the nipple and drip or squeeze milk or formula. Be creative and resourceful. Infant should be fed in the slightly upright position. Air swallowing can be prevented with more frequent and slower feedings. Monitor weight and feeding closely. A feeding specialist can be very helpful. All babies have different abilities, likes and dislikes. The urge to feed is strong, and the best technique is quickly determined. Sometimes it requires patience, time and determination but eventually all babies will learn to feed. Babies need time to learn and to strengthen the muscles of the mouth, tongue and throat.

Unilateral Cleft Lip

In the unilateral cleft lip, the defect involves only one side, extending from the gum, through the lip and into the nose. In some cases, a small band is present at the base of the nose, sometimes called an “incomplete” cleft lip. The muscles of the mouth and lips are split and do not form a sphincter so when the baby cries, the lip actually opens up. In addition, there is a gap in the bone of the maxilla through the gum and gingiva. This is addressed when the child is 7-8 years old. The columella of the nose is smaller and the entire nose can appear displaced or crooked including a deviated septum. These nasal deformities are corrected later.

Bilateral Cleft Lip

The bilateral defect is the most severe form of clefting. In this deformity, there are two gaps extending from the gum to the nose. The central part of the lip, called the prolabium, has nothing to hold it back so it tends to protrude. After repair, the prolabium will mold back into a more normal place. The orbicularis muscle of the mouth is widely split and will be realigned in the midline at the time of repair. The nasal deformity is greater with a small columella and displaced nostrils.

Variations of cleft palate:

Submucous Cleft Palate - A condition in which the uvula is split and it appears that the palate is intact, when, in fact, the muscles and underlying structures are split. Repair is often necessary if there is a risk of abnormal speech development.

Robin Sequence - A condition in which babies are born with an excessively short mandible or jaw, along with a very wide cleft of the palate, often called a “U-shaped” cleft. These babies sometimes require respiratory and feeding support shortly after birth. As the child grows, the mandible tends to grow as well. Cleft palate repair may be delayed until later.

FAMILY COUNSELING -

It is imperative that a member of our team meet with the parents as soon as the baby is born to assist in feeding. During consultation we review various aspects of clefts and show before-and-after pictures in preparation for the first surgery.

ULTRASOUND DIAGNOSIS

Many mothers are now finding out about their baby's cleft while still pregnant. With newer techniques in ultrasound, the details of the face can be determined. In this case, I will meet with the mother and family before birth and provide support and information. Knowing what to expect can be very helpful.

SURGERY in the FIRST YEAR OF LIFE

The two most important surgeries will take place in the first year of life. These operations are to correct the lip and palate. Timing and technique are critical.

Repair Cleft Lip - Age 3 months

The lip is repaired when the baby is 2½ to 3 months old. Performing surgery at this age insures that the immune system, airway, respiratory system and blood counts are strong enough to tolerate surgery and anesthesia.

The goals of surgical correction are to create a pleasing harmonious lip and eliminate the stigmata of cleft lip deformity, avoid secondary deformities and additional surgeries. This is done by using the triangular flap repair developed by Tennison and Randall. The technique involves reconstituting lip height, realignment of the orbicularis muscle, recreating the Cupid's Bow of the lip by aligning the white roll, creating a vermillion tubercle, releasing and realigning the nostril while maintaining an adequate labial sulcus. Sometime minor revisions are necessary at a later time.

Surgery takes 3 hours and includes closure of the most anterior portion of the hard palate with vomer flaps. An otologist usually inserts PE tubes in the ears at this time. Baby stays in the hospital with the mother 2 nights, sutures are removed in 7 days in the operating room.

Repair Cleft Palate - Age 9 months

Studies have shown that 9 months is the ideal time for cleft palate repair in terms of better speech results and fewer complications. This is done before meaningful speech patterns develop and healing is complete by the time the baby begins to form words. At this stage, the entire palate is closed. Vomer flaps are used to close the anterior palate at the initial lip repair.

The goal of palate surgery is to restore normal anatomy in order to achieve as normal speech as possible and to eliminate the stigmata of cleft palate hypernasal speech. The main defect associated with the cleft palate is an open communication between the nasal and oral cavities. Mucosal deficiency is present and the palate is shortened with the muscles of the soft palate displaced. They are split, retracted and have an abnormal insertion onto the posterior margin of the palate rather than in the midline. Repair consists of realignment of speech muscles. Most commonly used technique is the V to Y lengthening and repositioning with mucoperiosteal flaps. (Wardill-Kilner-Veau). This is done by mobilizing flaps from the sides. This lengthens the palate, closing the gap while repositioning the muscles.

