Feeding and Swallowing Issues in Infants With Craniofacial Anomalies

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Abstract

The problems with feeding and swallowing that occur as a result of clefts and craniofacial anomalies range in severity. The extent of clefting, as well as other structural, airway, and neurologic issues, are factors that contribute to potential difficulty with oral feeding mechanics and with the integrity of airway protection during swallowing. Oral motor dysfunction in conjunction with obstruction in the upper airway secondary to anatomic or physiologic anomalies has the potential to cause serious disruption to the necessary coordination of respiration and swallowing. Timely identification of problems is necessary to address threats to nutritional status and/or respiratory health. The pediatric speech-language pathologist has a primary role in the clinical assessment of oral motor/feeding skills, and in the recognition of clinical signs and symptoms of swallowing dysfunction that may warrant referral for instrumental assessment of swallowing function and airway protection. The objective of this article is to summarize the potential feeding difficulties and feeding strategy options for patients with Craniofacial Microsomia, 22q11.2 Deletion Syndrome, Treacher Collins Syndrome, and Pierre Robin Sequence. Awareness of the factors that impact upon the success of feeding and the efficacy of compensatory feeding strategies is key to best practice and successful patient outcomes.

Introduction

Craniofacial anomalies are likely to have a significant impact upon the efficiency and safety of infant feeding. Clefts in the lip and/or palate are the most common anomalies and may or may not be present in the context of orofacial congenital malformations and/or craniofacial syndromes (Arvedson & Brodsky, 2002; Cooper-Brown et al., 2008; Shprintzen et al., 1985). Problems with feeding mechanics and coordination of respiration and swallowing may occur with isolated cleft lip and/or palate, but are usually transient in nature (Masarei et al., 2007). In contrast, feeding and swallowing issues can be significant in the context of complex craniofacial anomalies secondary to upper airway obstruction, cranial nerve abnormalities, and neuromotor factors (Arvedson & Brodsky, 2002; Cooper-Brown et al., 2008; Kummer, 2013; Perkins, Sie, Milczuk, & Richardson, 1997). Inefficient feeding may result in inadequate volume of oral intake, poor weight gain, and ultimately compromised nutritional and developmental status. In addition, airway obstruction as a result of the craniofacial anomaly may cause an inability to achieve or
sustain airway protection during feeding and swallowing, posing a potential threat to respiratory health (Dinwiddie, 2004).

The management of infants with craniofacial anomalies reaches across the scope of multiple disciplines, including speech-language pathology, nutrition therapy, otolaryngology, gastroenterology, radiology, neonatology, plastic surgery, and the pediatrician, underscoring the importance of interdisciplinary communication. The pediatric speech-language pathologist (SLP) serves as a key member of the team in both the assessment and intervention of feeding and swallowing difficulties in infants with craniofacial anomalies. Speech-language pathologists evaluate oral sensorimotor strengths, limitations, and suck-swallow-breathe synchrony during the clinical assessment. Compensatory feeding strategies or interventions may be introduced during the clinical feeding assessment, depending upon the infant’s feeding presentation. Clinical signs and symptoms of airway protection compromise such as persistent coughing, choking, desaturations, color change, and congestion are documented and discussed with the medical team (Arvedson & Brodsky, 2002). An instrumental swallowing examination may be necessary, such as a videofluoroscopic swallowing study (VFSS) or Fiberoptic Endoscopic Evaluation of Swallowing (FEES), to assess swallowing physiology and to define the adequacy of airway protection. The SLP collaborates within the team of professionals who are involved in decision-making regarding oral feeding to design and implement an appropriate feeding treatment plan.

The prevalence, risk factors, and range of feeding problems encountered by infants with cleft lip and/or palate and complex craniofacial conditions is unknown as there are no epidemiologic studies to date that detail this information (Bessell et al., 2011). As such, there are no standardized feeding treatment algorithms and there is limited empirical data available regarding the efficacy of feeding interventions. The purpose of this article is to review feeding and swallowing problems that occur in relatively common craniofacial conditions encountered by the pediatric SLP in clinical practice, including: Cleft Lip and/or Palate, Craniofacial Microsomia, 22q11.2 Deletion Syndrome, Treacher Collins Syndrome, and Pierre Robin Sequence. The impact of comorbidities on feeding and swallowing mechanisms, possible strategies for feeding intervention, and available evidence to support the interventions are described. The anatomic and functional deficits associated with each condition and the possible implications for feeding are described in Table 1. Oropharyngeal deficits and corresponding treatment strategies are defined in Table 2, and available options for specialized feeding equipment are summarized in Table 3.

