

Achieving Excellence in Cleft Care:

From Birth to Adulthood

Joseph A. Napoli, MD, DDS¹ and Linda D. Vallino, PhD, MS, CCC-SLP/A, FASHA²

1. Division Chief, Plastic and Maxillofacial Surgery; Director, Cleft Lip and Palate and Craniofacial Anomalies Program, Nemours Childrens' Clinic, Alfred I. duPont Hospital for Children

2. Head, Craniofacial Outcomes Research Laboratory, Center for Pediatrics Auditory and Speech Sciences; Senior Research Scientist, Nemours/Alfred I. DuPont Hospital for Children

Orofacial clefts, comprised largely of cleft lip with or without cleft palate (CL/P) and cleft palate only (CPO) are among the most common congenital anomalies (Figure 1). Because of the anatomic location of the birth defect, a wide range of structural and functional impairments occur that are best managed by a variety of healthcare specialists. Although most associated problems are a result of the defect, some are by iatrogenic means. Moreover, while most issues are managed during childhood, adults with CL/P will often have cleft related treatment needs. The purpose of this article is to describe the issues associated with orofacial clefts, management of these issues, and discuss strategies to achieve excellence in care for our patients and families affected by these conditions throughout their lives.

Figure 1. Complete bilateral cleft lip and palate. Note teeth in premaxilla. Image from Joseph A. Napoli M.D., D.D.S., Plastic, Maxillofacial, and Craniofacial Surgery, Alfred I. DuPont Hospital for Children. Zajac and Vallino. (2017). Evaluation and Management of Cleft Lip and Palate: A Developmental Perspective, Plural Publishing, Inc. Reprinted with permission.



OVERVIEW OF CLEFT LIP AND PALATE AND ASSOCIATED PROBLEMS

The incidence of orofacial cleft nationally is approximately 1 in 600 births. In Delaware, the estimated incidence for cleft lip, with or without cleft palate is approximately 1 in 1270 births and 1 in 1600 for cleft palate only.¹

Among the earliest concerns for a child born with a cleft palate is airway obstruction, specifically in those children with CPO associated with Pierre Robin sequence (PRS). The airway impairment occurs at the level of the tongue base resulting from mandibular hypoplasia. Feeding difficulty may also occur either simply related to the cleft palate which prevents generation of the intraoral negative pressure necessary for suction, or airway compromise (i.e. Pierre Robin sequence), which can limit the rate of nutrition intake, causing failure to thrive.

