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Current Comments®

Cleft Lip and Palate: Good News, Bad News

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Sometimes when a human embryo is growing, something goes wrong, and an opening, or cleft, occurs in the lip, the palate (the roof of the mouth), or both. (The word *cleft* is derived from "to cleave.") Cleft lip with or without a cleft palate is one of the world's most common birth defects. It occurs in about one of every 700 to 800 births.[1] These figures roughly hold true worldwide for Caucasians. According to the American Cleft Palate Educational Foundation (ACPEF), more males than females are born with a cleft lip (with or without cleft palate). Among blacks, the incidence is somewhat lower—about one in every 1,000 or 1,300. Among Orientals, the incidence is slightly higher.[2]

ACPEF is a nonprofit organization which provides information to parents of children born with clefts. ACPEF can tell you which treatment centers exist in your area. The organization also publishes a number of free booklets. It is affiliated with the American Cleft Palate Association (ACPA), a professional organization of physicians, dentists, speech pathologists, audiologists, psychologists, and others. Similar organizations exist in other countries. For example, there is the Cleft Lip and Palate Association of Sweden in Akensberga, and the Canadian Cleft Lip and Palate Family Association in Toronto.

Speech pathologist Hughlett L. Morris, University of Iowa, Iowa City, points out that cleft palate presents a "good news-bad news" situation. The good news is that cleft palate can be repaired and its victims can lead normal lives. The bad news is that not everyone with cleft palate is reached, and that even in cases of successful repair, victims and their parents may have trouble adjusting.[3]

Cleft lip is sometimes called "harelip," a term now considered inaccurate and somewhat insensitive. According to Blair O. Rogers, New York University School of Medicine, the term may be derived from cleft lip's resemblance to the mouth of a rabbit.[4] In her 1924 novel, *Precious Bane*, Mary Webb tells the story of an early nineteenth-century woman born with cleft lip. Among the problems the woman faces is the accusation of witchcraft. When it was reprinted in 1978, literary critic Michelene Wandor wrote a preface to the novel, pointing out the superstition that children with "harelips" are born to women who, when pregnant, were frightened by the devil, who had assumed the shape of a hare.[5]

If clefting sparked superstition, it also challenged the ingenuity of the early surgeons. According to Rogers, cleft palate is not mentioned in the works of ancients such as Hippocrates or Galen. However, ancient Chinese surgeons may have known how to repair clefts. And the Arabian surgeon Albucasis (936-1013 AD) used needles and sutures to repair facial injuries. He may have used them to fix clefts. Saxon surgeons, colloquially known as "leeches," recom-

mended in 950 AD that the doctor "cut with a knife the false edges of the lip," sew the lip with silk, and apply a mastic and egg-white salve.[4]

According to Rogers, the first person known to have described cleft lip repair in detail was the fourteenth-century Flemish surgeon Jehan Yperman.[4] Flemish plastic surgeon J. Vrebos, St. Jean Hospital, Belgium, notes that Yperman so scrupulously described the details of needle-and-suture surgery that some of his advice is worthy of a modern plastic surgery text. Yperman even counseled surgeons to do a good job lest their reputations be compromised.[6]

Yperman also speculated on the origin of clefting. He debunked the idea that it was caused when the mother ate hare or red mullet during pregnancy. He pointed out that many of the mothers who bore children with clefts never ate those foods. He did believe, though, that clefting and other birth defects were sometimes caused by "the imagination of the mother during sexual intercourse." However, Yperman also considered more modern explanations for clefting's origins. He assumed, correctly, that heredity and the health of the parents are important factors.[6]

Today, researchers agree that cleft palate is caused by a combination of heredity and environment. It is almost impossible to tell, however, where genetic effects end and environmental ones, such as drugs, disease, or radiation, begin.[7] Many agents cause clefting in animals. But only a few have been proved to cause it in humans. According to F.C. Fraser, McGill University, Toronto, Canada, these causes include the disease rubella and the drug thalidomide.[8]

