

Feeding Issues and Interventions in Infants and Children with Clefts and Craniofacial Syndromes

Claire K. Miller, Ph.D., CCC-SLP¹

ABSTRACT

Problems with oral feeding occur in varying degrees in infants born with cleft lip/palate and/or craniofacial syndromes. The extent of clefting is associated with the severity of feeding problems, and if cleft lip/palate occurs in conjunction with a craniofacial syndrome, additional structural, airway, and neuromotor issues may be present. The infant's feeding and swallowing skills may be significantly impaired, characterized by inefficient oral feeding skills coupled with poor airway protection ability during swallowing. Inadequate airway protection during swallowing has serious implications for the infant's respiratory health as sequelae of chronic aspiration during feeding may include recurrent respiratory illness, pneumonia, and lung damage. Feeding difficulty in nonsyndromic and syndromic cleft lip/palate infants has been documented as source of considerable stress for parents and can have a potential negative effect on the parent-infant bonding process. Therefore, timely identification of feeding problems by the speech pathologist with subsequent intervention and modification in the feeding method is essential, along with provision of early feeding instruction to families. The objective of this article is to review expert opinion and available evidence regarding factors that influence feeding success and efficiency in infants with nonsyndromic and syndromic cleft lip/palate. The types of compensatory strategies or interventions that are effective in alleviation of feeding and swallowing difficulties will be described. Descriptive reports, expert opinion, and available evidence from clinical trials to support the use of feeding interventions in treatment are reviewed.

¹ Speech Pathologist III and Program Director, Aerodigestive and Sleep Center, Cincinnati Children's Hospital Medical Center, Cincinnati, Ohio.

Address for correspondence and reprint requests: Claire K. Miller, Ph.D., Aerodigestive and Sleep Center, Cincinnati Children's Hospital Medical Center, MLC

4011, 3333 Burnet Avenue, Cincinnati, OH 45229-3039 (e-mail: Claire.miller@cchmc.org).

Communication Disorders Related to Cleft Palate, Craniofacial Anomalies, and Velopharyngeal Dysfunction; Guest Editor, Ann W. Kummer, Ph.D., CCC-SLP, ASHA-F.

Semin Speech Lang 2011;32:115–126.
 Copyright # 2011 by Thieme Medical
 Publishers, Inc., 333 Seventh Avenue, New
 York, NY 10001, USA. Tel: +1(212) 5844662.
 DOI: <http://dx.doi.org/10.1055/s-0031-1277714>.
 ISSN 0734-0478.

115

KEYWORDS: Pediatric, craniofacial, feeding, swallowing, dysphagia

serious consequences to respiratory health.

It is generally reported that CL/CP, in the absence of other congenital structural, respiratory, or neurological issues, has only a minimal to moderate effect on feeding success initially.^{1,2} The degree of feeding difficulty will vary

Learning Outcomes: As a result of this activity, the reader will be able to (1) describe craniofacial issues that result in feeding and swallowing dysfunction, (2) discuss various assessment procedures, and (3) apply appropriate intervention techniques for management of feeding and swallowing difficulties.

Cleft lip/cleft palate (CL/CP) and craniofacial syndromes are often associated with special challenges in regard to the feeding and swallowing process in infancy, as well as beyond in certain circumstances. Problems vary in degree depending upon the degree of clefting and if a craniofacial syndrome is present. Severe feeding and swallowing issues have the potential to result in nutritional and/or respiratory compromise, as well as to create significant stress for families and caretakers. Most reports of feeding/swallowing problems and the efficacy of feeding interventions in nonsyndromic and syndromic CL/CP are from descriptive independent studies, though there is increasing availability of empirical evidence to guide interventions in practice.

The infant feeding process depends upon smooth synchronization of sucking, swallowing, and respiration, coordinated by the neurological system. Rapid, sequential swallowing occurs during the apneic phase of the inspiratory-expiratory respiratory pattern to ensure protection of the airway. Precise coordination of all components is critical; disruption in the process can occur if anatomic, neurological, or respiratory issues are present. Inefficient oral feeding may result in inadequate volume of intake for adequate growth, and ongoing problems with airway protection during swallowing present potentially

with the type and severity of the cleft (unilateral, bilateral, incomplete, complete) and may require some modification of feeding technique.^{3,4} For example, an isolated cleft lip generally does not interfere with an infant's ability to efficiently suck during bottle-feeding, whereas an opening in the palate has a more profound effect on sucking efficiency in terms of ability to generate sufficient sucking pressure. This can result in inadequate volume of oral intake if modifications in feeding method are not made. The degree of palatal clefting is reported to correspond to the degree of feeding difficulty, with more significant feeding problems reflective of more extensive palatal clefting.⁵ Usually, an initial assessment by a speech-language pathologist, with subsequent implementation of feeding interventions, such as specialized cleft palate bottles and compensatory feeding strategies and techniques, effectively alleviates the initial feeding difficulty.^{4,6}

In contrast, CL/CP, in conjunction with syndromes, sequences, and associations, presents a more complicated scenario. Oral motor dysfunction and pharyngeal swallowing dysfunction may occur secondary to the structural issues, but also because of cranial nerve abnormalities or neuromotor components that may be present. The infant may have problems with the oral phase of swallowing, coupled with an impaired ability to achieve and maintain airway protection during the pharyngeal phase of swallowing.^{1,6,7} Therefore, the infant with syndromic CL/CP and associated neurological issues will likely have feeding and swallowing issues that will require ongoing input from the pediatric speech pathologist in regard to treatment and management.