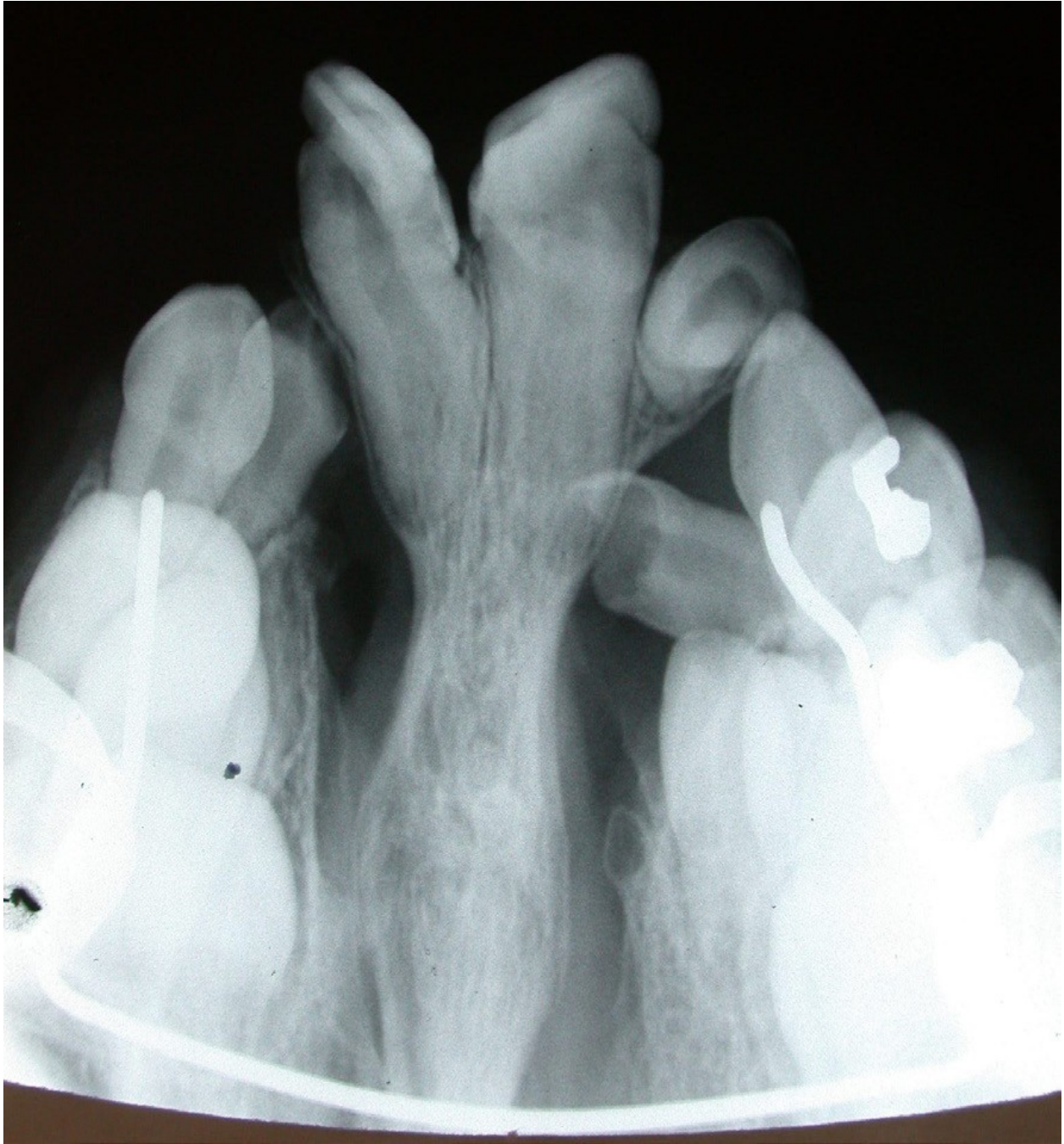
A cleft of the palate very often leads to eustachian tube dysfunction due to the anatomic arrangement of the tensor veli palatini and levator palatini muscles. Otitis media with effusion is almost universal among children born with cleft palate and, if left untreated may lead to permanent conductive hearing loss. It should be noted that some children with cleft palate will pass the newborn hearing screening exam, only later to develop fluid build-up in the middle ear, resulting in hearing loss identified on later examination. Hearing is critical to the development of normal speech. Hearing loss at an early age can impair speech and language development and this could be particularly significant in children with cleft palate who are already at risk for speech difficulty.

The soft palate is part of a complex valve mechanism necessary for normal speech production. When incomplete closure of the velopharyngeal port occurs (velopharyngeal dysfunction), speech will sound hypernasal and can limit intelligibility. In some children with velopharyngeal dysfunction (VPD), their attempts to compensate for the “air leak” during speech production can lead to other complex speech sound errors (known as “compensatory errors”) which may be very difficult to correct. In addition to the speech errors related to VPD, speech errors associated with

dental malocclusion may also occur. These latter types of errors are not amenable to speech therapy and require orthodontic and/or surgical procedures to allow correction.

In patients with cleft lip and palate, particularly if the anterior maxilla and the alveolar ridge are affected by the cleft, the alignment and position of the teeth are likely to be adversely affected. Dental anomalies are also prevalent in patients with cleft lip only.² This will often make dental hygiene difficult and lead to an increase in dental caries rate. Additionally, the bone defect resulting from the cleft creates inadequate support for teeth along the cleft which may lead to tooth loss (Figure 2). Furthermore, surgery to repair the cleft lip and palate may interfere with growth of the maxillary segments leading to severe malocclusion of the jaws. Occlusal defects can have an adverse effect on eating, speech and facial appearance.

Figure 2. The x-ray shows the boney defects in bilateral cleft lip and palate. Image from Joseph A. Napoli M.D., D.D.S., Plastic, Maxillofacial, and Craniofacial Surgery, Alfred I. DuPont Hospital for Children. In: Zajac and Vallino. (2017). Evaluation and Management of Cleft Lip and Palate: A Developmental Perspective, Plural Publishing, Inc. Reprinted with permission.