According to ACPEF, the risk for parents of bearing a second child with cleft *lip* (with or without cleft palate) is about one in 20 if there is no other family history of cleft. Parents with one child born with cleft *palate* run a one in 50 risk of having another, according to Hermine M. Pashayan, Cleft Palate Clinic, Tufts-New England Medical Center, Boston, Massachusetts.[7]

Unfortunately, there's little that can be done to warn parents that their child may be born with a cleft. Between about the second and eighth weeks of pregnancy, the embryo is extremely sensitive to teratogens. Unfortunately, at this stage most women are not yet aware that they are pregnant.[9] From the sixteenth to twentieth week of pregnancy, an ultrasound scan of the fetus may detect the presence or absence of a cleft lip, according to G. Savoldelli and colleagues, University of Zurich, Switzerland. However, this is a relatively new and untried procedure for cleft diagnosis and is not done routinely.[10]

The uncertainties surrounding clefting's origins are one source of emotional stress for parents. Obviously, it's a shock to find one's newborn is abnormal in any way. Sometimes the shock is compounded by guilt.

The parents' emotional problems are only one facet of clefting. Treatment of the child itself is a multidisciplinary problem. It requires expertise in pediatrics, surgery, otolaryngology, audiology, speech pathology, dentistry, and psychology. For that reason, the optimum treatment of clefting is carried out by a multidisciplinary cleft palate team.[11] The first such team or clinic was founded in 1938 by the late Herbert K. Cooper. Located in Lancaster, just a short distance from Philadelphia in Pennsylvania Dutch country, it was long known as the Lancaster Cleft Palate Clinic. It's now called the H.K. Cooper Clinic.[12] According to Ellen Cohen, ACPEF, there are about 120 such clinics throughout the US, and about 40 others throughout the world.

Surgery can fix the cleft, but feeding the newborn patient is the immediate problem. Babies with cleft have difficulty sucking. In fact, milk may flow into the nasal cavity. Depending on the circumstances, there are different ways to facilitate feeding. A regular formula bot-

tle with a soft nipple and a somewhat enlarged opening usually helps, according to an ACPEF pamphlet.[11] Other kinds of feeding devices may also help. For example, Jack L. Paradise and Betty Jane McWilliams, University of Pittsburgh, reported good results with a "compressible feeder" that allows the parent to gently pump milk into the baby's mouth.[13]

According to ACPEF, infant cleft patients sometimes experience certain problems. They may inhale too much air and need to be burped more frequently. They may have trouble consuming thicker foods. Feeding should take about 45 minutes and occur every three or four hours. If feeding lasts too long, it might mean the child isn't getting enough nourishment. Enlarging the hole in the nipple usually helps, according to Pashayan and colleagues.[7]

Some doctors use feeding appliances. For example, Jerome A. Markowitz, Elmhurst City Hospital, Queens, New York, developed a device which fits into the baby's mouth to allow normal feeding. The custom-fitted "over-extended denture without teeth" is made from quick-setting acrylic after a wax impression is made of the baby's mouth. The prosthesis allows for easy breast- or bottle-feeding, and reassures the parents by allowing the baby to cry and gurgle normally. Markowitz says it seems to help close the cleft, presumably because it keeps the baby's tongue out of the opening and allows bone to form.[14]

It's important to keep in mind that no one feeding method works best for every infant. According to the Cooper Clinic, some babies will even feed best drinking from a cup. And some doctors report little need for special feeding devices, apart from the enlarged nipple hole. Position is also important. Perhaps the single most important aid to feeding is to hold the child upright and tilted slightly backward.[12]

Surgery for clefting is done fairly early. The object is to close the clefts to minimize scarring, provide a pleasing appearance, and to forestall speech problems. According to ACPEF, the lip is usually repaired within the first three months after birth. The palate is usually repaired later, perhaps between a year and 18 months of age. The precise timing depends on many factors, including severity of the cleft. To close the palate may require only one operation, or several may be needed. In addition, further operations may be required as the child grows.[2,11]