CHARACTERISTICS OF FEEDING PROBLEMS DUE TO CL/CP AND CRANIOFACIAL CONDITIONS

Nonsyndromic Clefts

Descriptive reports of infants with CL/CP without accompanying medical issues document varying degrees of feeding difficulty. Usually the feeding difficulties are attributed to the specific structural problem in the oral cavity and are considered oral phase in nature.⁶ The pharyngeal phase of the swallow, where swallowing is initiated and coordinated with airway protection, is intact.

Infants with unilateral or bilateral clefts of the lip may have some initial difficulty in learning how to latch onto the nipple; however, they usually quickly adapt once the nipple is placed intraorally with the assistance of the feeder. The infant will initiate sucking, using reflexive tongue and jaw movements that are sufficient for efficient compression of the nipple against the intact part of the alveolus and palate. There may be some transient difficulty in achieving negative pressure suction, depending upon the degree that the lip seal on the nipple is compromised, but in general, the infant is able to adapt. The feeder can assist with use of a wide base nipple, as well as external provision of lip or cheek support to help maintain lip closure on the nipple.^{4,6}

In contrast to infants with cleft lip, infants with cleft palate have difficulty achieving adequate suction due to the coupling of the oral and nasal cavities. As a result, breast-feeding may be impossible. There are reports that describe the lack of efficiency in the sucking patterns of children with cleft palate. For example, Masarei et al compared sucking patterns of infants with nonsyndromic complete unilateral cleft lip and palate or cleft palate only with a noncleft infant group and found significant differences in regard to sucking

efficiency.⁸ Shorter sucking bursts, faster sucking rates, higher suck-swallow ratios, and increased positive pressure generation were noted in the infants with CL/CP or cleft palate only in comparison with the noncleft cohort. The oral phase feeding problems that are typically encountered in these patients have been described by multiple investigators and include poor oral suction, lengthy feeding times, nasal regurgitation, excessive air intake, and inadequate volume of oral intake.^{4-6,9,10}

Descriptive reports of infants with a minimal cleft of the palate suggest that these infants are often able to adapt and feed without special modifications during bottle as well as breast-feeding.^{1,11} With a small posterior cleft, infants may occlude the cleft with the tongue during sucking and thereby create the condition for negative pressure to be generated normally. In contrast, infants who present with a cleft in the hard and soft palate region may have more significant feeding difficulty initially with both bottle- and breast-feeding. Use of a modified bottle/nipple or cup feeder has been reported as helpful in assisting with feeding efficiency.^{9,12,13} If the mother wishes to breast-feed, consultation with a lactation consultant who has experience with infants with special needs such cleft lip and palate can be advantageous.¹⁴ Interventions may also include nutritional assessment and recommendations to maximize nutritional intake, as well as specific feeding techniques including feeder assisted squeezing for milk expression, pacing of oral intake to provide fluid in rhythm with the infant's movement and reactions, and oral facilitation techniques, such as jaw and cheek support to assist with increasing oral control during feeding.^{6,14}

Craniofacial Syndromes with Cleft Lip with/without Cleft Palate

Overall, there is an increased likelihood of dysphagia in infants and children with craniofacial malformations with and without associated CL/CP.^{15,16} The oral phase of feeding may be affected secondary to factors that impact upon the position and mobility of lip, jaw, and tongue

movements necessary for effective and efficient feeding. Craniofacial anomalies that involve maxillary or mandibular hypoplasia and/or hypoplasia of the midface create the potential for airway obstruction. Therefore, the need for airway management is the first priority.^{17,18} In addition, multiple cranial nerve palsies or abnormalities may be present, leading to impairment of the essential oral-motor and sensory nerve functions necessary for safe and effective feeding. Poor airway protection during swallowing with chronic aspiration may lead to recurrent respiratory illness, pneumonia, and lung damage, which presents significant implications overall in terms of long-term respiratory health.

The potential for parental stress regarding the feeding issues and need for specific feeding instruction are significant issues that need to be addressed by the professionals involved. Overall, it is generally agreed that the risk for syndromic CL/CP-associated feeding difficulty and swallowing dysfunction is great, and there are multiple issues that arise in regard to adequacy and ease of nutritional intake, respiratory health, and the potential for parental/ caretaker stress.¹⁵

The presence of oral, oropharyngeal, and/ or pharyngeal dysphagia has been documented in conjunction with syndromes associated with cleft lip, with or without cleft palate, including trisomy 13, Van der Woude syndrome, Opitz syndrome, orofacioidigital syndrome, Wolf-Hirschhorn syndrome, hemifacial microsomia, CHARGE association, and fetal alcohol syndrome. Perhaps the most severe feeding and swallowing dysfunction occurs in infants with trisomy 13, who are born with multiple birth defects including severe brain abnormalities, congenital heart defects, spina bifida, and often unilateral or bilateral CL/CP. It is known that most infants with trisomy 13 do not live beyond their first birthday, though there are reports of longer-term survivors. These

infants have such severe oral and pharyngeal swallowing impairment that total enteral feedings are necessary to meet their nutritional needs.¹⁵