FACIAL SKELETON and DENTAL ABNORMALITIES

Children with clefts exhibit a wide variety of facial skeleton and dental abnormalities from malocclusion to gross maxillary deficiency. These defects consist of hypoplastic maxilla on cleft side and malalignment of dento-alveolar arches. With a bilateral cleft, the defects are more severe and the premaxilla often grossly deficient in bone. Dental abnormalities include supernumerary teeth, dystrophic teeth, congenitally missing teeth and malocclusion in nearly all patients. Ectopic tooth buds may be present as infants. The most common orthodontic deformity involves a crossbite usually occurring on the side of the cleft. Surgical treatment with alveolar bone graft is performed by an Oral and Maxillofacial surgeon between ages 7-9 years to allow permanent teeth to grow. A maxillary expander is placed prior to bone graft. When the permanent teeth erupt, orthodontics is then applied to realign the teeth. If maxillary retrusion or severe malocclusion occur, further orthognathic surgery may be required at age 15-18 years.

HEARING ISSUES

Virtually all patients with a cleft involving the palate (with or without cleft lip) will develop middle ear effusions due to abnormal drainage of Eustachian tube. If untreated, this can lead to chronic ear infections and permanent hearing loss. Therefore, all patients with clefts are evaluated early by an Otologist (an ENT who specializes in hearing) for the need for ventilation tubes (PE tubes or BMT). The majority will demonstrate fluid build up and require tubes to be placed at the time of lip and/or palate surgery. With this aggressive approach, most children can expect normal hearing.

SPEECH OUTCOMES

After the repair of the cleft palate, the child will gradually begin to speak, first with a few words and then with sentences. This is the critical time for development. A speech therapist will monitor for any signs of language, speech and hearing problems. If there is any question of speech problems, the child may be referred for further testing including videofluoroscopy and nasendoscopy. Speech results are better with early recognition of surgically correctable problems. The most common problems we see are language delay and velopharyngeal insufficiency, or VPI. VPI is a result of abnormal healing or scarring of the palate, and may need a second operation to correct.

SECONDARY SURGERY

- Not all procedures turn out perfect the first time. Minor revisions or corrections are sometimes necessary for the lip, palate and nose.
- After full skeletal maturity in the teenage years, the final operation to be performed is a complete rhinoplasty and septoplasty to reshape the nose.
- Major revisions may be required at any age, especially if the surgery was done elsewhere and there exists a significant problem with either the appearance of the lip or with palatal function.

ANESTHESIA, HOSPITALIZATION AND RECOVERY

Babies and children with clefts are managed by an experienced team of pediatric specialists. All surgery is done at Presbyterian Hospital due to the availability of state of the art care. Anesthesiologists use specialized techniques making the procedures safe with good recovery. For most surgeries, baby or child will remain hospitalized for one to two nights, until feeding has returned to normal. Infants will be fitted with soft restraints on the arms, they will be fed with a special feeding bottle called a Zip-n-Squeeze. For lip repairs, sutures are removed the following week by placing the baby under light anesthesia. All palate repairs are done with dissolving sutures, and no suture removal is required. A small pack is placed in the palate, which will fall out on its own. After surgery the baby will be admitted with IV fluids, antibiotics, and small doses of narcotics for pain. When the baby can feed fully, he or she will be switched to oral medications. Pain after surgery is well controlled in this way, but many babies will be fussy and irritable. Drainage and low grade fever is normal after surgery. Most bottle feeding can be resumed after one week. You will be given detailed instructions on wound care, diet and follow-up. Recovery and healing generally takes 3 weeks.

SPECIALISTS INVOLVED IN CLEFT CARE:

Anesthesiologist	Nutritionist	Pediatric Surgeon
Audiologist	Neurosurgeon	Pediatric Intensivist
Dental Hygienist	Nurses	Pediatrician
Developmental Specialist	Obstetrician	Pedodontist (Dentistry)
Dietician	Operating Room Nurses	Perinatologist
Dysmorphologist	Ophthalmologist	Plastic Surgeon
Feeding Specialist	Oral Maxillofacial surgeon	Psychologist
Genetics Counselor	Orthodontist	Radiologist
Insurance Case Manager	Otolaryngologist	Social Worker
Lactation Specialist	Otologist	Speech Pathologist
Neonatologist	Parent Volunteer	

CLEFT PALATE CLINIC AND TEAM

As part of ongoing care and evaluation, all children and families with clefts are invited to attend the Cleft Palate Clinics. These outreach clinics meet throughout the year and are attended by group of specialists. The purpose of the clinics is to gather the patients and specialists in one location in order to coordinate. The cleft palate clinics are managed by Children's Medical Services (CMS) through the NM Department of Health. Children are followed from birth to 21 years.