Much of the additional surgery involves steps to improve appearance. For example, often appearance will benefit from rhinoplasty, or reshaping of the nose. Usually this operation is delayed until adolescence, when the nose is fully formed.[2]

Unfortunately, patients are at risk for ear infection early in life. With a cleft, the muscles in the palate don't function properly. These muscles are needed to open the eustachian tubes. When they're malfunctioning, the tubes don't open effectively, and air can't enter the middle ear. When that happens, fluid accumulates in the resulting vacuum. This condition is called otitis media. The fluid may become infected and cause earache and fever. Otitis media's widespread occurrence in cleft patients was first reported in 1967 by Sylvan E. Stool and Peter Randall, Children's Hospital Cleft Palate Clinic, Philadelphia.[15] Two years later, Paradise and colleagues found it to occur in every one of 50 patients.[16]

The result of such problems is that the otolaryngologist is an important part of the cleft palate team. Otitis media may be cured by medication which dries up the fluid. Or the otolaryngologist might perform a minor surgical procedure called myringotomy. The doctor makes a small slit in the eardrum to drain fluid from the middle ear. Afterward, tiny tubes may be inserted in the slits to allow air to enter.[11]

Otitis media may recur, so children born with clefts need frequent ear ex-

aminations. Sometimes ear infections lead to hearing loss. Thus, audiological tests are also essential. Even children as young as three months can be effectively tested. The tests are important because hearing loss can go undetected. A case of otitis media may not lead to infection, and there will be no telltale symptoms such as earache.[1]

There is another reason regular ear checkups are crucial. Hearing loss can impair the child's ability to distinguish words. Thus, acquisition of language and vocabulary can be affected.[2]

The issue of speech acquisition is complicated by other problems that may occur in cases of cleft lip or palate. With proper treatment, and in the absence of developmental problems, the child will learn to speak like other children. But many children with clefts begin to speak a few months later. The child will learn sounds and perhaps words before the cleft is closed. As with any child, parents should encourage all early attempts at speech.[1]

A speech pathologist should be part of the cleft palate team. This specialist is needed to monitor the child's language development. Among the things he or she will observe are the child's articulation and tone quality. By the time the child reaches the age of three or four, the speech pathologist should be able to tell how much, if any, speech therapy is needed.[1]

Dental problems are also common among cleft patients. Several types of dental specialists may be needed from birth. These include pedodontists (children's dentists), orthodontists (to align and straighten the teeth), oral surgeons, and prosthodontists (to fit dental appliances, if needed).[1]

As mentioned earlier, the parents may need psychological counseling. Obviously, this holds true for the child as well. The child may undergo several operations and hospitalizations early in life. This can be traumatic, as can the fact of being born with a cleft. Counseling may be needed throughout adolescence. But this isn't always the case.[2]

Some of the best news about cleft victims is that, with the proper counseling and surgical treatment, they usually

Table 1: This entry from the 1981 *SCI® JCR™* shows the journals that most cited *Cleft Palate Journal* in 1981. The number to the left of each journal is its 1981 impact factor. The number at the right is the total number of citations given to *Cleft Palate Journal*.

.60	Cleft Palate J.	350
.60	Cleft Palate J.	98
1.05	Plast. Reconstr. Surg.	41
.12	Int. J. Ped. Otorhinolaryngol.	25
	J. Craniofac. Genet. Dev. Biol.	18
.41	Scand. J. Plast. Reconstr. Surg.	11
.67	Amer. J. Orthodont.	10
.66	Laryngoscope	10
	Sprache-Stimme-Gehoer	10
.35	Child Psychiat. Hum. Develop.	9
.63	Brit. Dent. J.	8
.45	Brit. J. Plast. Surg.	8
.45	Clin. Pediat.	8
1.55	Arch. Dis. Child.	7
	J. Commun. Disord.	6
.31	Otolaryngol. Head Neck Surg.	6
	All Others (53)	105

Table 2: This entry from the 1981 *SCI® JCR™* shows the journals that were most cited by *Cleft Palate Journal* in 1981. The number to the left of each journal is its 1981 impact factor. The number at the right shows the total number of citations in 1981 that *Cleft Palate Journal* gave.