Van der Woude syndrome is considered to be among the most common syndromic causes of CL/CP. Overall development is normal, and the feeding issues that occur are similar to those experienced by infants with nonsyndromic CL/CP.¹⁵ In contrast, individuals with Opitz syndrome (also known as hypertelorism-hypospadias syndrome, Opitz BBB syndrome, and Opitz Frias syndrome) have multiple features including CL/CP. In addition, a laryngeal cleft may be present, thereby creating the potential for significant airway protection compromise with feeding. The laryngeal cleft may vary in degree, and therefore objective assessment of swallowing by instrumental means, such as videofluoroscopy or endoscopy, is valuable in determining whether airway compromise is occurring.¹⁹ If aspiration occurs in association with oral feeding, subsequent surgical intervention of the laryngeal cleft may be indicated, as well as implementation of the feeding modifications for cleft lip/palate.²⁰

Infants born with orofacioidigital syndrome type I often present with a midline cleft lip with multiple oral tissue webs, cleft palate, as well as tongue abnormalities, such as notching or lobulations.¹⁵ Feeding and swallowing issues occur, not only because of the cleft palate, but also secondary to the restrictions in tongue range of motion necessary for efficient oral transfer and for swallowing initiation.

Wolf-Hirschhorn syndrome is a rare chromosome disorder caused by a deletion or missing portion of the short arm of chromosome 4. Features of this syndrome include cleft lip and palate, hypotonia, microcephaly, and a characteristic facial appearance (described as a "Greek warrior helmet") with hypertelorism and a prominent nasal bridge.¹⁵ Feeding/sucking difficulties have been reported secondary to the CL/CP and poorly coordinated swallowing with aspiration. Poor growth is frequent, along with heart defects and seizures.²¹

Hemifacial microsomia (also referred to as Goldenhar syndrome, oculoauriculovertebral

dysplasia, or facioauriculovertebral spectrum) is characterized by orofacial clefts, facial nerve weakness, limited jaw range, unilateral velar paresis, as well as cleft palate. The facial nerve weakness limits active cheek and lip movements during feeding. The restriction in jaw excursion limits sucking efficiency, and the velar paresis and/or cleft palate contribute to difficulty with generation of negative pressure during sucking and with effective posterior transfer of liquid for swallowing.⁶

Infants with CHARGE syndrome (acronym for coloboma, heart defect, choanal atresia, retarded growth and/or development, genitourinary anomalies, and ear anomalies and/or deafness) have been documented to have significant oral feeding dysfunction. The choanal atresia and resulting respiratory issues, as well as significant cranial nerve abnormalities impacting sensory and motor function in the pharynx and larynx, create severe oral-motor and swallowing difficulty in some individuals.^{22,23}

Lastly, infants born with fetal alcohol syndrome, caused by intrauterine exposure to significant amounts of alcohol, may also present with significant feeding and swallowing issues, secondary to cleft lip and palate. Structural and neurological issues are reflected in varying degrees of oral-motor and swallowing dysfunction. In addition, fetal alcohol syndrome is one of the more common causes of Pierre Robin sequence (PRS) and cleft palate.¹⁵

Craniofacial Syndromes and Sequences with Cleft Palate

Varying degrees of oral and pharyngeal dysphagia have been described in patients with sequences and syndromes known to be associated with cleft palate.^{1,7,15,16,24} Such sequences and syndromes include:

PRS, Stickler syndrome, velocardiofacial syndrome, and Moebius syndrome.

PRS is characterized by retrognathia, posterior placement of the tongue, and often a cleft palate. The posterior position of the tongue (glossoptosis) causes a blockage in the pharynx and airway. The resultant airway obstruction can cause difficulty with coordination of respiration during feeding and swallowing and seriously compromised airway protection, resulting in aspiration.^{1,16,24} In a prospective longitudinal study of feeding skills in a cohort of infants with clefts and syndromes, infants with PRS were found to have a significantly higher risk of feeding problems.⁷ Lidsky et al reported the benefit of early airway intervention for infants with PRS.²⁵ In study of infants divided into groups according to isolated PRS and PRS with additional disorders and syndromes, the investigators found that the infants with isolated PRS who received early airway intervention to relieve upper airway obstruction were more successful with oral feeds. A greater number of infants with isolated PRS who received later airway intervention required gastrostomy tubes, and the infants with PRS with additional disorders and syndromes tended to require gastrostomy feedings regardless of the timing of airway intervention.

Reports indicate that airway management interventions for PRS vary across centers, and treatment decisions are individualized. The options include the use of prone or side-lying positioning, placement of a nasopharyngeal tube, tracheostomy to bypass the area of upper airway obstruction, or mandibular distraction to improve posterior airway space.

