Cleft lip and palate are among the most commonly encountered congenital malformations, and they represent a spectrum of abnormality – from the microform cleft lip with minimal apparent disruption or functional impairment, to the complete bilateral cleft lip and palate, to the atypical (Tessier) clefting and to those associated with syndromes. In the interest of brevity, this article will focus on the standard, commonly encountered cleft lip and palate.

Incidence is thought to be approximately 1 in 600 live births, and may be increasing. The etiology remains unknown in most cases, but prenatal exposure to viruses, drugs, cigarette smoking and other commonly suspected factors have been implicated. Additionally, they are common features of some well-defined syndromes (Van der Woude, Stickler and Velocardiofacial, to name a few). Today, diagnosis routinely is made prenatally by 20-week ultrasound, though cleft lips usually are easier to discern than isolated, especially incomplete, cleft palates. At birth, a thorough inspection of the lips, nose and oral cavity will confirm the diagnosis, even in subtle forms of the malformation (such as disrupted vermillion, flattened nostril on one side, notched gum, bifid uvula, and V-notch at posterior hard palatal midline).

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Anatomically, the palate is divided into primary and secondary segments. The division is the incisive foramen, which is a point in the midline immediately posterior to the central incisors. The middle upper lip originates from the same embryonic structure as the primary palate. The bony structure of the primary palate (premaxilla) usually contains only the incisors, and the canines, premolars and molars reside in what is referred to as the lateral segments. These structures, as well as the hard palate and soft palate (to include the musculature) constitute the secondary palate. The clefts almost always follow this line between incisors and canine in the primary palate, then at the midline in the secondary palate, whether complete or incomplete, unilateral or bilateral. The reason to note this anatomic point is that it is more useful to think of the common forms of cleft lip and palate as falling into two categories: those that are confined to the secondary palate, and all others.

The clinical significance of this distinction is apparent when one recognizes that functional ramifications of this condition usually follow similar lines. That is, clefts confined to the secondary palate are more likely to be associated with compromised airway and broader syndromic presentations, whereas those that involve the primary palate, with or without involvement of the secondary palate, tend not to have airway obstruction, but always have greater disruption of the dentition.

**CRITICAL FUNCTIONAL ISSUES**

The initial encounter of the newborn with a cleft should focus on critical functional issues. In order of importance they are airway, feeding and associated anomalies (such as cardiac). Children with Pierre Robin Sequence (retrognathia, glossoptosis, airway obstruction) are a special subset of these children, and are rare. The palatal cleft typically is wide and U-shaped, as opposed to the more commonly encountered V-shape, and there is usually a dramatically dysmorphic facies, with virtually no chin projection in profile. The lower gum is well recessed as well. When these features are present, the child should be admitted to the Neonatal Intensive Care Unit, and plans made for intervention. Options include conservative measures such as positioning and use of a nasopharyngeal airway, as well as surgical interventions, including tongue-lip adhesion, mandibular distraction and even tracheostomy in the most severe cases. The intensity of care is guided by input from neonatology, pulmonary, otolaryngology and plastic surgery, and includes imaging, endoscopy and polysomnography to objectively define the problem and exclude non-mechanical etiologies.

With regard to feeding, a child with a cleft palate is unable to generate negative intraoral pressure, so must be aided. A variety of cleft nursers are available, but the most commonly employed allow for a controlled amount of formula or breast milk to be delivered to the infant by the parent. It takes some training and practice and is generally the focus of the early visits to the cleft surgeon. The Cleft Palate Foundation (www.cleftline.org) has a number of educational materials available to families, including a detailed instructional online video that focuses on feeding babies with clefts.

Most children with clefts do not have associated congenital anomalies, though orofacial clefting has been described as a feature of more than 300 syndromes. When associated anomalies do occur the most common are CNS, cardiac (16 percent of syndromic forms) and clubfoot. No formal consensus has been reached with respect to screening for CNS or cardiac disease beyond standard clinical evaluation.
COMPREHENSIVE LONG-TERM TREATMENT REQUIRED

Beyond the immediate, critical issues discussed above, the long-term, comprehensive treatment plan for each child with a cleft anticipates five distinct functional issues: facial growth, speech, hearing, dental development and psychosocial maturation.

