

## The Changing Faces of Children with Cleft Lip and Palate

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Related article, page 769

Many expectant parents worry, consciously or unconsciously, that their baby will be born with a malformation. Cleft lip with or without cleft palate is one of the most common birth defects and is certainly the most visible. The incidence varies among ethnic groups, ranging from 3.6 per 1000 live births among Native Americans to 2.0 per 1000 among Asians, 1.5 per 1000 among Indians, 1.0 per 1000 among people of European ancestry, and 0.3 per 1000 among Africans. Cleft lip with or without cleft palate is more frequent among boys. In contrast, isolated cleft palate is twice as common among girls and occurs in approximately 0.4 of every 1000 live births in all ethnic groups.

Parents who have a newborn with a cleft lip desperately want to know why it occurred, and in their distress they may inquire about birth defects in distant relatives or even become suspicious of each other. Explanations by geneticists generally invoke multifactorial inheritance, meaning that a cleft results from a complex interplay between genetic and environmental factors. Indeed, as discussed by Zuccherro et al. in this issue of the *Journal* (pages 769–780), a number of mutations in specific genes have recently been identified. An interruption of the cascade of molecular events that normally leads to a coalescence between the medial nasal process and maxillary prominence at around four to six weeks of gestation may result in cleft lip, cleft alveolus, and nasal deformity. Embryologists call this anatomical site of initial closure the primary palate.

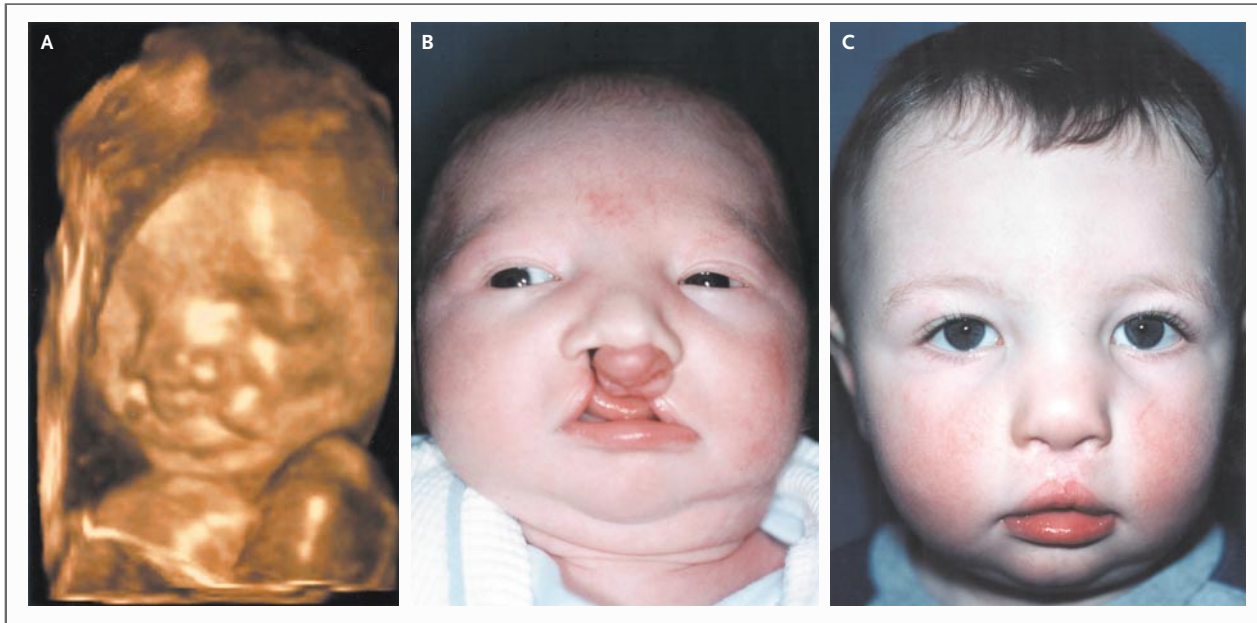
The palatine shelves emerge from the maxillary prominences during the sixth week of embryonic development and grow obliquely downward on each side of the tongue. Between 8 and 12 weeks, the palatine shelves become horizontal (in a matter of hours) and begin fusing with each other and the nasal septum to form the secondary palate. Cleft palate results from a disturbance in the elevation, fusion, or mesenchymal penetration of the palatine shelves.

A cleft in the fetal lip may be visualized with the use of transvaginal ultrasonography as early as 11

weeks of gestation and may be detected by means of transabdominal ultrasonography at 16 weeks. If a cleft lip is detected, the fetus has an increased likelihood of having another malformation, a chromosomal aberration, or both. Yet the fetal face is not included in the basic structural survey recommended by the American Institute of Ultrasound in Medicine. Less than 25 percent of cleft lips are identified in ultrasonographic facilities in this country. However, specialists in high-volume prenatal centers can probably diagnose cleft lip in as many as 80 percent of affected fetuses, particularly if they use three-dimensional ultrasonography (see Figure). Cleft of the secondary palate is difficult to visualize; its presence must be inferred from subtle sonographic findings. Prenatal magnetic resonance imaging appears to have better diagnostic accuracy than ultrasonography for cleft of the secondary palate.