Feeding trials are possible while the infant is placed in prone positioning, and there are some reports that indicate this strategy is effective in improving feeding ability.^{4,6} Sidelying positioning, with the infant's trunk straight and well supported by the feeder, has also been reported as effective in helping the retracted tongue fall forward, and therefore facilitating feeding.⁴ Mandibular distraction is a recent advancement in the treatment of airway obstruction that involves an osteotomy (mandibular incision) and placement of a device that allows separation of the two ends of bone. New bone is created in the gap, and

thereby elongates the mandible. This causes forward movement of the tongue base and surrounding soft tissues away from the pharyngeal wall, thus alleviating the upper airway obstruction. Infants who undergo mandibular distraction are unable to feed during the distraction as their jaws are immobilized and they are unable to suck. However, once the distraction procedure is completed, feeding can be resumed. Several reports indicate significant improvement of oral feeding following mandibular distraction.^{26,27}

Stickler syndrome can be variable in terms of the clinical presentation, but is characterized by the presence of PRS including cleft palate, myopia, and sensorineural hearing loss. The issues with feeding are similar to those in infants with PRS.¹⁵

Velocardiofacial syndrome (also referred to as DiGeorge syndrome or 22q11.2 deletion syndrome) is variable in presentation. It typically includes palatal anomalies, such as cleft palate and velopharyngeal insufficiency, congenital heart defects, and dysmorphic facial features.²⁸ Feeding issues have been described in patients with velocardiofacial syndrome, which range from oral-motor dysfunction secondary to low orofacial muscle tone and difficulty with motor sequencing of oral motor movements necessary for efficient feeding. More serious swallowing issues have been reported in patients with velocardiofacial syndrome with accompanying laryngotracheal abnormalities and pharyngeal hypotonia.¹⁵

Moebius syndrome is characterized by bilateral facial and oral musculature weakness secondary to absence or underdevelopment of the abducent and facial nerves. Therefore, the infant may have limited ability to achieve and maintain an adequate lip seal on the nipple for efficient sucking. A high palatal vault may

be present, resulting in potential difficulty in maintaining an adequate tongue-palate seal for efficient sucking. The possible presence of cleft palate contributes to the overall feeding difficulties experienced by the infant with Moebius syndrome.^{29,30}

Craniosynostosis Syndromes

Craniosynostosis is the premature fusion of one or more of the cranial sutures, resulting in abnormal growth of the skull. Children with isolated craniosynostosis generally do not have other malformations or accompanying issues and tend to have an excellent prognosis.¹⁵ In contrast, craniosynostosis syndromes, such as Crouzon syndrome, Apert syndrome, Pfeiffer syndrome, and Saethre-Chotzen syndrome, have varying clinical features that can include feeding problems.¹⁷ Midface hypoplasia is a common presenting feature in Saethre-Chotzen syndrome, and a small number of infants will also have an accompanying cleft palate, creating the potential for difficulty with the oral-motor mechanics necessary for efficient feeding. Infants with Crouzon syndrome have cranial and facial involvement that can also interfere with oral-motor movements necessary for feeding. Cleft palate may also occur with Crouzon syndrome, though this is reported to be uncommon.^{15,17} Infants with Apert syndrome have midface hypoplasia, a narrow palate, and an increased frequency of cleft palate in comparison to Crouzon syndrome.¹⁵ In addition, choanal stenosis and narrowing in the nasal and pharyngeal airway create the potential for respiratory obstruction, which must be resolved prior to presentation of oral feeding. Lastly, infants with Pfeiffer syndrome may present with significant midface hypoplasia and airway compromise resulting from tracheal anomalies and upper airway stenosis. The children have been reported to have significant difficulty with feeding and swallowing in infancy, with poor growth.³¹

Other Craniofacial Anomalies

Treacher-Collins syndrome and Beckwith-Wiedemann syndrome are additional genetic disorders that are often linked with feeding and swallowing issues. Treacher-Collins syndrome has a variable presentation, but usually includes PRS with the accompanying feeding difficulties.¹⁷ Of note is that most individuals with Treacher-Collins and PRS do not have clefts. Infants with Beckwith-Wiedemann syndrome have macroglossia (large tongue), which has the potential to cause respiratory and feeding problems. The tongue may be obstructive and interfere with normal breathing. Limitations in the movement of the tongue may cause difficulty with the degree of motion needed for an efficient oral phase of feeding.⁶

COMMONLY USED FEEDING INTERVENTIONS

Best practice dictates that interventions are evidence-based; however, the availability of evidence supporting specific strategies in the management of feeding and swallowing issues in patients with CL/CP and craniofacial syndromes is limited. Interventions described in the literature vary between centers and between individual practitioners. Often, combinations of interventions are implemented, which makes determination of specific effect on outcomes problematic. Variation in the severity of clefting, the presence and features of accompanying syndromes, as well as other complicating medical issues contribute to the difficulty of determining the effects of specific intervention on patient outcome.

The following is a summary of data-driven reports, as well as reports based upon expert opinion. There is also be a description of feeding interventions found to be effective in the clinical environment.

Oral Feeding Facilitation Techniques
Commonly used techniques to facilitate

oral feeding in infants with nonsyndromic and syndromic CL/CP and craniofacial syndromes include positioning, oral facilitation techniques, assisted fluid delivery, pacing of fluid delivery, and changes in the viscosity of fluid. These techniques are summarized in Table 1.

Optimal positioning as key to successful feeding for facilitation of coordinated jaw, cheek, lip, and tongue movements for sucking/swallowing is frequently described in reports of oral-motor intervention.^{4,10} General positioning recommendations for feeding include provision of head support, with arms forward, trunk in midline, and hips flexed.¹⁰ Positioning the infant with a cleft palate in an upright position of at least 60 degrees will allow gravity to assist with posterior transfer of fluid and with swallowing. It also helps to prevent nasal regurgitation, which may occur secondary to the cleft palate.^{4,6,32}

Direct oral-motor techniques to facilitate nutritive intake may include lip, cheek, and jaw support provided by the feeder.³³ Stabilization of the jaw by placing the middle finger under the chin and the index finger between the chin and the lower lip assists in providing a stable platform for movements of the tongue, lips, and cheeks.⁴ Provision of proprioceptive input to the tongue by use of slight downward pressure to the midline may help to initiate an appropriate sucking pattern.