**Cleft Lip and/or Palate**

Clefts in the lip and/or palate are the most frequent craniofacial defects with a worldwide estimated incidence of 1 in 700 births (Murray & Schutte, 2004). Research shows that there are multiple genetic and environmental causes (Murray, 2002; Murray & Schutte, 2004). The feeding difficulties demonstrated by infants with cleft lip and/or palate (syndromic or non-syndromic) vary and are based on: (1) the extent of the cleft and the infant’s ability to achieve positive and negative pressures for efficient sucking and (2) the infant’s ability to coordinate sucking, swallowing, and respiration during feeding (de Vries et al., 2014; Reid, 2004).

**Infants With Cleft Lip/Palate**

Infants with minimal or incomplete clefts of the lip may exhibit difficulty initially with achieving a lip seal on the nipple, but generally adapt well with minimal feeding alterations (Bessell et al., 2011; Clarren, Anderson, & Wolf, 1987). The choice of nipple (typically wide base) may compensate for the opening in the lip (Clarren et al., 1987). Infants with incomplete and complete clefts of the soft palate demonstrate variable feeding difficulty, characterized by nasal regurgitation secondary to insufficient velopharyngeal closure during swallowing or poor ability to achieve sufficient negative intraoral pressure while sucking (Clarren et al., 1987). The nasopharyngeal regurgitation often can be decreased or eliminated with positional changes (upright) and by the use of specialized feeding systems with placement of the nipple away from
the cleft during feeding. Breastfeeding and/or bottle feeding can usually be achieved with use of the aforementioned feeding modifications (Bessell et al., 2011; Clarren et al., 1987).

In comparison, infants with more extensive clefts of the hard and soft palate have difficulty with liquid extraction as the open palate provides little surface area for compression of the nipple. The infant’s ability to generate sufficient positive pressure (compression) and negative pressure (suction) for efficient sucking is impaired (Clarren et al., 1987; Masarei et al., 2007). Specialized nipple and bottle systems are necessary for the infant to achieve oral feeding (Bessell et al., 2011; Clarren et al., 1987; Reid, 2004) and are described in Table 2. If rooting and sucking reflexes are intact, the aim of treatment is to choose a nipple/bottle system that is compatible with the infant’s anatomy and oral sensorimotor skills. Infants with intact sucking movements are good candidates for infant-directed or self-paced feeding systems. Such systems are designed with a one-way valve in the nipple. When the infant compresses the nipple during feeding, the liquid is released, thus allowing the infant to independently modulate the flow of the fluid.

Alternatively, there are specialized or assisted delivery bottles whereby the feeder assists the infant by squeezing the feeding system in synchrony with the infant’s sucking efforts. Infants with difficulty initiating and maintaining a rhythmic suck-swallow pattern may benefit from the gentle assistive squeezing of fluid by the feeder to help create a liquid flow and stimulates a sucking action (Shaw, Bannister, & Roberts, 1999). It should be noted that squeeze force and duration may vary depending on the feeder; thus care must be taken to stay in synchrony with the sucking efforts of the infant. The feeder must monitor physiologic responses such as color, oxygen saturation, and respiratory rate while feeding, and be alert to any of the infant’s cues that signify the need for pausing or changes in rate of fluid delivery (Glass & Wolf, 1999). A Cochrane review of feeding interventions showed that flexible bottles used for assisted fluid delivery were more effective when used with non-syndromic infants with a cleft lip and/or palate compared to rigid bottles; however, there were no differences in growth outcomes between the groups (Bessell et al., 2011).

In addition to the use of modified nipple/bottle systems, compensatory feeding strategies may be indicated for infants with cleft lip and/or palate (Table 3). The use of external pacing or imposed pauses during feeding have been found to facilitate airway protection in infants who respond with a rapid or poorly coordinated sucking pattern (Law-Morstatt, Judd, Snyder, Baier, & Dhanireddy, 2003). Changes in liquid viscosity to slow bolus transit and facilitate airway protection may be considered for infants who demonstrate poor timing of airway protection during swallowing; however, this strategy must be discussed with the medical team before implementation because of potential medical and nutritional implications (Gosa, Schooling, & Coleman, 2011). Additionally, the provision of thickened fluids with the specialty feeding systems described may be challenging due to limited nipple flows available and the valves used in the self-paced bottles. Last, oral facilitation strategies (lip, cheek, and chin support) may assist with provision of a stable platform for active movements of the tongue, lips, and cheeks during sucking efforts (Clarren et al., 1987; Hwang, Lin, Coster, Bigsby, & Vergara, 2010).