.60	Cleft Palate J.	678
.60	Cleft Palate J.	98
1.05	Plast. Reconstr. Surg.	69
	J. Speech Hear. Res.	25
	Birth Defects—Orig. Art. Ser.	23
.67	Amer. J. Orthodont.	20
.72	J. Speech Hear. Disord.	15
.90	Arch. Otolaryngol.	14
.28	Angle Orthodont.	13
1.16	Amer. J. Dis. Child.	12
.45	Brit. J. Plast. Surg.	11
1.23	J. Acoust. Soc. Amer.	9
1.69	Teratology	9
.41	Scand. J. Plast. Reconstr. Surg.	8
1.55	Arch. Dis. Child.	7
.41	Acta Morphol. Neer. Scand.	6
	Communication	6
.42	J. Maxillofac. Surg.	6
2.59	J. Pediat.	6
.48	Oral Surg. Oral Med. Oral Patho.	6
2.53	Pediatrics	6
	All Others (216)	309

grow up to be well-adjusted, productive adults. There is no evidence that treated patients suffer severe and permanent psychological damage. Pediatric psychologist Lynn C. Richman, University of Iowa, however, reports that some teenagers are self-conscious about their appearance and do have some problems socializing.[17] Richman and Michele Eliason,[18] University of Iowa, also report that some cleft children, although they perform normally on intelligence tests, get lower grades. Part of the problem might be self-consciousness or speech or hearing difficulties. But lower grades might also be a result of lowered expectations on the part of teachers.

Little is known about how people with clefts adapt in their adult lives. McWilliams[19] notes that, despite lower grades in childhood and adolescence, cleft victims achieve higher educational levels than their fathers. However, they do end up with relatively lower incomes and lower-level jobs. This could be because of prejudice, or a result of lingering self-consciousness. McWilliams also says that adults with cleft have fewer and later marriages. They also have fewer children and postpone having them. Part of the reason may be fear of having a child with clefting.[19]

Edward Clifford, Duke University Medical Center, Durham, North Carolina, says that though these factors are problems, they "are hardly descriptions of pathology." Although we really know little about the life outcomes of typical cleft patients, most of them seem to do well, probably because of the treatment available now.[20]

Probably the most important single factor in successful treatment is awareness on the part of parents and physicians. ACPEF, in addition to its pamphlets for parents and patients, also publishes *Cleft Palate Journal*. Other journals that regularly publish cleft palate research include *Plastic and Reconstructive Surgery*, *Clinical Genetics*, and *Pediatrics*. These periodicals are covered in *Science Citation Index®*, *Current Contents®/Life Sciences*, and *CC®/Clinical Practice*.

The literature on cleft palate is interdisciplinary. This is illustrated by Tables 1 and 2. Table 1 shows *Journal Citation Reports®* (*JCR™*) listings of the journals that most frequently cite *Cleft Palate Journal*. Table 2 shows those journals

Table 3: A list of the ten most-cited articles published in *Cleft Palate Journal* from its inception in 1964 to the present.

Bluestone C D, Paradise J L, Beery Q C & Wittel R. Certain effects of cleft palate repair on Eustachian tube function. *Cleft Palate J.* 9:183-93, 1972.

Kitamura H. Epithelial remnants and pearls in the secondary palate in the human abortus: a contribution to the study of the mechanism of cleft palate formation. *Cleft Palate J.* 3:240-57, 1966.