Table 1 Oral Feeding Facilitation Techniques

Technique	Rationale
Positioning	Maintaining flexion, midline orientation, and neutral alignment of the head and neck facilitates oral-motor patterns, airway protection, and efficient feeding. Positioning at 60 degrees assists with posterior transfer of fluid and decreases tendency for nasopharyngeal reflux.
Assisted feeding	Assistive squeezing, in synchrony with infant's sucking efforts, help to compensate for infant's inability to create suction for fluid extraction. This requires specialized feeding equipment.
External pacing	Imposing pauses during infant feeding helps to maintain appropriate respiratory phase pattern for airway protection.
Lip, cheek, and chin support	Supporting lips, cheek, and chin by external support facilitates sucking movements.
Liquid viscosity	Thickening liquids slightly creates a more cohesive bolus that moves more slowly through the hypopharynx, allowing more time for airway closure to occur. Medical and nutritional consultation is essential so that caloric, fluid, and nutritional needs are met.

Assisted feeding requires the feeder to squeeze the nipple in synchrony with the infant's sucking efforts. Turner et al demonstrated that assisted feeding using a specialized nipple/bottle, such as the Special Needs feeder, was effective in a small prospective study of lactation education and use of an obturator on feeding efficiency.³⁴ Though there are no data-driven studies that provide a strong evidence basis for this technique, there are numerous descriptive reports of the benefit in regard to feeding success and increased volume of oral intake.^{4,6,35}

Pacing the rate of intake is done so that organization in sucking, swallowing, and respiration will be maintained during feeding.³⁶ The feeder monitors infant reaction (color, oxygen saturation, rhythm of sucking, feeding efficiency, respiratory rate) during feeding and identifies cues that signify the need for pausing or changing the pace of intake to help the infant maintain a stable and organized swallowing pattern. Whether the implementation of pacing interferes with the usual rhythm of feeding and creates more difficulty remains to be seen. Current research in identification of respiratory airflow and apneic pause during

swallowing will likely provide a means for objective evidence in the future to support the use of pacing as an evidence-based feeding strategy.³⁷

Increasing the viscosity of fluids to assist with maintenance of airway protection during swallowing is another frequently described interventional strategy with little objective evidence to support. The rationale supporting changes in liquid viscosity or thickened liquid is that the slower flow of the liquid will assist the infant in maintaining airway protection during swallowing. At this time, the reports of the efficacy of using thickened fluid are primarily anecdotal in nature. Medical and nutritional consultation is necessary prior to implementation of thickened fluids to confirm that the infant's caloric, fluid, and nutritional requirements are met.

Table 2 Specialized Feeding Equipment

Type of Equipment	Rationale
Nipple type	<p>Considerations include nipple shape, nipple length, and type and size of nipple hole.</p> <p>Selection depends upon individual infant anatomy and oral motor skills.</p> <p>Must support appropriate oral-motor patterns during sucking.</p>
Special Needs Feeder (formerly Haberman and Mini-Haberman Feeder)	<p>The nipple is designed to release milk from infant compression efforts, so suction is not required.</p> <p>The nipple is soft and pliable with one-way valve that prevents rapid fluid flow and opens only when infant sucks.</p> <p>The nipple is designed for assistive squeezing.</p> <p>Flow control can be regulated by feeder.</p> <p>Mini-special needs feeder is designed for smaller or premature infants with cleft lip/palate.</p>
Soft Cup Advanced Cup Feeder (Medela; McHenry, IL)	<p>The cup is made of soft pliable silicone and is contoured.</p> <p>A control valve, with self-filling cuplike reservoir with flow rate, is controlled by the feeder.</p> <p>Active sucking is not required.</p>
Ross Syringe Nipple with compressible bottle (Abbott Nutrition; Abbott Park, IL)	<p>This very thin, long nipple requires assistive squeeze for formula delivery with close monitoring.</p>
Pigeon Nipple/Bottle (Pigeon Corp.; Chuo-ku, Tokyo, Japan)	<p>The nipple has one side with thick wall for placement against roof of mouth and one side with a thin wall for the infant to suck.</p> <p>The bottle is soft and compressible, allowing assisted feeding.</p> <p>The nipple can be used with other bottles.</p>
Mead Johnson Cleft Palate Nurser (Mead Johnson Nutrition; Glenview, IL)	<p>Soft, compressible bottle is easily squeezed.</p> <p>It is fitted with a cross-cut nipple.</p> <p>A standard nipple can be used with a modified hole as necessary.</p>

Specialized Feeding Equipment

There is a wide variety of specialized nipples, bottles, and cups available for use with infants who present with CL/CP or other feeding difficulties (Table 2). Likewise, there is a wide variety of independent descriptive studies regarding these specialized items that report varying degrees of effectiveness with feeding. Of note are two randomized controlled trials specific to infants with CL/CP that compared feeding equipment and provide moderate to strong evidence regarding the use of specific type of equipment.^{12,13,38} The Mead Johnson cleft palate feeder, combined with a nutrition intervention protocol, was found to be effective in

support of normal growth in infants with cleft palate or combined CL/CP.³⁸ A comparison of