Reports on the feasibility of breastfeeding in infants with cleft lip and/or palate and craniofacial anomalies offer expert opinion and existing evidence to guide practice (Alperovich, Frey, Shetye, Grayson, & Vyas, 2016; Donovan, 2012; Garcez & Giugliani, 2005; Mei, Morgan, & Reilly, 2009; Reid, 2004; Reilly et al., 2013). The Academy of Breastfeeding Medicine (ABM) has developed clinical protocols to serve as guidelines for breastfeeding infants with cleft lip, cleft palate, or cleft lip and palate (Reilly et al., 2013). Non-syndromic infants with a cleft lip may be able to successfully latch on to the breast with the mother’s tissue filling any void in the infant’s labial musculature (Reilly et al., 2013). However, infants with cleft palate and/or associated craniofacial anomalies have difficulty with efficient expression of breastmilk due to their inability to obtain adequate intraoral suction (Reilly et al., 2013). Coordination with a lactation consultant may be helpful to determine optimal positioning during breastfeeding, to assist with latching problems, and to prevent and manage concerns such as inadequate milk supply (Donovan, 2012;
Reid, 2004; Wambach et al., 2005). Pacing or the use of a supplemental nursing system (Table 3) may be employed during breastfeeding depending on the array of issues present; variations in the treatment protocol will depend on each infant’s clinical presentation (Mei et al., 2009; Reilly et al., 2013). Counseling and support for pumping and providing breast milk via bottle is efficacious, especially given the known benefits of breast milk (Alperovich et al., 2016; Reilly et al., 2013).

**Craniofacial Microsomia**

Craniofacial microsomia (CFM) is characterized by underdevelopment of the facial structures (mandible, maxilla, ears, soft tissues, and facial nerves) and is usually a unilateral malformation, although bilateral involvement is reported in 10% of cases (Caron et al., 2015; Heike et al., 2013). CFM is reported to be the second most common craniofacial defect, with a reported incidence ranging from 1 in 3,500 to 1 in 20,000, although there is some variability in prevalence estimates due to diverse inclusion criteria among CFM studies and the existing population-based resources (Caron et al., 2015; Heike et al., 2013). Severity ranges from subtle facial asymmetry to bilateral involvement (typically asymmetric), ear malformations, and respiratory compromise secondary to mandibular hypoplasia (Heike et al., 2013). Accompanying malformations include vertebral, renal, cardiac, and limb anomalies (Heike et al., 2013). The etiology of CFM is unknown but has been associated with prenatal maternal exposures (diabetes and vasoactive medications) and rare genetic variations (Heike et al., 2013).

Infants with CFM are likely to have problems with sucking mechanics secondary to restricted mandibular excursion, facial and masticatory muscle weakness, and abnormalities in the position and range of motion of the tongue (Caron et al., 2015; Heike et al., 2013). Upper airway anomalies in CFM are well documented and negatively impact upon the infant's ability to coordinate sucking, swallowing and respiration, creating the potential for compromised airway protection during feeding (Dinwiddie, 2004; Heike et al., 2013). In addition, the contractile force of the pharyngeal phase of the swallow may be affected by the decreased movement of the pharynx on the affected side secondary to anomalies at the level of the oropharynx (Caron et al., 2015). Co-morbidities associated with CFM include gastrointestinal malformations and congenital heart disease, which may compound the feeding difficulties secondary to digestive, motility, and/or endurance issues (Caron et al., 2015; Heike et al., 2013).

Securing an adequate airway in infants with CFM is the primary concern prior to the introduction of oral feeding trials, as with all craniofacial conditions. The use of complete enteral nutrition via nasogastric or gastrostomy tube to ensure adequate nutrition may be necessary, especially in the period prior to medical intervention of upper airway obstruction. Once adequate respiratory support has been achieved, decisions regarding the appropriateness of oral feeding trials can be made. The SLP’s assessment of the infant’s oral sensorimotor skills (range of movement, strength of sucking, rate of sucking) guides the choice of feeding system. The system chosen should facilitate appropriate tongue position and movement to allow for efficient sucking and the proper flow rate to ensure airway protection (Table 3).