Kremenak C R, Huffman W C & Olin W H. Growth of maxillae in dogs after palatal surgery: I. *Cleft Palate J.* 4:6-17, 1967.

Meskin L H, Gorlin R J & Isaacson R J. Abnormal morphology of the soft palate: I. The prevalence of cleft uvula. *Cleft Palate J.* 1:342-6, 1964.

Pruzansky S. Pre-surgical orthopedics and bone grafting for infants with cleft lip and palate: a dissent. *Cleft Palate J.* 1:164-87, 1964.

Skolnick M L. Videofluoroscopic examination of the velopharyngeal portal during phonation in lateral and base projections—a new technique for studying the mechanics of closure. *Cleft Palate J.* 7:803-16, 1970.

Skoog T. The use of periosteal flaps in the repair of clefts of the primary palate. *Cleft Palate J.* 2:332-9, 1965.

Spriestersbach D C, Dickson D R, Fraser F C, Horowitz S L, McWilliams B J, Paradise J L & Randall P. Clinical research in cleft lip and cleft palate: the state of the art. *Cleft Palate J.* 10:113-65, 1973.

Stool S E & Randall P. Unexpected ear disease in infants with cleft palate. *Cleft Palate J.* 4:99-103, 1967.

Warren D W & DuBois A B. A pressure-flow technique for measuring velopharyngeal orifice area during continuous speech. *Cleft Palate J.* 1:52-71, 1964.

that *Cleft Palate Journal* cites often. The tables show a variety of disciplines, including journals of pediatrics, communication disorders, teratology, otolaryngology, dentistry, and psychiatry. Table 3 lists the most-cited articles published in *Cleft Palate Journal*.

In a telephone conversation, Laird Jackson, Division of Medical Genetics, Thomas Jefferson University, Philadelphia, told us that cleft palate research is geared mainly toward the practical matter of treating the patient. It does not display a lot of citation activity, but it is part of teratology, which is an active discipline. According to Jackson, teratologists often look for cleft palate in laboratory animals as a marker indicating teratological effects of a particular substance.[21]

Society has come a long way since it linked clefts with witchcraft. But as social research on the effects of clefting indicates, some prejudice against people with clefts remains even after the clefts are repaired. Organizations like ACPEF and ACPA enhance progress by informing people about the treatment and, perhaps, changing people's attitudes about the condition. Both organizations can be contacted at 331 Salk Hall, University of Pittsburgh, Pittsburgh, Pennsylvania 15261, (412) 681-9620.

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3. **Morris H L.** The child with cleft lip and palate: 20 years of progress. *Int. J. Ped. Otorhinolaryngol.* 3:93-9, 1981.
4. **Rogers B O.** Harelip repair in Colonial America. *Plast. Reconstr. Surg.* 34:142-62, 1964.
5. **Wandor M.** Preface. (Webb M.) *Precious bane.* New York: Dial Press, 1978. p. 8-12.
6. **Vrebos J.** Jehan Yperman, medieval cleft lip surgeon. *Plast. Reconstr. Surg.* 70:762-5, 1982.
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13. **Paradise J L & McWilliams B J.** Simplified feeder for infants with cleft palate. *Pediatrics* 53:566-8, 1974.
14. **Markowitz J A, Gerry R G & Fleishner R.** Immediate obturation of neonatal cleft palates. *Mt. Sinai J. Med.* 46:123-9, 1979.
15. **Stool S E & Randall P.** Unexpected ear disease in infants with cleft palate. *Cleft Palate J.* 4:99-103, 1967.
16. **Paradise J L, Bluestone C D & Felder H.** The universality of otitis media in 50 infants with cleft palate. *Pediatrics* 44:35-42, 1969.
17. **Richman L C.** Self-reported social, speech, and facial concerns and personality adjustment of adolescents with cleft lip and palate. *Cleft Palate J.* 20(2):108-12, 1983.
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