Nipple Type	Pliability	Flow Rate	Shape	Hole Type and Size
Nuk orthodontic nipple (Gerber Co.; Fremont, MI)	Soft	High	Broad base	Hole on surface tip
Standard Nipples	Moderately firm	Low to variable	Round, cross section; straight, taper to flared base	Single, cross-cut or multiple holes
Ross Cleft Palate Syringe Nipple	Soft	Feeder regulated	Long, thin	Large hole
Mead Johnson Cleft Palate Feeder Nipple	Soft	Feeder regulated	Customized	Cross-cut
Special Needs Feeder Nipple	Soft	Feeder regulated	Customized	Slit

a compressible bottle used with an orthodontic nipple was shown to lead to greater weight gain and head circumference in a group of nonsyndromic infants with CL/CP.¹³

There are varying reports from experimental evidence regarding the type of nipples that are most effective, considering the following four nipple parameters: pliability, shape, size, and hole type (Table 3).^{6,39} There is general agreement that the nipple needs to be pliable enough to allow release of formula or breast milk with limited compression, but that the nipple must also be firm enough to provide an appropriate degree of proprioceptive input to stimulate sucking. Reports regarding appropriate flow rate are varied. Overall, there is currently no definitive evidence, other than expert opinion and clinical experience, to guide decision making regarding nipple selection. Generally, the type of nipple chosen should be based on each infant's oral-motor skill ability and on the type and degree of clefting.⁴ Therefore, clinical decision making at this time is based on the infant's success and ease of feeding with a particular nipple, as well as the instrumental assessment of airway protection and swallowing while feeding.

Parental/Caretaker Education on Feeding Technique

Feeding is one of the most immediate challenges faced by parents and caretakers following the birth of a baby with a CL/CP or craniofacial anomaly. Therefore, the clinician's role in assisting with appropriate feeding adaptations is

Table 3 Nipple Characteristics

essential. Prenatal diagnosis of orofacial clefts and early provision of information regarding feeding techniques has been found to be effective in improving patient satisfaction.⁴³ This is important to consider as the early maternal bonding process has been reported to be more disrupted by the initial feeding problems than by the facial defect.⁴⁴ Studies have shown that instruction regarding feeding methods in the newborn period is a parent/caretaker priority immediately following birth.^{8,40-42} Of concern are parental reports detailing the poor quality of advice regarding appropriate feeding techniques and interventions and the fact that they often receive conflicting information regarding the recommended feeding technique within the same institution.⁴⁴

Palatal Obturators

An additional feeding intervention is a feeding obturator, which consists of an acrylic plate inserted into the mouth over the hard palate, essentially closing the palatal defect. Separation between the nasal cavity and the oral cavity can thus be obtained. There are differing reports in the literature regarding the effectiveness of a

palatal obturator for improvement of oral feeding ability.^{45,46} The obturator provides a surface by which the infant can achieve nipple compression; however, there are differing opinions regarding the ability to generate negative pressure. The combined use of a palatal obturator, Haberman feeder, and education by a lactation consultant has been reported to reduce feeding time and significantly increase volume of oral intake.³⁴ Disadvantages of obturators include the expense and the difficulty of use. Intraoral placement of the obturator can be challenging, irritation to the oral tissues may occur, and ongoing adjustment and replacement to accommodate growth is necessary.

Palatal Obturators

An additional feeding intervention is a feeding obturator, which consists of an acrylic plate inserted into the mouth over the hard palate, essentially closing the palatal defect. Separation between the nasal cavity and the oral cavity can thus be obtained. There are differing reports in the literature regarding the effectiveness of a palatal obturator for improvement of oral feeding ability.^{45,46} The obturator provides a surface by which the infant can achieve nipple compression; however, there are differing opinions regarding the ability to generate negative pressure. The combined use of a palatal obturator, Haberman feeder, and education by a lactation consultant has been reported to reduce feeding time and significantly increase volume of oral intake.³⁴ Disadvantages of obturators include the expense and the difficulty of use. Intraoral placement of the obturator can be challenging, irritation to the oral tissues may occur, and ongoing adjustment and replacement to accommodate growth is necessary.

Interventions to Assist with Breast-Feeding

Whether or not breast-feeding is feasible in the infant with CL/CP (syndromic or nonsyndromic) is often an immediate question, and historically opinions have varied. There are descriptive studies that indicate infants with CL/CP are able to successfully breast-feed at least to some degree, though supplemental bottle-feeding may be necessary to meet nutritional needs.⁴⁷ Other reports cite less success with breast-feeding for infants with cleft palate than for infants with cleft lip or no cleft.⁵ In general, the success of breast-feeding (as in bottle feeding) will depend upon the degree of clefting and the status of the airway. Infants with a small cleft in the soft palate may be able to breast-feed with some success; in contrast, infants with more significant palatal clefting will likely have considerable difficulty and require an alternative feeding method.

Most pediatricians and health care providers agree that breast milk is best for infants, and evidence does show that breast-feeding appears to be protective against otitis media in children with CL/CP.^{48,49} Fortunately, clinical protocols have been developed by the Academy of Breastfeeding Medicine to manage common medical problems that may impact upon breastfeeding success, including CL/CP.¹⁴ The protocols serve as guidelines and acknowledge that individual variations in treatment are appropriate according to the needs of each infant. In infants with a cleft lip, the guidelines advise that the infant should be positioned so that the cleft lip is oriented at the top of the breast. The mother may occlude the cleft with her thumb or her finger, or provide cheek support to decrease the width of the cleft and increase closure around the nipple.^{6,14} Semiupright positioning is advised to minimize nasal regurgitation.