**22q11.2 Deletion Syndrome**

22q11.2 deletion syndrome (22qDS) results from a microdeletion on the long arm of chromosome 22 in a location designated as q11.2. Clinical features are varied, but often include conotruncal cardiac anomalies, palatal dysfunction, immunodeficiency, and hypocalcemia (McDonald-McGinn, Emanuel, & Zackai, 2013). Because of the varied signs and symptoms of 22qDS, groupings of these features historically been classified by a variety of other names, including: DiGeorge Syndrome, Velocardiofacial Syndrome, Conotruncal Anomaly Face Syndrome, and Shprintzen Syndrome.
22qDS is reported to be the most common microdeletion syndrome (Bassett et al., 2011). Population studies have reported the incidence of 22qDS to vary from 1 in 4,000 births to 1 in 5,950 births; reports describe the probable under-diagnosis of 22qDS due to the variability of presenting features (McDonald-McGinn et al., 2013). Congenital heart defects are frequent and have been reported in approximately 75–80% of cases (Bales, Zaleski, & McPherson, 2010; Kobrynski & Sullivan, 2007). The frequency of palatal abnormalities has been reported to be from 69–100%, and includes anomalies such as submucous cleft palate, velopharyngeal insufficiency, and bifid uvula (Kobrynski & Sullivan, 2007).

Feeding difficulties are widely reported in infants with 22q11DS (Bales et al., 2010; Eicher et al., 2000; McDonald-McGinn et al., 2013). Velopharyngeal incompetence and/or hypotonia of the velopharyngeal musculature is a common aspect of 22qDS and may result in nasal regurgitation during feeding (Rommel, Davidson, Cain, Hebbard, & Omari, 2008). Underlying cardiac issues may contribute to poor endurance during feeding and affect overall volume of oral intake. In addition, gagging, choking, and disorganized suck-swallow-breathe coordination during feeding are reported (Eicher et al., 2000; Rommel et al., 1999). Prominence of the cricopharyngeal muscle and abnormal cricopharyngeal function during swallowing may be observed (Eicher et al., 2000; McDonald-McGinn et al., 2013). Cricopharyngeal dysfunction may result in incomplete hypopharyngeal clearance (Rommel et al., 2008) and the potential for aspiration of the pharyngeal residue. Gastrointestinal involvement is a frequent feature of 22qDS with symptoms of dysmotility, vomiting, gastroesophageal reflux disease (GERD), and constipation, all of which may impact upon successful oral feeding (Giardino et al., 2014). Furthermore, laryngotracheoesophageal anomalies, such as laryngeal webs or vascular rings, are occasionally present in infants with 22qDS, which may affect the infant’s ability to maintain a coordinated suck-swallow-breathe sequence during feeding (McDonald-McGinn, Emanuel, & Zackai, 1993; McDonald-McGinn et al., 2013).

Management of feeding issues in infants with 22qDS varies based on the infant’s medical diagnoses and feeding challenges. Sucking dysfunction due to palatal involvement or velopharyngeal incompetence can be effectively managed with the use of specialized cleft bottles and/or nipples. If cricopharyngeal dysfunction is found to negatively impact on swallow efficiency or safety, collaboration with gastroenterology or otolaryngology may be warranted to determine whether medical intervention is required. A variety of techniques, including Botox injections, cricopharyngeal dilation, and surgical interventions such as cricopharyngeal myotomy may be indicated for pediatric patients with significant cricopharyngeal dysfunction (Rommel et al., 2008). Syndromic infants with cardiac involvement may require reduced oral feeding duration or formula with increased caloric density to optimize intake during feeding. Reflux medications may be indicated for GERD symptoms if deemed necessary by the medical team. Infants with severe dysphagia may require nasogastric or gastrostomy tube feedings to ensure appropriate nutritional intake.

