

Prenatal Diagnosis of Cleft Lip

What the Sonologist Needs to Tell the Surgeon

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High-resolution ultrasonography enables the detection and characterization of many types of fetal facial anomalies. Although the fetal face is still not part of the basic sonographic structural survey recommended by the American Institute of Ultrasound and Medicine, accurate evaluation of craniofacial malformations is usually possible. After the sonologist makes the diagnosis of a fetal cleft lip/palate (and chromosomal aberrations are excluded), consultation with an appropriate surgeon is essential. The parents need to be educated about the severity of the deformity and the predicted outcome of repair. A recent survey of parents showed that only one third thought they were given adequate information about antenatally diagnosed cleft lip by the physician supervising the ultrasonographic examination or by the obstetrician.¹

The purpose of this commentary is to highlight the importance of the report from the sonologist to the surgeon. We underscore the anatomic findings that a surgeon needs to know to present a plan of management to the parents. As in any communication between physicians, there must be common terminology and understanding of the role that each plays. Thus, the surgeon must be aware of the difficulties in ultrasonographic diagnosis of cleft lip and palate, whereas the sonologist should have basic knowledge about the repair of various types of cleft lip and palate.

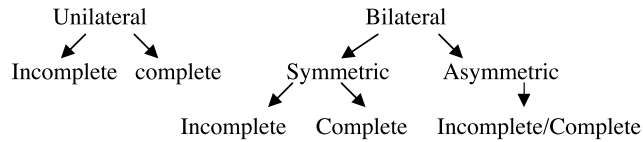
Prenatal ultrasonography permits visualization of the midfacial structures that are formed by 4 to 6 weeks, i.e., the nose, lip, alveolus, and hard palate anterior to the incisive foramen. Embryologists call this the *primary palate*. The *secondary palate* (defined as posterior to the incisive foramen) fuses between 8 and 12 weeks' gestation. Isolated cleft palate is rarely identified antenatally. The sonologist's report to the surgeon who will counsel the parents should include the specific type of cleft lip and details about the extent of the anomaly. Figure 1 is a data form that we have used to compose such a report.

Unilateral Versus Bilateral and Incomplete Versus Complete

The sonologist should comment on whether the cleft is unilateral or bilateral and whether the cleft is complete or incomplete. Unilateral versus bilateral indicates whether one or both sides of the lip are involved. The term *incomplete* denotes that the labial cleft does not extend through the nasal sill or floor (Fig. 2). The term *complete* signifies that there is no tissue connection between the alar base and medial labial ele-

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CLEFT LIP :



CLEFT PALATE; Yes No

Figure 1. Template for report generated by the sonologist to the consulting plastic surgeon. The appropriate cleft type is circled and identified as left or right.

ment or premaxilla (Figs. 3 and 4). In rare instances, there may be a *microform* (tiny) cleft on the “normal” side of a unilateral labial cleft. Such a minor cleft can be difficult to visualize antenatally (Fig. 5).

Some surgeons use the term *Simonart's band* to designate a thin bridge of tissue connecting the lateral and medial sides of the cleft lip. This is an undeserved eponym, a misattribution to a 19th century German surgeon named Simon, plus

arzt (meaning doctor).² Nevertheless, prenatal discovery of such a small band of tissue between the 2 sides of a complete labial or alveolar cleft is very important. Such a band may well determine the number of procedures, whether presurgical dentofacial orthopedics is necessary, timing of repair, outcome, and need for revisions (Fig. 5). Sonologists may not be aware of the surgical significance of this band. Furthermore, the ability of ultrasonography (two- or three-dimensional) to detect the presence or absence of this band is undetermined.

