Cleft Lip and Palate

David J. Crockett, MD, Steven L. Goudy, MD*

KEYWORDS

• Cleft lip • Cleft palate • Orofacial clefting • Cleft lip repair • Cleft palate repair • Palatoplasty • Multidisciplinary care

KEY POINTS

- Cleft lip with or without cleft palate is the most common congenital malformation of the head and neck.
- Each patient should be evaluated for congenital anomalies, developmental delay, neurologic disorders, and psychosocial concerns before surgery.
- A multidisciplinary team is necessary to ensure that every aspect of the child's care is treated.
- The surgeon should be aware of the needs of the cleft patient and be able to educate and assist caretakers as necessary.
- A fundamental understanding by the surgeon of the surgical options for cleft repair is warranted.

OVERVIEW

Cleft lip with or without cleft palate is the most common congenital malformation of the head and neck. The impact on quality of life for the child and the family can be severe, particularly in unsuspecting families. Emotional and psychological needs must be recognized and addressed, in addition to surgical care, for all those involved with the patient. Assessment and treatment of those with cleft lip and/or palate requires a multidisciplinary approach. Access to and evaluation by speech-language pathology, surgery, psychology, psychiatry, social work, audiology, genetics, dentistry, otolaryngology, and pediatric primary care are all recommended by the American Cleft Palate–Craniofacial Association.[1] The recommendation for a team approach allows the child to be able to obtain complete and coordinated care. This article discusses the assessment and treatment recommendations for children born with cleft lip and/or cleft palate. This article focuses on the surgical management and treatment of these special patients.

Incidence and Genetics

Clefts of the lip and/or palate affect approximately 1 in 700 live births.[2] The incidence varies widely depending on geographic origin, racial and ethnic group, environmental exposures, and socioeconomic status. Asian and Native American populations have reported prevalence rates as high as 1 in 500. European populations are approximately 1 in 1000, whereas African populations have a reported prevalence close to 1 in 2500.[2] Clefts of the lip and/or palate can be categorized as syndromic or nonsyndromic. Syndromic clefts are those that occur in association with a recognized pattern of human malformation or syndrome. The cause of a syndromic cleft may be associated with gene transmission, chromosomal aberrations, teratogens, or environmental factors.[3] Identifying an associated syndrome is important, because it

Disclosures: There are no actual or potential conflicts of interest, including employment, consultancies, stock ownership, patent applications/registrations, grants, or other funding.
Department of Otolaryngology – Head and Neck Surgery, Vanderbilt University Medical Center, Vanderbilt University, 1211 Medical Center Drive, Nashville, TN 37232, USA
* Corresponding author.
E-mail address: steven.goudy@vanderbilt.edu

Facial Plast Surg Clin N Am 22 (2014) 573–586
http://dx.doi.org/10.1016/j.fsc.2014.07.002
1064-7406/14/$ – see front matter

can have prognostic implications that may be helpful to the patient and the family.

Classification

Orofacial clefts include all variations of cleft lip and cleft palate. A variety of classification schemes have been suggested and recommended for typical and atypical orofacial clefts.[4] The features used to initiate the classification of an orofacial cleft include the laterality, completeness, severity (wide vs narrow), and presence of any abnormal tissue. Diminutive orofacial clefts may also be described as microform, occult, or minor.[5] The cleft lip laterality includes unilateral and bilateral. A complete cleft lip extends through the lip and the nasal sill, whereas an incomplete cleft lip extends through the orbicularis oris and skin, but intact lip tissue persists. The cleft alveolus can also be considered complete or only notched. A weblike piece of tissue may extend from the lip's cleft side to the noncleft side at the nasal sill. This abnormal tissue is called a Simonart band and, if present, it is not considered the same as an incomplete cleft.

A cleft palate can be unilateral, when 1 palatal shelf attaches to the nasal septum, or bilateral. The classification scheme introduced by Victor Veau[6] is the most popular system. Clefts of the palate were placed into 4 groups. A group I defect includes a cleft of the soft palate only. Group II clefts exist when the defect involves the soft palate and the hard palate to the incisive foramen (secondary palate). Groups III and IV are unilateral and bilateral defects extending through the entire palate and alveolus, respectively.

PATIENT ASSESSMENT

Multidisciplinary Care

Patients with cleft lip and/or palate may present as early as during the prenatal period. Surgical consultation before birth is becoming more common because of the ability to make the diagnosis on prenatal ultrasonography. Discussion regarding general care issues and surgery can help improve some of the anxiety the expecting mother may be experiencing. After birth, the initial management includes ensuring proper feeding of the neonate and evaluation for other comorbidities. As previously discussed, a multidisciplinary approach should be used in the assessment of the child. The patient should undergo initial evaluations by a pediatrician, geneticist, surgeon, feeding specialist, and social worker. Such services allow immediate education and support for the caretakers and the patient. Future assessments need to be performed by audiology, otolaryngology, dental, maxillofacial, speech-language pathology, and psychosocial practitioners.

Surgical Assessment

Immediately after birth, the surgeon should evaluate and examine the neonate. The various anatomic sites for clefting are assessed, including the upper lip, alveolar arches, nostrils, and primary and secondary palates. These areas should be palpated and inspected under direct visualization. Microform cleft lip and submucous cleft palate can often present with only subtle findings on clinical examination. Particular attention should be given to the nasal characteristics in the setting of a cleft lip. The nasal alar symmetry, tip projection, and alar base position and width should all be assessed. The extent of the clefting can be classified, as described previously. A thorough physical examination is necessary to evaluate for any signs of dysmorphia that may lead to the identification of other congenital anomalies or a syndromic diagnosis.

Regular clinic visits with the patient and caretakers allow the surgeon to provide counsel and guidance before surgery. Feeding and weight gain are important aspects to monitor with each visit. Associated congenital anomalies, developmental delay, and neurologic disorders should also be followed. Any concern for cardiac defects or airway obstruction needs to be identified and evaluated promptly. A multidisciplinary approach is warranted to ensure proper management of all of the needs of the child. The surgeon should be aware of these needs and assist with any referrals to the necessary specialists.

UNILATERAL CLEFT LIP AND NASAL DEFORMITY

Preoperative Planning and Preparation

Before surgical treatment of a unilateral cleft lip, adequate weight should be established, with the child weighing at least 4.5 kg (10 lb). Breast feeding is recommended, when possible, but often this is difficult for the infant. However, pumped breast milk may be taken via bottle feeds with the use of a specialized nipple that controls the flow rate, such as a Haberman or pigeon-type nipple based on an evaluation by speech therapy. Most surgeons prefer an average of 28 g (1 ounce) of weight gain per day beginning 2 after birth.

Adequate management for any cardiopulmonary anomalies should be ensured. If there are any concerns with the ability of the child to tolerate general anesthesia, evaluation by an anesthesiologist before surgery is warranted. The anesthesiologist