For infants with cleft palate, positioning the breast toward the side of the palate that has the most bone promotes better compression and prevents the nipple from being pushed into the area of the cleft. The guidelines state that assessment of breast-feeding potential in infants with syndromic CL/CP should be done on a case-by-case basis, taking into consideration the additional features of the syndrome that might have an impact on breast-feeding success.

CONCLUSION

Feeding and swallowing difficulties vary in severity in the nonsyndromic and syndromic CL/CP population. There are a multitude of medical and neurological factors that impact feeding success and efficiency that the pediatric speech-language pathologist must consider. Compensatory strategies and interventions to assist with the feeding and swallowing difficulties have previously been described, primarily in descriptive studies and case reports rather than extensive systematic research. However, the availability of evidence to support the use of specific techniques, such as specific equipment and assisted feeding methods, is emerging. Continued research to develop a strong evidence base to support the efficacy of specific feeding interventions is needed for best clinical practice.

REFERENCES

- Cooper-Brown L, Copeland S, Dailey S, et al. Feeding and swallowing dysfunction in genetic syndromes. *Dev Disabil Res Rev* 2008;14(2):147–157
- Mizuno K, Ueda A, Kani K, Kawamura H. Feeding behavior of infants with cleft lip and palate. *Acta Paediatr* 2001;91:1227–1232
- Clarren SK, Anderson B, Wolf LS. Feeding infants with cleft lip, cleft palate, or cleft lip and palate. *Cleft Palate J* 1987;24(3):244–249
- Wolf L, Glass R. *Feeding and Swallowing Disorders in Infancy: Assessment and Management*. Tucson, AZ: Therapy Skill Builders; 1992
- Reid J, Reilly S, Kilpatrick N. Sucking performance of babies with cleft conditions. *Cleft Palate Craniofac J* 2007;44(3):312–320
- Arvedson J, Brodsky L. *Pediatric Swallowing and Feeding*. 2nd ed. New York, NY: Singular; 2002
- Reid J, Kilpatrick N, Reilly S. A prospective, longitudinal study of feeding skills in a cohort of babies with cleft conditions. *Cleft Palate Craniofac J* 2006;43(6):702–709
- Masarei AG, Sell D, Habel A, Mars M, Sommerlad BC, Wade A. The nature of feeding in infants with unrepaired cleft lip and/or palate compared with healthy noncleft infants. *Cleft Palate Craniofac J* 2007;44(3):321–328
- Clarren SK, Anderson B, Wolf LS. Feeding infants with cleft lip, cleft palate, or cleft lip and palate. *Cleft Palate J* 1987;24(3):244–249
- Morris S, Klein M. *Prefeeding skills: A Comprehensive Resource for Feeding Development*. Tucson, AZ: Therapy Skill Builders; 1987
- Biancuzzo M. Clinical focus on clefts. Yes! Infants with clefts can breastfeed. *AWHONN Lifelines* 1998;2(4):45–49
- Reid J. A review of feeding interventions for infants with cleft palate. *Cleft Palate Craniofac J* 2004;41(3):268–278
- Shaw WC, Bannister RP, Roberts CT. Assisted feeding is more reliable for infants with clefts—a randomized trial. *Cleft Palate Craniofac J* 1999; 36(3):262–268
- Reilly S, Reid J, Skeat J; Academy of Breastfeeding Medicine Clinical Protocol Committee. ABM Clinical Protocol #17: Guidelines for breastfeeding infants with cleft lip, cleft palate, or cleft lip and palate. *Breastfeed Med* 2007;2(4):243–250
- Saal H. The genetics evaluation and common craniofacial syndromes. In: Kummer AW ed. *Cleft Palate and Craniofacial Anomalies: Effects on Speech and Resonance*. 2nd ed. Clifton Park, NY: Thomson Delmar Learning; 2008:86–118
- Baudon JJ, Renault F, Goutet JM, et al. Assessment of dysphagia in infants with facial malformations. *Eur J Pediatr* 2009;168(2):187–193
- Perkins JA, Sie KC, Milczuk H, Richardson MA. Airway management in children with craniofacial anomalies. *Cleft Palate Craniofac J* 1997;34(2): 135–140
- Andrew T. Airway obstruction in craniofacial anomalies. In: Myer C, Cotton R, Shott S, eds. *The Pediatric Airway: An Interdisciplinary Approach*. 1st ed. Philadelphia, PA: J.B. Lippincott; 1995:247–261
- Willing JP. Benefit of feeding assessment before pediatric airway reconstruction. *Laryngoscope* 2000; 110(5 Pt 1):825–834
- Rahbar R, Rouillon I, Roger G, et al. The presentation and management of laryngeal cleft: a 10-year experience. *Arch Otolaryngol Head Neck Surg* 2006;132(12):1335–1341
- Battaglia A, Filippi T, Carey JC. Update on the clinical features and natural history of WolfHirschhorn (4p-) syndrome: experience with 87 patients and recommendations for routine health supervision. *Am J Med Genet C Semin Med Genet* 2008;148C(4):246–251
- Dobbelsteyn C, Peacocke SD, Blake K, Crist W, Rashid M. Feeding difficulties in children with CHARGE