**Treacher Collins Syndrome**

Treacher Collins Syndrome (TCS) is an inherited disorder with variable expressivity and is the result of bilateral symmetric anomalies in the first and second branchial arches. The most characteristic feature of Treacher Collins is hypoplasia of the malar bones, maxilla, and mandible with variable effects on the muscles of mastication and the temporomandibular joints (Posnick, Tiwana, & Costello, 2004). Patients with TCS may have an accompanying cleft palate and/or cleft lip, choanal atresia, and an Angle Class II anterior open bite malocclusion (Kummer, 2013; Posnick et al., 2004; Trainor, Dixon, & Dixon, 2009). The occurrence of TCS ranges from 1 in 25,000 to 1 in 50,000 live births (Posnick et al., 2004).

Airway compromise is a primary concern at birth, which arises secondary to micrognathia, posterior tongue position, and the resultant obstruction of the oropharynx and hypopharynx (Trainor et al., 2009). Infants with TCS may have such severe airway obstruction that adequate oxygenation cannot be maintained at rest; therefore, immediate intervention (tracheostomy) is
needed (Trainor et al., 2009). Once the airway is stabilized and the medical team confirms that oral feedings may be initiated, feeding strategies can be trialed by the SLP.

The extent of the mandibular micrognathia and retrodisplaced tongue may result in significant feeding and swallowing difficulties (Posnick et al., 2004). Adaptations in positioning (prone or side-lying) may assist in relieving the upper airway obstruction (Posnick et al., 2004), and can be introduced during the sensorimotor feeding assessment. Depending upon the infant’s non-nutritive oral skills (range and strength), the SLP may present small amounts of nutritive stimulus. Based on the infant’s responses, the SLP will select the nipple, choosing the nipple type/flow rate that corresponds with the infant’s oral sensorimotor skills. If the infant has a cleft palate in addition to micrognathia, use of an assisted delivery bottle with the selected nipple or an infant-paced bottle system may be helpful to assist with an appropriate rate of formula delivery. The use of prone positioning is generally not successful if a cleft is present, as adequate suction will not be present to transfer the liquid for swallowing. Use of upright positioning to at least 60 degrees and pacing may be necessary to ensure the infant sustains a coordinated suck-swallow pattern (Law-Morstatt et al., 2003).

Pierre Robin Sequence

Pierre Robin sequence (PRS) is a congenital condition, characterized by a triad of features: respiratory obstruction, micrognathia, and glossoptosis (Evans et al., 2011; Robin, 1994). A characteristic U-shaped cleft palate is present in 73–90% of infants with PRS (Butow, Hoogendijk, & Zwahlen, 2009; Costa et al., 2014). The reported incidence of PRS is approximately 1 in 8,500 to 1 in 14,000 births (Bush & Williams, 1983; Printzlau & Andersen, 2004). Pierre Robin sequence is a sequence, not a syndrome, which means that an initial primary malformation results in a cascade of secondary anomalies. Underdevelopment of the mandible is considered to be the primary anomaly, reducing the oropharyngeal area (Rathe et al., 2015). This causes posterior and superior displacement of the tongue, which results in glossoptosis, airway obstruction, and a cleft palate due to the incomplete fusion of the palatal plates (Hanson & Smith, 1975; Tan, Kilpatrick, & Farlie, 2013). Feeding and swallowing difficulties are common in infants with PRS (Cruz, Kerschner, Beste, & Conley, 1999; Dudkiewicz, Sekula, & Nielepiec-Jalosinska, 2000) because of these anatomical abnormalities.

Airway obstruction is the primary cause of feeding issues in infants with PRS (Evans et al., 2011). The respiratory effort associated with feeding can result in increased work of breathing and tachypnea, negatively influencing the coordination of airway protection during swallowing (Evans et al., 2011). Sucking efficiency is disrupted due to the cleft palate. Furthermore, glossoptosis prevents central grooving of the tongue around the nipple for effective sucking. Respiratory inefficiency may also increase the infant’s work of breathing during feeding, resulting in early fatigue and cardiopulmonary instability (Rathe et al., 2015). Close monitoring is necessary to determine whether oral feeding is safe or if non-oral nutrition is required until airway stability is obtained. The upper respiratory obstruction noted in PRS may reduce the infant’s suck-swallow-breathe coordination resulting in increased risk of aspiration during feeding. Gastroesophageal reflux (GER) is a frequent comorbidity secondary to altered intrathoracic pressures due to the upper respiratory tract obstruction (Dudkiewicz et al., 2000). Persistent GER may exacerbate feeding difficulties if inflammation or tissue irritation occurs as a result of the reflux episodes (Evans et al., 2011; Mercado-Deane et al., 2001; Suskind et al., 2006).