Oronasal fistulas associated with cleft of the maxilla may result in regurgitation of food and fluid leaking out of the nose, and possibly causing embarrassment (Figure 3).

Figure 3. Occlusal view of an alveolar cleft. Image from Joseph A. Napoli M.D., D.D.S., Plastic, Maxillofacial, and Craniofacial Surgery, Alfred I. DuPont Hospital for Children. In: Zajac and Vallino. (2017). Evaluation and Management of Cleft Lip and Palate: A Developmental Perspective, Plural Publishing, Inc. Reprinted with permission.



As already mentioned, poor dental alignment increases the risk of caries. Other risk factors for caries include extensive need for orthodontic care, presence of scar in the labial vestibule which may reduce access for tooth brushing, and other associated medical issues which may reduce dental care as a priority for parents whose children have multiple congenital anomalies.

Finally, but not least in importance, are the psychosocial burdens associated with cleft lip and palate. From the aforementioned issues, one can easily see the financial, emotional, and social burden carried by families afflicted with this condition. Teasing and bullying have long been a concern for families of individuals with facial difference or speech abnormalities and may lead to poor self-esteem. Much has been written about effect of facial differences on the psychological welfare of children and adults alike. Although much of this information appears in health care literature, the topic has also been addressed in popular media, for example, Wonder.³

The cost of cleft care is significant, and certain aspects of care may not be covered by insurance. While most of the surgical procedures are covered, associated dental (especially orthodontic care and implant replacement of missing teeth) and psychologic services may not be covered.

TREATMENT

Obstructive airway issues in neonates and infants with cleft palate are most commonly associated with Pierre Robin Sequence. In this condition, the mandible is small and retrognathic. This can result in glossoptosis with the base of tongue positioned posteriorly, obstructing the airway. Treatment options range from non-surgical maneuvers such as positioning the child in the side lying or prone position in mild cases, to tracheostomy in the most severe cases. Before committing to tracheostomy, however, other options may be considered including tongue to lip adhesion (glossopexy) or mandibular lengthening by distraction osteogenesis (MLDO). A hypoplastic mandible is sometimes identified prenatally and may alert the obstetrician and neonatologist of a potential newborn with an at-risk airway. In the absence of airway obstruction, mandibular hypoplasia is not treated during infancy.

Feeding difficulty attributed to cleft palate is most commonly associated with inability to create negative intraoral pressure which is necessary for suction. Another cause of feeding difficulty is airway obstruction, (typically in patients with Pierre Robin sequence). While breastfeeding may

be possible for infants with a small palatal cleft or isolated cleft of the lip, the majority of infants with cleft palate are unable to breastfeed.

Feeding challenges are addressed by establishing a safe airway, use of special bottles and nipples designed for infants with cleft palate, and supportive coaching for the select infants that may be able to breastfeed. Successful management relies on the expertise of multiple specialists working together including surgeons with expertise in airway management, nurses, occupational therapists, and speech-language pathologists with experience in feeding therapy, and lactation consultants.

Management of the effects of eustachian tube dysfunction is critically important for otologic health, in particular as it relates to infection and hearing loss. Otitis media in children with cleft palate frequently requires otologic surgery, specifically bilateral myringotomy with pressure equalizing tube placement. If myringotomy with tubes is required, this can be combined under the same general anesthetic as another procedure, commonly at time of cleft palate repair. Vigilance must be maintained from infancy, through childhood, and into adulthood to assure the ongoing otologic health and maintenance of optimal hearing as this is so critical not only to speech and language development, but also for the ability to succeed in the classroom and during one's employment.

Cleft palate repair is usually done at approximately one year of age. The speech-language pathologist (SLP) begins to address speech sound development at about this time and continues to follow the patient's communication progress during the course of his or her development. The majority of children who have repair of cleft palate will have normal speech, especially when done by an experienced surgeon. However, due to various factors, complete velopharyngeal closure may not be achieved even after cleft palate repair, resulting in hypernasal speech and nasal air emission. The SLP with experience in cleft associated speech problems provides guidance to patients-and surgeons-during management of these issues. With proper diagnostic tests, including videonasoscopy, the site and anatomic configuration of velopharyngeal portal and inadequate closure can be identified, information that is useful in determining the need for surgical correction of VPD. When surgery is not indicated or cannot correct the problem, prosthetic management such as a palatal lift or palatal bulb appliance, can be fabricated by a maxillofacial prosthodontist to treat VPD (Figure 4).