Major advances in the care of children born with cleft lip and palate occurred in the last quarter of the 20th century. Rotation-advancement (Millard) repair of unilateral cleft lip has replaced the zigzag scar that was created by earlier methods with a gentle, curvilinear scar that simulates the philtral ridge. Nasal asymmetry can now be corrected at the same time that labial closure is accomplished. It was once accepted that the repair of bilateral cleft lip was twice as difficult as the repair of unilateral cleft lip and that the results were only half as good. However, during the past decade, the old staged operations have been superseded by synchronous repair of the bilateral labial cleft, gums, and nasal deformity. Now children born with such malformations can look as good as their counterparts with a unilateral cleft (see Figure). In major centers, the prevalence of “cleft-palate speech” has diminished from 30 percent to approximately 10 percent.

I believe that every newborn with a cleft lip with or without cleft palate should be sent to a regional cleft-lip-and-palate center. Scandinavia has a long tradition of centralized cleft care, with one center in Denmark, two in Finland, two in Norway, and six



**Figure. Cleft Lip and Palate.**

A three-dimensional ultrasonograph (Panel A) reveals bilateral cleft lip at 22 weeks of gestation. Courtesy of Dr. Beryl Benacerraf. Panel B shows an infant with bilateral complete cleft lip and palate, and Panel C shows the same child at 18 months of age, after synchronous nasolabial repair and palatal closure (performed at a second stage).

in Sweden. A six-center European cleft study demonstrated that standardization, centralization, and the participation of surgeons who perform a high volume of procedures are associated with better results and fewer revisions.<sup>1</sup> It is clear that the quality of the initial nasolabial and palatal repairs is the critical determinant of outcome, in terms of appearance, speech, and facial growth.

In the United Kingdom, the “threshold” volume for primary repair, in order to maintain and improve expertise, has been determined through an outcome assessment of the country’s 57 cleft-lip-and-palate centers.<sup>2</sup> On the basis of this study, the National Health Service has implemented a drastic reduction in the number of centers and has mandated that newborns with a cleft be sent only to teams composed of two surgeons, each of whom treats 40 to 50 such babies annually. Governmental funding has been redirected to only eight teams in England, two in Scotland, and one each in Wales and Northern Ireland.

Such sweeping regulations would be unlikely to be enacted in the United States, with its decentral-

ized health care system in which change occurs slowly and usually on a voluntary basis. The American Cleft Palate–Craniofacial Association (ACPA) has established standards for cleft care and surveyed caseloads in various centers. The mean number of new cases of cleft lip seen per year is only 17, according to the responses from 229 North American centers. Furthermore, 37 percent of the centers (84 teams in the United States and Canada) report performing 10 or fewer primary labial repairs each year.<sup>3</sup> These low numbers are even more disturbing than they appear at first glance, because there is usually more than one plastic surgeon on the team, and the reported number of cases would be divided among them. On the basis of such data, the ACPA is moving toward addressing the level of activity that is necessary for maintaining competency and providing optimal care.

In the meantime, savvy parents are crossing the traditional referral lines and seeking advice in medical cyberspace. On the Web, they find parental support groups that have arisen around the major centers, and they log on to electronic mailing lists. After

the completion of an online crash course in cleft care, parents with an affected child are likely to seek out a surgeon who specializes in this field. Sometimes their choice of a surgeon is unacceptable to their health maintenance organization, which insists that the infant be assigned to a less qualified plastic surgeon within its network. Knowledgeable parents learn to ask the appropriate questions that address the number of cleft-lip repairs performed per year, specific protocols, and the way in which a cleft-lip-and-palate center runs its team. They often ask to see representative photographs showing a surgeon's work. The consulting plastic surgeon may answer the question about annual volume by saying, "I do more than 20 cleft-lip operations every year." The well-informed parent is quick to respond, "Are you referring to infants in your hospital or procedures performed overseas?"

In countries with evolving economies, children with cleft lip and palate are often low on the list of health care (and budgetary) priorities. Although the plastic surgeons in these countries may be quite able, many prefer not to operate on these children because they would receive little or no reimbursement. Into this void have come volunteer surgical teams from wealthy countries, such as the United States, Australia, Germany, and the United Kingdom. There are many such humanitarian organizations, of which the best known in the United States are Interplast, Operation Smile, and the Smile Train. Corporate and individual donors to these enterprises should know that cleft-lip-and-palate missions are the objects of both praise and derision. Many of these missions emphasize head counts — the number of clefts repaired per trip. Often, there is little effort to involve local surgeons, who are left to manage postoperative problems once the visitors have departed. Some physicians in the host countries consider these expeditions to be "surgical safaris." Usually, there is no continuity of care. These children need help with speech, dental and orthodontic services, and often secondary surgical procedures,

and they must be followed until their facial growth is complete.

Many of these humanitarian organizations are currently in transition. It is no longer acceptable to send a team composed of junior residents or practicing plastic surgeons who are not actively involved in cleft care at home. There has been a major shift toward educating the local cleft-care teams, either through direct supervision or by financing visits to cleft-care centers in the United States. This change in philosophy led to the establishment of the Smile Train (headquartered in New York), and now a few other groups are coming on board with the same concept. This organization selects cleft centers in developing countries on the basis of their potential for high-quality care and self-sufficiency; it then directly supports the hospitals and staff, as well as monitoring the surgical outcomes of the designated units.

If a young surgeon is passionate about helping children with cleft lip and palate in other countries, I would advise becoming a specialist who has unique and important skills to offer. A surgical specialist should concentrate on one country and should work within the local infrastructure, returning frequently until the local team is fully capable and outside help is no longer needed. Alternatively, a young surgeon who is interested in this field could stay at home, refining the craft of labiopalatal repair, writing, teaching, and trying to change the U.S. health care system in order to provide the best care for every child born with cleft lip and palate.

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