Specialized cleft bottles and/or nipples may be necessary when feeding trials are initiated. Alterations in feeding position, such as prone or side-lying, use gravity to pull the tongue forward thereby decreasing the likelihood of airway obstruction during feeding (Litman et al., 2005; Park, Thoyre, Knafl, Hodges, & Nix, 2014). Longer nipples may compensate for the posterior displacement of the tongue and aid in attaining the compressive forces necessary for effective sucking (Glass & Wolf, 1999). Pacing and short feeding durations may help to ensure coordinated
airway protection during swallowing (Arvedson & Brodsky, 2002). Despite these strategies, some infants may require nasogastric or gastrostomy tubes to obtain adequate nutrition.

Surgical management may be needed to improve respiratory status in infants with PRS. Opinions differ regarding the optimal management of upper airway obstruction in PRS and interventions vary across centers. Common interventions include nasopharyngeal tubes (to create an airway in mild cases), tongue-lip-adhesion (TLA), mandibular distraction osteogenesis (MDO), or tracheostomy (Cohen, Simms, & Burstein, 1998; Evans et al., 2011; Monasterio et al., 2004). The TLA is performed by suturing the tongue to the lower lip to reduce upper airway obstruction, with a later take-down procedure to remove the suture. MDO is used to lengthen the mandibular bones thus increasing the oropharyngeal space.

**Summary**

The SLP must have an in-depth understanding of the complex interface between sucking, swallowing, and breathing in the treatment of infants with craniofacial anomalies. Understanding of the medical diagnoses is essential, with careful consideration of the interactions between the airway and gastrointestinal tract. Speech-language pathologists are encouraged to gain knowledge about embryology and the medical/genetic etiologies underlying feeding dysfunction. In each of the above described conditions, feeding interventions are individualized to correspond with the infant’s anatomy, oral sensorimotor feeding characteristics, and ability to achieve and sustain airway protection during oral feeding. The availability of evidence to support the use of specific feeding equipment and techniques is accumulating, but more evidence-based research is needed. Current feeding treatment protocols are based upon expert opinion, clinical reports, review articles, and only a small number of randomized control trials (Bessell et al., 2011). There are many opportunities for the future development of standardized treatment algorithms and protocols across conditions, which will facilitate tracking of outcomes. Such data will inform our practice going forward and promote the best feeding outcomes in our patients.
<table>
<thead>
<tr>
<th>Craniofacial Anomaly Description</th>
<th>Potential Impact on Feeding</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cleft lip in isolation, unilateral or bilateral</td>
<td>Variable impact; Incomplete lip seal during breast/bottle feeding may interfere with efficient sucking</td>
</tr>
<tr>
<td>Cleft lip and/or palate, unilateral or bilateral</td>
<td>Variable effect on breast or bottle feeds; may have nasopharyngeal reflux if there is incomplete velopharyngeal closure during swallowing</td>
</tr>
<tr>
<td>Cleft extends into hard palate</td>
<td>Bilateral cleft results in inability to create negative intraoral pressure for efficient sucking</td>
</tr>
<tr>
<td>Bilateral cleft</td>
<td>Nasal regurgitation during feeding due to the inability to seal off nasal cavity</td>
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<tr>
<td></td>
<td>Disorganization of sucking, swallowing and respiration during feeding may result in compromised airway protection</td>
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<td></td>
<td>Prolonged feeding times, fatigue</td>
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<tr>
<td></td>
<td>Breastfeeding is rarely feasible; lack of suction limits ability to latch and transfer milk</td>
</tr>
<tr>
<td>Craniofacial Microsomia</td>
<td>Inefficient sucking due to restricted mandibular excursion and decreased sucking strength due to facial and masticatory muscle weakness</td>
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<tr>
<td></td>
<td>Inability to generate sufficient intraoral pressure for efficient sucking</td>
</tr>
<tr>
<td></td>
<td>Intraoral placement of the nipple onto the tongue body is difficult due to posterior position of tongue</td>
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<tr>
<td></td>
<td>Inefficient sucking secondary to limitation in the range and strength of tongue movements</td>
</tr>
<tr>
<td>Upper airway anomalies</td>
<td>Upper airway obstruction may be exacerbated with respiratory effort of feeding</td>
</tr>
<tr>
<td>22q11.2 Deletion Syndrome</td>
<td>Nasopharyngeal reflux during feeding</td>
</tr>
<tr>
<td></td>
<td>Inability to generate sufficient intraoral pressure for efficient sucking</td>
</tr>
<tr>
<td></td>
<td>Incomplete hypopharyngeal clearance during swallowing, poor velopharyngeal closure during swallowing resulting in pharyngonasal retrograde flow</td>
</tr>
<tr>
<td>Pharyngeal hypotonia</td>
<td>Partial or complete obstruction of flow through the pharyngoesophageal segment; failure of cricopharyngeal muscle relaxation; aspiration</td>
</tr>
<tr>
<td>Criocopharyngeal dysfunction</td>
<td>Fatigue during feeding secondary to cardiac issues</td>
</tr>
<tr>
<td>Cardiac Issues</td>
<td>(Continued)</td>
</tr>
</tbody>
</table>
### Treacher Collins Syndrome