INSURANCE AND PAYMENT ISSUES

Dr. Cuadros and the members of the cleft palate team are committed to taking care of all children with clefts regardless of payment source. Dr. Cuadros is contracted with all major insurance carriers, HMO's, Salud programs, CMS and Medicaid. It is my philosophy that there should be no barriers to offering these children the best care possible.

RESOURCES for PARENTS and HEALTH CARE PROVIDERS
for immediate information call 1-800-24-CLEFT

LOCAL RESOURCES:

Dr. Luis Cuadros, MD, FACS, Plastic Surgery.

1-505-243-7670, or toll free in New Mexico 1-888-217-6120. Our office is available at any time to discuss issues regarding cleft management. We will arrange for consultation, provide feeding information and supplies. Dr. Cuadros has been in practice in New Mexico since 1988. He is a graduate of Columbia University and trained at Harvard Medical School. He is a member of the American Society of Plastic Surgeons and American Cleft Palate Association.

Susanne R. Hays, MS, RN, CRRN. Pediatric Rehab Nurse and Feeding specialist: 1-505-294-8338. Able to consult by phone or home visit for all feeding issues in newborns and babies.

Presbyterian Ear Institute

415 Cedar St. S.E, Albuquerque, NM 87106

Telephone: (505) 224-7020, Fax: (505) 224-7023

Email: pei@oraldeafed.org

<http://www.oraldeafed.org/schools/pei/index.html>

Children's Medical Services - Cleft Palate Clinics (CMS)

CMS offers evaluation, diagnosis and treatment for congenital deformities using an interdisciplinary team approach.

CMS Statewide Office - Toll Free 1-877-890-4692

Albuquerque - District I CMS Office - 897-5700 or 897-3311

Farmington - San Juan County Health Office - 505-327-7606

Gallup Health Office - 505-722-6830

Santa Fe County Health Office - 505-476-2603

Dr. Dan Shover - Pediatric Gastroenterology - 563-6550- Presbyterian Feeding Clinic - Nutritionist Cindy Chall Silva. If your baby is having trouble gaining weight, your pediatrician can refer you to this clinic.

INSTITUTIONAL AND ORGANIZATIONAL RESOURCES

CleftLine: Managed by the Cleft Palate Foundation. 1-800-24-CLEFT . Call this number 24 hours/ 7 days to obtain information on clefts.

<http://www.cleftline.org/>

WideSmiles: organization begun by a parent with three adopted cleft children.

PO Box 5153, Stockton, CA 95205-0153

email WideSmiles@aol.com

<http://www.widesmiles.org>

American Cleft Palate-Craniofacial Association/Cleft Palate Foundation

1504 East Franklin Street, Suite 102, Chapel Hill, NC 27514-2820 USA

(919) 933-9044 ,

info@cleftline.org

<http://www.acpa-cpf.org/>

Prescription Parents - support group with cleft lip and palate information.
45 Brentwood Circle, Needham, MA 02492
617-431-1398.

<http://www.samizdat.com/cleft.html>

A Parent's Guide to Cleft Lip & Palate - University of Minnesota Guides to Birth and Childhood Disorders. Moller, Starr and Johnson. Univ of Minn Press. 1990. Available from Dr. Cuadros and from <http://www.amazon.com>.

The Cleft Advocate -providing educational opportunities, on- and off-line support networks, social interaction and advocacy guidelines, cleftAdvocate gives families the tools they need to obtain the absolute best medical care from their craniofacial team and the best service from their insurer, while encouraging children, teens and adults with cleft lip and/or palate and other craniofacial anomalies to reach their highest level of self-esteem.

<http://www.cleftadvocate.org>

The Cleft Palate-Craniofacial Journal is an international, interdisciplinary journal reporting on clinical and research activities in cleft lip/palate and other craniofacial anomalies, together with research in related laboratory sciences. Official Publication of the American Cleft Palate-Craniofacial Association (ACPA)

<http://cpcj.allenpress.com/cpcjonline/?request= index-html>

PRODUCTS

Zip-n-Squeeze bottles

<http://www.zip-n-squeeze.com/softsipp.html>

Arm restraints: Pedi-Wrap, PO Box 5398, Hemet, CA 92544. Tel 909-925-8800.

<http://www.medi-kid.com/>

Pigeon Nipples. PIGEON has developed the nursing bottle which enables a baby to suck in the same manner as they would from a mother's nipple by studying the shape, size, elasticity and structure of the nipple.

<http://www.galtak.com/nipples.htm>

Haberman Feeders

Specialized feeders designed for babies with facial or oral problems that hamper their ability to nurse.

<http://www.medela.com/New Files/specialtyfdg.html>