Anatomy of the Cleft Determines Strategy

Dental and surgical protocols for management differ among cleft lip/palate centers. In general, an incomplete cleft lip, either bilateral or unilateral, is repaired in a single stage, usually around 3 to 4 months of age. For a complete cleft lip, either unilateral or bilateral, presurgical premaxillary orthopedics is generally recommended. The pediatric dentist or orthodontist takes an alginate impression of the alveolus and anterior cleft palate from which a plaster cast is made as a

Figure 2. Unilateral incomplete cleft lip. **A.** Coronal two-dimensional sonographic view through the fetal upper lip showing a subtle cleft. **B.** Postnatal photograph of the same infant showing the left unilateral incomplete cleft lip.

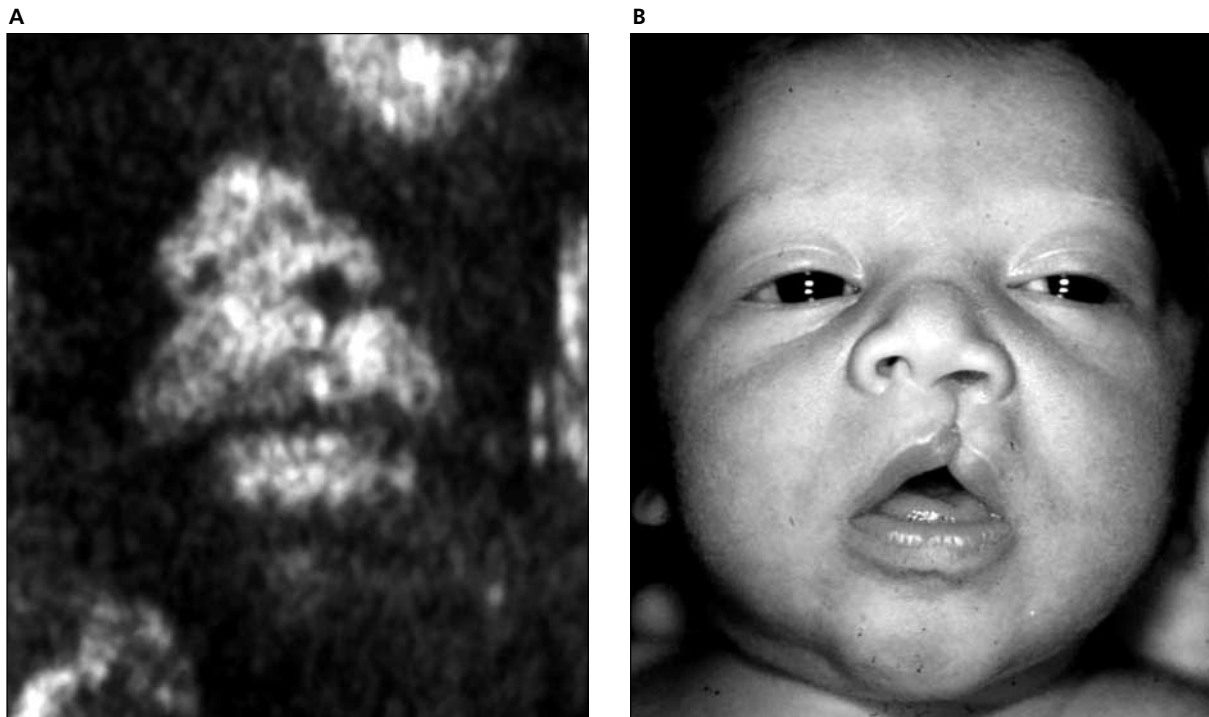




Figure 3. Unilateral complete cleft lip and palate. **A**, Three-dimensional sonogram showing a unilateral cleft lip that extends into the nose, consistent with an associated cleft of the secondary palate. **B**, Postnatal photograph of the same infant.