- Hypoplasia of the malar bones, maxilla, and mandible
- Micrognathia and posterior tongue position → obstruction of oropharynx and hypopharynx
- Variable effects on the muscles of mastication and the temporomandibular joints (TMJ)
- Cleft palate
- Choanal atresia (nasal obstruction)

- Restricted range of jaw motion during sucking
- Retrodisplaced tongue prevents nipple from making contact with the tongue body for central grooving around nipple and efficient sucking
- Respiratory effort of feeding may cause increased airway obstruction
- ↑ work of breathing with sucking may cause fatigue during feeding and cardiopulmonary instability
- Reduced strength and efficiency of sucking; reduced range of jaw motion
- Inability to generate sufficient intraoral pressure during sucking if cleft palate present
- Difficulty with coordination of breathing and swallowing, ↑ risk of aspiration

#### Pierre Robin Sequence

- Micrognathia
- Glossoptosis
- Airway obstruction
- "U" shaped cleft palate

- Upper airway obstruction may be exacerbated with respiratory effort of feeding, coordination of airway protection during swallowing is disrupted, ↑ risk of aspiration
- Glossoptosis may prevent placement of nipple onto tongue body for efficient sucking
- Inability to generate sufficient intraoral pressure for efficient sucking

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**Table 2. Feeding Deficits Associated With Craniofacial Conditions and Compensatory Treatment Strategies**

<table>
<thead>
<tr>
<th>Feeding Deficit</th>
<th>Treatment Strategy &amp; Rationale</th>
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<tbody>
<tr>
<td>Nasal regurgitation</td>
<td>Reposition nipple to avoid the nasopharynx during feeding</td>
</tr>
<tr>
<td></td>
<td>Modify positioning to upright (at least 60 degrees) to promote gravity assist with posterior transfer of fluid and limit the amount of liquid entering the nasal passage</td>
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<tr>
<td></td>
<td>May use upright cradle position or elevated side-lying</td>
</tr>
<tr>
<td>Incomplete lip seal during bottle or breast feeding</td>
<td>Wide based nipple to help with occlusion of the cleft</td>
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<tr>
<td></td>
<td>Feeder manually assists with lip closure during bottle or breast feeding</td>
</tr>
<tr>
<td>Inability to compress nipple</td>
<td>Use soft, compressible nipple that delivers fluid with any sucking effort</td>
</tr>
<tr>
<td>Poor initiation of sucking</td>
<td>Use assistive squeezing for small volume (&lt;1mL) of fluid delivery to provide sensory stimulus of taste to help stimulate sucking movements, monitor the infant’s cues closely</td>
</tr>
<tr>
<td>Inability to create negative pressure for fluid extraction</td>
<td>Use specialized nipple that is easily compressed and does not require suction for expression of fluid; self-paced systems allow infant to control flow rate</td>
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<tr>
<td></td>
<td>Use an assisted delivery bottle system</td>
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</tbody>
</table>

(Continued)
Exacerbation of upper airway obstruction with feeding secondary to micrognathia, glossoptosis, midface hypoplasia, upper airway structural anomalies

Modification of position to relieve upper airway obstruction
- Side-lying positioning: less anti-gravity movement during respiration, may promote patency of the upper airway due to the decreased gravitational effects on the tongue and soft palate
- Prone positioning: may relieve upper airway obstruction by promoting anterior positioning of the tongue
*not indicated if cleft palate present