Figure 4. Speech bulb appliance combined with a partial denture to replace missing anterior teeth. Image from Dennis Ruscello, Ph.D., West Virginia University. In: Zajac and Vallino. (2017). *Evaluation and Management of Cleft Lip and Palate: A Developmental Perspective*, Plural Publishing, Inc. Reprinted with permission.



Fortunately, a number of secondary operations have been designed over the years to correct VPD. Caries is largely a preventable disease. Oral hygiene instruction should be initiated well before the full complement of primary teeth erupt. We recommend hygiene instruction for parents at time of eruption of primary incisors allowing infants to become used to examination of the mouth and teeth.

Support for the permanent teeth in the vicinity of the cleft requires healthy bone and stability of the maxillary segments affected by the cleft. This is accomplished by placing a bone graft, usually autogenous bone harvested from the patient's iliac crest, across the cleft gap in the alveolar process to bridge the bone defect and unite the maxillary segments. This procedure is known as alveolar cleft bone grafting and is usually done when patients are in mixed dentition prior to eruption of maxillary permanent canine, sometime between age 7 and 10 years.⁴ There is usually a crossbite on the side of the cleft; bilateral crossbite in patient with bilateral cleft lip and palate. Prior to placing the bone graft, transverse maxillary expansion is accomplished with an orthodontic palatal appliance. This will correct the maxillary width and allow reconstruction of the alveolar bone defect with bone graft. Benefits of this procedure include closure of the oronasal fistula, provision of bone for support of teeth in region of cleft, and skeletal support for later nasal reconstruction.

As children with cleft lip and palate enter the teen years and young adulthood, particularly those with bilateral cleft lip and palate, the malalignment of the jaws worsens, creating a class III malocclusion. Both the crossbite and class III malocclusion are iatrogenic and associated with surgical procedures done to repair cleft lip and palate during infancy and childhood. If severe enough, orthognathic surgery is required to correct the malocclusion and midface deficiency.⁵ If

the anterior-posterior discrepancy is very severe, conventional orthognathic surgery may be insufficient to correct the occlusion and advancing the maxilla by distraction osteogenesis may be necessary (Figure 5a – Figure 5d). There is significant risk in attempting to correct the skeletal malocclusion with orthodontics alone including resorption of tooth roots, especially the maxillary incisors, with tooth loosening, and in severe cases, loss of teeth. Images from Joseph A. Napoli M.D., D.D.S., Plastic, Maxillofacial, and Craniofacial Surgery, Alfred I. DuPont Hospital for Children. In: Zajac and Vallino. (2017). Evaluation and Management of Cleft Lip and Palate: A Developmental Perspective, Plural Publishing, Inc. Reprinted with permission.

Figure 5a. Preoperative occlusal view of a young adult with cleft lip and palate.



Figure 5b. Preoperative facial view of a young adult with cleft lip and palate.