model for construction of an acrylic palatal appliance. There are 2 basic types of appliances: active (pin-retained) and passive (removable). The palatal appliance is gradually adjusted to bring the cleft maxillary segments into alignment, thus facilitating single-stage labial repair and, whenever possible, closure of the alveolar cleft.^{3,4} After presurgical orthopedics for a unilateral complete cleft lip, many surgeons prefer a single-stage repair. However, some surgeons use a two-stage nasolabial correction, beginning with labial-nasal adhesion.⁴ Successful presurgical orthopedics permits synchronous repair of the bilateral complete cleft lip, correction of the nasal deformity, and closure of the alveolar clefts (Fig. 4).^{3,5}

Premaxillary orthopedic manipulation cannot be used in an infant with a complete cleft of the primary palate (unilateral or bilateral) and an intact secondary palate. For a unilateral cleft of the lip and alveolus, preliminary labial adhesion (a simple 3-layered apposition of the lateral and medial elements) before more definitive nasolabial repair serves to align the maxillary segments.⁶ For the infant with bilateral complete cleft lip and alveolus and an intact secondary palate, usually the maxillary and premaxillary

segments are in an acceptable position to permit synchronous (1-stage) nasolabial repair.

The dental and surgical maneuvers are more complicated in the rare instance of the infant with an asymmetric bilateral cleft lip, i.e., complete on 1 side and incomplete on the other. One strategy is to use premaxillary orthopedic manipulation to narrow the complete cleft, followed by labial adhesion and alveolar gingivoperiosteoplasty. These preliminary maneuvers level the surgical field for synchronous bilateral nasolabial repair.

Parents may request to see “before” and “after” photographs. The surgeon is obligated to show representative results of children born with a similar anatomic cleft from the surgeon’s practice, not from textbooks. Should the parents be encouraged to meet a family with a child who has had a repaired cleft lip? Parent support groups have arisen around most large craniofacial and cleft centers, and often designated parents have been trained as postnatal advisors. However, we agree with Eiserman and Strauss,⁷ who cautioned that it is usually unwise to delegate prenatal counseling to parents or patients. In this regard, Jones⁸ reported

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that most parents in her area declined the opportunity to meet other parents during the prenatal period of decision making.

Alveolar Cleft

If possible, the sonographic report should include a statement about the extent of the alveolar cleft and degree of premaxillary proclination (Fig. 4). Nyberg and colleagues⁹ described protrusion of the intermaxillary segment as a feature of a bilateral complete cleft, which can be readily seen on a sagittal sonogram of the fetal face. Details such as the degree of protrusion are useful to the surgical consultant because they determine whether presurgical orthopedics will be necessary. The parents usually ask about the whether any teeth will be missing or malformed.

Cleft of the Secondary Palate

Sonographic visualization of the secondary palate is difficult and unreliable. Tongue movements above the level of the hard palate suggest that there is likely to be a cleft. Color Doppler ultrasonography, showing reflux of amniotic fluid into the nasal cavity, may be useful in this regard.¹⁰ A cleft of the hard and soft palate is often assumed to be present, particularly with a wide labial cleft, although not actually seen. If indeed there is a complete cleft lip and alveolus, usually the hard and soft palate are also involved; however, in about 10% of these infants, the secondary palate is intact.

If the sonologist were ever certain (highly unlikely) that there is a cleft of the secondary palate, the parents could be informed about the implications for speech and the predictable problems with drainage of the middle ears. A cleft of the hard and soft palate or soft palate only is usually closed in a single procedure before

Figure 4. Bilateral complete cleft lip and palate. **A**, Coronal two-dimensional sonographic view through the fetal upper lip showing a cleft lip on both sides of the premaxilla. The cleft extends into the floor of the nostrils, indicating that it is complete. **B**, Longitudinal view of the same fetal face showing premaxillary proclination secondary to the absence of the alveolar attachment and tongue movement. **C**, Postnatal photograph of the same infant. The child underwent dentofacial orthopedic manipulation. **D**, Photograph of the infant 1 month after synchronous repair of the bilateral cleft lip, nasal deformity, and alveolar clefts.