Fatigue during feeding due to effort exerted during feeding and/or underlying cardiac issues

Use of small, frequent feedings
Ensure duration of oral feeding remains under 30 minutes
Careful assessment of appropriate nipple flow rate considering respiratory effort of feeding: faster flow=greater frequency of swallowing, less ventilation time; slower flow=less swallow frequency, more time for ventilation
Increase caloric density of feeds
*requires collaboration with medical team

Poor coordination of sucking, swallowing, and breathing during feeding

Consider nipple flow rate—may need to decrease flow rate to achieve maintenance of airway protection and adequate ventilation during feeding
Consider assistive squeeze pattern—may need to limit volume and rate of each assistive squeeze
Use of external pacing, whereby the feeder imposes external pause intervals to help the infant achieve and maintain a stable and organized feeding pattern throughout the feed
Alteration of liquid viscosity to slow bolus transit and facilitate airway protection during feeding
*requires collaboration with medical team

Table 3. Specialized Feeding Equipment

<table>
<thead>
<tr>
<th>Type of Equipment</th>
<th>Use and Indications</th>
</tr>
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<tbody>
<tr>
<td>• Nipple shape &amp; length</td>
<td>Select nipple shape and length that will provide adequate contact of the nipple on the tongue for central grooving /effective tongue movements during sucking</td>
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<tr>
<td>• Nipple pliability</td>
<td>Firm: provides proprioceptive input, requires adequate sucking strength for compression and generation of negative pressure</td>
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<tr>
<td></td>
<td>Soft: easy to compress with jaw and tongue movements, for example, silicone</td>
</tr>
<tr>
<td>• Flow rates vary per nipple type</td>
<td>Fast flow: requires faster swallowing rate and less ventilation time</td>
</tr>
<tr>
<td></td>
<td>Slow flow: may require infant to exert increased sucking effort and energy expenditure, but may provide more ventilation time during feeding</td>
</tr>
<tr>
<td>Dr. Brown’s Specialty Feeding System®</td>
<td>Infant-Paced Feeding Valve® (unidirectional flow valve) fitted within the base of Dr. Brown silicone nipple allows infant to use spontaneous tongue and jaw movements during sucking to express fluid during feeding at their own pace, does not require suction, feeder does not assist with flow</td>
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<td></td>
<td>Bottle is fully vented to create a positive pressure flow, which reduces air intake during feeding</td>
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<th>Product Description</th>
<th>Details</th>
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| Pigeon™ Nipple/Bottle | Nipple with one-way valve that works with compression effort only, feeder does not assist with flow  
Firm and soft side to nipple—soft side placed on tongue, firm side placed against gum line and palate  
Small notch near rim of nipple serves as air vent  
Pigeon nipple can be used with any bottle type  
Two nipple sizes/flow rates, small size with slow flow, larger size with fast flow |
| Mead Johnson Cleft Lip/ Palate Nurser™ | Assisted delivery bottle with a long cross-cut amber nipple and soft, flexible bottle  
Nipple can be replaced with a shorter, softer nipple based on infant’s sucking characteristics and needs  
Designed for assistive squeezing by the feeder as the baby is sucking, with close monitoring of infant’s cues |
| Medela® SpecialNeeds™ Feeder | Assisted delivery system  
Soft silicone nipple with “Y” cut and one-way valve to prevent backflow of fluid  
Designed to facilitate sucking compression, does not require suction  
Three lines on nipple indicate flow rate, desired flow rate line should be oriented under infant’s nose: shortest line=slow flow, medium line=medium flow, longest line=fast flow  
Fluid delivered in response to infant’s compression efforts; assistive squeeze may be used to deliver fluid in synchrony with the infant’s compression efforts  
Must monitor the infant’s cues during feeding if using assistive squeeze method  
Assistive squeeze may be used to stimulate sucking |
| Bionix Controlled Flow® Baby Feeder | Silicone  
Adjustable flow rate  
0=no flow, for non-nutritive sucking  
1=tastes → 5 =equivalent flow to standard nipple |
| Supplemental Breastfeeding Systems | Feasibility of breastfeeding is determined by type and extent of cleft  
Assisted milk flow during breast feeding  
Multiple systems available  
Lactation consultation for breastfeeding options |

**References**


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