- syndrome: prevalence, risk factors, and prognosis. *Dysphagia* 2008;23(2):127–135
23. White DR, Giambra BK, Hopkin RJ, Daines CL, Rutter MJ. Aspiration in children with CHARGE syndrome. *Int J Pediatr Otorhinolaryngol* 2005; 69(9):1205–1209
 24. Shprintzen RJ. The implications of the diagnosis of Robin sequence. *Cleft Palate Craniofac J* 1992; 29(3):205–209
 25. Lidsky ME, Lander TA, Sidman JD. Resolving feeding difficulties with early airway intervention in Pierre Robin sequence. *Laryngoscope* 2008; 118(1):120–123
 26. Rothchild D, Thompson B, Clonan A. Feeding update for neonates with Pierre Robin sequence treated with mandibular distraction. *Newborn Infant Nurs Rev* 2008;8(1):51–56
 27. Mandell DL, Yellon RF, Bradley JP, Izadi K, Gordon CB. Mandibular distraction for micrognathia and severe upper airway obstruction. *Arch Otolaryngol Head Neck Surg* 2004;130(3):344–348
 28. Shprintzen RJ. Velo-cardio-facial syndrome: 30 years of study. *Dev Disabil Res Rev* 2008; 14(1):3–10
 29. Sjögreen L, Andersson-Norinder J, Jacobsson C. Development of speech, feeding, eating, and facial expression in Möbius sequence. *Int J Pediatr Otorhinolaryngol* 2001;60(3):197–204
 30. De Serpa Pinto MVX, De Magalhães MHC, Nunes FD. Moebius syndrome with oral involvement. *Int J Paediatr Dent* 2002;12(6):446–449
 31. Cohen MM Jr. Pfeiffer syndrome update, clinical subtypes, and guidelines for differential diagnosis. *Am J Med Genet* 1993;45(3):300–307
 32. Tisza VB, Gumpertz E. The parents' reaction to the birth and early care of children with cleft palate. *Pediatrics* 1962;30:86–90
 33. Einarsson-Backes LM, Deitz J, Price R, Glass R, Hays R. The effect of oral support on sucking efficiency in preterm infants. *Am J Occup Ther* 1994;48(6):490–498
 34. Turner L, Jacobsen C, Humenczuk M, Singhal VK, Moore D, Bell H. The effects of lactation education and a prosthetic obturator appliance on feeding efficiency in infants with cleft lip and palate. *Cleft Palate Craniofac J* 2001;38(5):519–524
 35. Haberman M. A mother of invention. *Nurs Times* 1988;84(2):52–53
 36. Law-Morstatt L, Judd DM, Snyder P, Baier RJ, Dhanireddy R. Pacing as a treatment technique for transitional sucking patterns. *J Perinatol* 2003; 23(6):483–488
 37. Kelly BN, Huckabee ML, Jones RD, Frampton CM. Nutritive and non-nutritive swallowing apnea duration in term infants: implications for neural control mechanisms. *Respir Physiol Neurobiol* 2006;154(3):372–378
 38. Brine EA, Rickard KA, Brady MS, et al. Effectiveness of two feeding methods in improving energy intake and growth of infants with cleft palate: a randomized study. *J Am Diet Assoc* 1994;94(7): 732–738
 39. Mathew OP. Nipple units for newborn infants: a functional comparison. *Pediatrics* 1988;81(5): 688–691
 40. Chuacharoen R, Ritthagol W, Hunsrisakhun J, Nilmanat K. Felt needs of parents who have a 0- to 3-month-old child with a cleft lip and palate. *Cleft Palate Craniofac J* 2009;46(3): 252–257
 41. Kuttenger J, Ohmer JN, Polska E. Initial counselling for cleft lip and palate: parents' evaluation, needs and expectations. *Int J Oral Maxillofac Surg* 2010;39(3):214–220
 42. Young JL, O'Riordan M, Goldstein JA, Robin NH. What information do parents of newborns with cleft lip, palate, or both want to know? *Cleft Palate Craniofac J* 2001;38(1):55–58
 43. Robbins JM, Damiano P, Druschel CM, et al. Prenatal diagnosis of orofacial clefts: association with maternal satisfaction, team care, and treatment outcomes. *Cleft Palate Craniofac J* 2010; 47(5):476–481
 44. Johansson B, Ringsberg KC. Parents' experiences of having a child with cleft lip and palate. *J Adv Nurs* 2004;47(2):165–173
 45. Choi BH, Kleinheinz J, Joos U, Komposch G. Sucking efficiency of early orthopaedic plate and teats in infants with cleft lip and palate. *Int J Oral Maxillofac Surg* 1991;20(3):167–169
 46. Crossman K. Breastfeeding a baby with a cleft palate: a case report. *J Hum Lact* 1998;14(1): 47–50
 47. Garcez LW, Giugliani ER. Population-based study on the practice of breastfeeding in children born with cleft lip and palate. *Cleft Palate Craniofac J* 2005;42(6):687–693
 48. Paradise JL, Elster BA, Tan L. Evidence in infants with cleft palate that breast milk protects against otitis media. *Pediatrics* 1994;94(6 Pt 1): 853–860
 49. Aniansson G, Svensson H, Becker M, Ingvarsson L. Otitis media and feeding with breast milk of children with cleft palate. *Scand J Plast Reconstr Surg Hand Surg* 2002;36(1):9–15