A



B



C



D



A



B



C



D

1 year of age. In some centers, the soft palate is closed first, and the hard palate is repaired in early childhood. If the secondary palate is intact, the parents can be told that the risk of chronic middle ear infection (otitis media) will be no greater than that in a child born without a cleft palate. Furthermore, the parents can be assured that the child's speech will not be affected.

The Sonologist As Referring Physician

Referral patterns are changing for infants with cleft lip/palate, whether diagnosed prenatally or postnatally. Notwithstanding the advances in ultrasonography, in most infants cleft lip/palate is still diagnosed in the delivery room. Thus, traditionally, the referral to a surgeon or cleft team continues to be the responsibility of the neonatologist or pediatrician. However, with the increasing likelihood of antenatal diagnosis of cleft lip, the sonologist may choose to guide the parents to a plastic surgeon. Where should the parents go and when?

Multidisciplinary or interdisciplinary cleft lip/palate clinics began in the 1930s and continue to be the model of care. There is accumulating evidence that these specialized centers provide improved outcomes, in terms of facial appearance, complications, and need for secondary procedures. The plastic surgeon with an abiding interest in children with cleft lip is the pivotal consultant for the parents both prenatally and postnatally. Parents benefit immensely from a prompt referral to such a specialized pediatric plastic surgeon, who should also be a member of the most experienced regional cleft lip/palate team.

Figure 5. Unilateral complete cleft lip and palate with a cutaneous band. **A** and **B**, Two-dimensional (**A**) and three-dimensional (**B**) sonograms showing a unilateral cleft lip involving the nostril floor, consistent with an associated cleft of the secondary palate. The mucosal band, so important to the repair process, was not looked for and was not seen. **C**, Postnatal photograph of the same infant showing a right unilateral complete cleft lip and palate. Also note the left microform cleft lip. **D**, Postnatal submental photograph of the same infant showing the small cutaneous band bridging the alveolar gap and holding the maxillary segments in relatively good alignment. The parents had been counseled that the infant would need premaxillary orthopedics and a 2-stage nasolabial repair. Because of the band, presurgical manipulation was not necessary.

References

1. Matthews MS, Cohen B, Viglione N, Brown AS. Prenatal counseling for cleft lip and palate. *Plast Reconstr Surg* 1998; 101:1–5.
2. Gibson T. Gustav Simon (1824-1876): Simonart(s)(z) of the band? *Br J Plast Surg* 1977; 30:255–260.
3. Mulliken JB. Bilateral complete cleft lip and nasal deformity: an anthropometric analysis of staged to synchronous repair. *Plast Reconstr Surg* 1995; 96: 9–23.
4. Mulliken JB, Martinez-Peréz D. The principle of rotation advancement for repair of unilateral complete cleft lip and nasal deformity: technical variations and analysis of results. *Plast Reconstr Surg* 1999; 104: 1247–1260.
5. Mulliken JB. Primary repair of bilateral cleft lip and nasal deformity. *Plast Reconstr Surg*. 2001; 108: 181–194.
6. Van der Woude DL, Mulliken JB. Effect of lip adhesion on labial height in two-stage repair of unilateral complete cleft lip. *Plast Reconstr Surg* 1997; 100: 567–572.
7. Eiserman W, Strauss RP. The early diagnosis of cleft lip and the decision-making process [letter]. *Cleft Palate Craniofac J* 1999; 36:542–545.
8. Jones MC. Prenatal diagnosis of cleft lip and palate: experiences in Southern California. *Cleft Palate Craniofac J* 1999; 36:107–109.
9. Nyberg DA, Hegge FN, Kramer D, Mahony BS, Kropp RJ. Premaxillary protrusion: a sonographic clue to bilateral cleft lip and palate. *J Ultrasound Med* 1993; 12:331–335.
10. Monni G, Ibba RM, Olla G, Cao A, Crisponi G. Color Doppler ultrasound and prenatal diagnosis of cleft palate. *J Clin Ultrasound* 1995; 23:189–191.