FACIAL GROWTH

Orofacial clefts are not confined to the lip and palate. The internal structure of the nose is usually affected, as is the maxilla. The standard of care is to intervene surgically to correct the lip and nose at about 3 months of age, and the palate at about 12 months. Different techniques are described, but the effectiveness of the surgical procedures may not be readily apparent. Ongoing growth impacts the outcomes of lip and nose repairs, and the act of surgery itself creates some measure of unpredictability with regard to its effect on growth. It is not uncommon to have to correct residual deformity that becomes more apparent over time. Moreover, the projection of the maxilla may ultimately demonstrate growth deficiency and result in severe malocclusion and the potential for temporomandibular joint problems. The timing of these procedures is dependent upon the severity of the deformity and what, if any, negative impact it has on the child’s normal functional and psychosocial development.

SPEECH AND HEARING

The issue of hearing is related to the aberrant insertion of the muscles of the soft palate. As a result, these children usually manifest Eustachian tube dysfunction, requiring ventilation tubes. It is not uncommon for middle ear effusion to be evident after the first few months, and maintenance with tube replacement on a regular basis is required to prevent conductive hearing loss. Hearing is repeatedly checked during team visits throughout childhood, and usually by age 7 or so the effusions seem to resolve, and hearing stabilizes. The need for hearing aids is occasionally seen, however, as is the need for myringoplasty.

The threat of conductive hearing loss is only one of many factors impacting normal speech development in these children. In fact, speech and hearing development is the primary focus of team care from age 2 to about 7 when they are seen for routine follow-up. Impaired hearing, velopharyngeal dysfunction related to the cleft palate and its repair, disrupted dental development and oronasal fistula formation all contribute to speech “hazards.” Some children develop maladaptive compensatory articulation and many require relatively intense speech therapy throughout childhood to correct this. Others require surgical intervention in the form of a pharyngeal flap, for example, to address anatomic deficiency that cannot be overcome with conservative measures.

DENTAL DEVELOPMENT

Children with cleft palate have neonatal teeth, missing teeth and supernumerary teeth. Both primary and permanent dentition are crowded and can erupt in the palate and the cleft itself. They often have enamel hypoplasia as well, and these factors contribute to a higher incidence of caries and decay. Routine dental care must be established at an early age. A critical issue occurs when the cleft involves the alveolar ridge. In these cases, orthodontic preparation must be initiated at eruption of the first permanent molars (about age 6) in order to align the alveolar ridges for bone grafting before eruption of the permanent canine. Failure to do so may result in ankylosis of the tooth in the maxilla, with no way to restore it. Complex orthodontic treatment is always a key part of the coordinated care of these children.
PSYCHOSOCIAL MATURATION

Psychosocial development merits its own classification because it addresses the whole child. All of the surgical, orthodontic and speech therapeutic intervention is meant to restore what we regard as “normal.” What we really strive to do is to prevent the children from feeling different. A truly successful treatment plan is more than a series of office visits and surgical procedures.

NEW DEVELOPMENTS

With respect to new developments in cleft care, two stand out in particular. The first, nasoalveolar molding, is a form of presurgical orthopedics whereby the alveolar ridges are aligned by passive means using constant traction on an intraoral appliance coupled with active stretching of the deficient nasal alar dome. The result is improved shape and form, ultimately translating to superior functional outcomes with respect to occlusion and nasal airway. The second is the technological development of intraoral distraction devices, allowing much improved comfort and earlier intervention in the management of midface growth deficiency and malocclusion. The result here is a diminished burden on children with severe forms of these problems, and the ability to intervene prior to skeletal maturity without any apparent effect on growth potential.

Generally speaking, the long-term prognosis for children with orofacial clefting is excellent. While it is a long journey, often with several surgical procedures, it is a field with a long and rich history of coordinated, interdisciplinary care. It was early recognized that this population of patients represented a unique clinical problem that by its nature required early intervention to prevent permanent disability, and that also required ongoing care to address the inevitable evolution of the problem as the children grew and changed. The creation of cleft teams more than 50 years ago for the expressed purpose of coordinated care by several disciplines established the standard of team care. This essential concept remains as relevant today and is the model for other team-care initiatives that have since been developed. Plastic surgery, otolaryngology, speech pathology, audiology, genetics, dentistry, orthodontics and psychology represent the core disciplines. The participation of each is now a standard of care as this approach has demonstrably improved outcomes for these children.

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