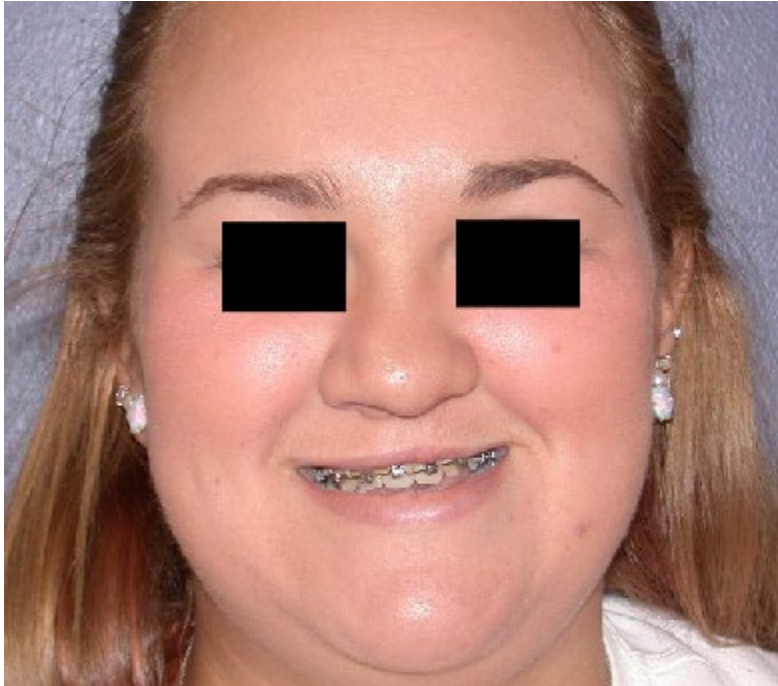


Figure 5c. Postoperative frontal view of same patient after maxillary advancement using distraction osteogenesis.



Figure 5d. Postoperative profile view of same patient after maxillary advancement using distraction osteogenesis.



Perhaps the most significant issue associated with facial differences associated with cleft lip and palate is the psychological burden carried by both the affected individuals and their family members. Additionally, the costs of services to care for these individuals can significantly add to the stress experienced by affected families and may even become a barrier to adequate care. While the stigmata of the cleft cannot always be removed, and the financial obligations completely mitigated, much can and should be done to reduce the burden and assist individuals and their families in their ability to manage these issues. Psychologists and other mental health professionals, and social workers are vital members of the cleft and craniofacial team and actively play a role in caring for these patients. Access to psychological care for children and families is considered standard of care by,⁶ and is often initiated during team visit.

PRINCIPLES OF HEALTHCARE EXCELLENCE

The Oxford dictionary defines excellence as “the quality of being outstanding or extremely good.”⁷ Achieving excellence in healthcare for patients with CLP compels us to provide a comprehensive approach to care that will deliver to a patient the best possible outcome and satisfaction with care, leading to a positive impact on the person’s quality of life. To do this effectively, we need to create a culture of excellence that at least includes a standard set of guidelines such as that set forth by the ACPA Standards of Care.⁶ The culture should be that of patient-centered care, innovation without compromising quality outcomes, and continual improvement.

Creating excellence in healthcare is an ongoing process. There are at least five core features central to a model of healthcare excellence: it is patient-centered, accessible, collaborative, includes measurable outcomes, and cost- effective. Each feature will be described below.

Patient-Centered Care

A hallmark of excellence is that treatment is patient-centered. Each patient and caregiver(s) brings to the team or provider a set of values, preferences, and desired healthcare outcomes based on their experiences and perceived needs. Patient-centeredness is a unique blend and appropriate balance of a clinician's findings and input, active participation of parent/patient, and the services that can be provided within a given context.⁸

Certainly, for some decisions, there is clearly one optimal path to take (i.e., cleft palate repair prior to onset of speech), but for most decisions, there are other options (i.e., surgical or prosthetic management of hypernasal speech), including no intervention (as in the case where a teenager does not want surgery to improve speech). This patient/family involvement in the decision adds value to the management plan.

Accessible

Beginning in infancy, most individuals with CLP receive the services of a team starting in early infancy (some prenatally). A connected referral network should be barrier free.⁹

This means that the patient and family should be able to locate a team where services are provided, or community-based providers (i.e., dentist, general care providers, psychologists) who can manage the health care needs of the person with a CL/P. The latter is particularly vital as the young adult with cleft palate transitions from a pediatric interdisciplinary team to an adult-centered team.

In addition to accessibility of care, care needs to be affordable. Recognizing the reimbursement limitations and lack of or inadequate health insurance for some individuals, the move towards excellence needs to actively address the financial issues for individuals with cleft lip and palate.

Collaborative

Collaboration begins early in the child's care to establish care that will carry over into adulthood. Collaboration, especially with providers outside the team is critically important, especially for young adults who are beginning to transition between settings whether it is an adult-centered interdisciplinary team or a community-based system with independent providers.⁸ This interprofessional collaboration provides and assures seamless care for the patient and families.

Measuring Outcomes

Evaluating outcomes and satisfaction is a critical component to health care excellence. It is too important to overlook. Measuring outcomes is difficult and sometimes seemingly impractical but it is a very necessary feature of healthcare excellence. We need to begin by considering measures that are multidimensional, those that are both biomedical and psychosocial (e.g., patient-report of satisfaction with outcome). Robust measures and the means by which it is organized will produce evidence of positive (or negative) outcomes, which may lead to changes in care delivery. Evaluating outcomes is a powerful motivator for continuous improvement.

Cost-Effective

Lastly, healthcare must be cost effective without compromising care. Needless or unnecessary treatment can lead to high cost of care. The use of electronic medical records in both hospital and physician offices between these providers is increasing.¹⁰ Its use and availability can enhance

communication and collaboration between the hospital-based team providers, thereby reducing redundancy in care,¹¹ and costs.

These elements should be nurtured by the leadership of teams involved in cleft care and the organizations they serve to sustain a culture of excellence and continual improvement.

ACHIEVING EXCELLENCE IN CLEFT LIP AND PALATE CARE

From the aforementioned sections, one can begin to see the complex interrelationship of all the issues associated with an orofacial cleft. The impact of the cleft and the perceived impact on the quality of life evolves over the course of the individual's life.^{8,12}

No individual specialist or caregiver is equipped to provide all the necessary care associated with cleft lip and palate and the term "team" has been introduced earlier in the article. Fortunately, K. Herbert Cooper, an orthodontist in Lancaster, Pennsylvania recognized the benefit of having clinicians caring for children with cleft lip and palate see the children together for evaluating and planning care. In 1938, the concept of team care was launched. Dr. Cooper established the Lancaster Cleft Palate Clinic with the initial team consisting of a surgeon, an orthodontist, and a speech- language pathologist. Since then, team care has become recognized as standard practice in managing the health care needs of these individuals with orofacial clefts in this country and around the world. Initially, the team dealt with the obvious issues: visible facial deformity, dental problems, and speech abnormalities but over time many more needs were recognized and addressed.

As greater understanding of the complexity and wide- ranging effect of orofacial clefts developed, other specialties were added to the clinical team including otolaryngology, genetics, dysmorphology, audiology, psychology, and social work. This has been driven by a greater understanding and awareness of the importance of psychologic health, relationship of otitis media to hearing loss and speech development, advancements in medical genetics and teratogenicity, association with other congenital anomalies, especially cardiac and renal, and finally the complex financial considerations as barriers to care. Given the wide range of services required, a team patient care coordinator is necessary to assure that all these needs are met.

Excellence in care begins when an expectant family first learns of a pregnancy with a fetus having a cleft, or any other congenital anomaly for that matter. There is often an associated level of anxiety and sometimes despair surrounding what was hoped to be a very joyous beginning. Prior to high quality prenatal ultrasound, orofacial clefts were not identified until the actual birth, or in the case of CPO, perhaps days to weeks after birth, often rendering care givers ill prepared to deal with both the emotional as well as the physical needs of the infant. Since the advent of high quality prenatal ultrasound, most cleft lips are identified at about 20 weeks of gestational age. This could be considered a mixed blessing, with increased months of anxiety during the remainder of the pregnancy. But it also provides opportunity for education and counseling, very often allaying fears and possibly returning the expected birth into more joyous and celebratory event. Prenatal identification allows care givers to prepare and anticipate the need for higher level airway management and/or availability of special feeding items. It also alerts for the need to evaluate for other congenital anomalies. A prenatal visit with family should be arranged with the hope of providing increased understanding of the issues and support for the family.

Excellence continues at the birth institution, for example, where staff identify the child at risk for airway obstruction and initiate measures to have the delivery occur in a setting where the airway

can be secured if necessary. After birth, excellence in care requires coordinated treatment with correct timing. This is influenced by team cohesion and how well the members work together. It also requires that the team understands that its members cannot always provide all the care themselves, but must very often collaborate with clinicians outside of the team's home institution, especially if the team covers a wide geographic area and insurance programs dictate where patients can receive services. In a way, the "team" would be expanded to include school based speech-language pathologists, community based dentists, including orthodontists, or others who provide care to the patients of a given team.

Excellence requires an understanding that care begins as early as possible and continues into adulthood as needed. It includes an understanding as to how this condition not only affects the individual but other family members who may be a source of support for the patient.

TRANSITION OF CARE

Some teams are limited by age regulations of the home institution, a particular issue in some free-standing children's hospitals. Team care through a general hospital with an associated pediatric unit is less likely to be affected. Much of the care for cleft lip and palate is provided prior to early adulthood but for some patients, unresolved needs remain into adulthood which can and should be addressed. These may include scar revision, correction of nasal deformity, treatment of malocclusion, replacement of missing teeth, ongoing dental care, counseling for job and relationships. As highlighted earlier, collaboration among institutions and caregivers between pediatric facilities and facilities managing adults, can ensure satisfactory, and hopefully seamless transition for affected individuals and continue to provide excellent care in people born with cleft lip and palate.

SUMMARY

Over the past half century, much has been done to improve the lives of those afflicted with orofacial clefts. Maintaining these gains by continuing team care and adopting accepted pathways toward healthcare excellence should be the goal of all involved with caring for this very common condition.

References

1. Mai, C. T., Isenburg, J., Langlois, P. H., Alverson, C. J., Gilboa, S. M., Rickard, R., . . . Kirby, R. S., & the National Birth Defects Prevention Network. (2015, November). Population-based birth defects data in the United States, 2008 to 2012: Presentation of state-specific data and descriptive brief on variability of prevalence. *Birth Defects Research. Part A, Clinical and Molecular Teratology*, 103(11), 972–993. [PubMed](#)
2. Vallino, L. D., Zuker, R., & Napoli, J. A. (2008, September). A study of speech, language, hearing, and dentition in children with cleft lip only. *Cleft Palate Craniofac J*, 45(5), 485–494. [PubMed https://doi.org/10.1597/06-207.1](https://doi.org/10.1597/06-207.1)
3. Palacio, R. J. (2012). Wonder. Alfred A. Knopf.
4. Napoli, J. A. (2017). Alveolar cleft repair. In D.J. Zajac and L.D. Vallino (Eds.). Evaluation and management of cleft lip and palate: A developmental perspective. pp. 339-354. San Diego, CA: Plural Publishing, Inc.

5. Napoli, J. A., & Vallino, L. D. (2017). Maxillary advancement. In D.J. Zajac and L.D. Vallino (Eds.). *Evaluation and management of cleft lip and palate: A developmental perspective*. pp. 357-378. San Diego, CA: Plural Publishing, Inc.
6. American Cleft Palate-Craniofacial Association. (2009). *Parameters for the Evaluation and Treatment of Patients with Cleft Lip/Palate or Other Craniofacial Anomalies*. http://www.acpa-cpf.org/uploads/site/Parameters_Rev_2009.pdf
7. Oxford Dictionary. (2017). Excellence. Retrieved from <https://en.oxforddictionaries.com/definition/excellence>. Accessed December 8, 2017.
8. Vallino, L. D., & Louw, B. (2017). We've got some growing up to do: An evidence-based service delivery model for the transition of care for the young adult with cleft lip and palate. *Perspectives of the ASHA Special Interest Groups*, 2(5), 4–17. <https://doi.org/10.1044/persp2.SIG5.4>
9. Vallino, L. D., Ruscello, D. R., & Zajac, D. J. (2018). Referring to a cleft palate-craniofacial team. In: *Cleft Palate Speech and Resonance: An Audio and Video Resource*. Plural Publishing.
10. Rumball-Smith, J., Shekelle, P. G., & Bates, D. W. (2017, January 17). Using the electronic health record to understand and minimize overuse. *JAMA*, 317(3), 257–258. [PubMed](https://doi.org/10.1001/jama.2016.18609) <https://doi.org/10.1001/jama.2016.18609>
11. Grady, D., & Redberg, R. F. (2010, May 10). Less is more: How less health care can result in better health. *Archives of Internal Medicine*, 170(9), 749–750. [PubMed](https://doi.org/10.1001/archinternmed.2010.90) <https://doi.org/10.1001/archinternmed.2010.90>
12. Zajac, D. J., & Vallino, L. D. (2016). *Evaluation and management of cleft lip and palate: A developmental perspective*. Plural Publishing.

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