ABSTRACT: Purpose: The purpose of this article is to review historical and current literature and provide clinical relevance related to early development in areas of speech, language, cognition, psychology, and feeding specific to children with velocardiofacial syndrome (VCFS).

Method: The authors reviewed the literature on VCFS in the areas of speech, language, cognition, psychology, and feeding pertaining to the birth-to-3 population. This body of knowledge was then synthesized with relevant literature from the fields of early intervention, language development, articulation/phonology, feeding/swallowing, cleft lip/palate, resonance disorders, and psychology in order to provide speech-language pathologists with a gestalt of evidence-based practices to apply in clinical practice with this population.

Discussion: Diagnosis of VCFS is occurring earlier through fluorescence in situ hybridization testing; thus, early intervention speech-language pathologists (EIs) are more likely than ever before to have children with VCFS on their caseloads (Jones, 2006; Shprintzen, 2005). With an increasing number of children being diagnosed with VCFS in infancy and toddlerhood, and therefore seeking and qualifying for services, it is imperative that speech-language pathologists (SLPs) working as EIs have a working knowledge of the syndrome and appropriate treatment methods.

The prevalence of cleft palate in children with VCFS often indicates a need for SLPS to focus on early speech development. Scherer, D’Antonio, and Kalbfleisch (1999) compared the speech and language of young children (ages 6–30 months) with cleft lip and/or palate in isolation to the speech and language of same-age children with VCFS and cleft lip and/or palate. Findings indicated that the children with VCFS had greater delays in speech and language than the children with cleft palate or cleft lip and palate (Scherer et al., 1999). In addition, young children with VCFS often present with specific needs related to feeding, learning, and socioemotional development. Although the range of behavioral presentation varies considerably among children with VCFS, there is a body of literature that describes early development in children with VCFS.

VCFS, also called Shprintzen syndrome, DiGeorge syndrome, or 22q11 deletion syndrome, was first described as a nosological entity by Shprintzen in 1978 (Shprintzen et al., 1978). He listed the main signs of VCFS as cleft palate, velopharyngeal insufficiency, ventricular septal defects, typical facies, and learning disabilities. Shprintzen described the facies as having a very consistent phenotype expression, including downward slanting palpebral fissures,
wide nasal bridge, flat malar region, retrognathic jaw, and limited facial expression. In the past 30 years since Shprintzen’s report, research on VCFS and the knowledge base of the syndrome has expanded to include more than 180 clinical findings associated with VCFS (Shprintzen, 2005). Some common clinical findings include learning disabilities, cleft palate, pharyngeal hypotonia, lymphoid tissue hypoplasia, cardiac anomalies, retrogrowthia, obtuse cranial base, malar flatness, minor ear anomalies, slender hands and digits, microcephaly, mental retardation, retinal anomalies, small stature, Pierre Robin sequence, and scoliosis (Goldberg, Motzkin, Marion, Scrambler, & Shprintzen, 1993). Indeed, structural anomalies affect nearly every part and system of the body (Robin & Shprintzen, 2005).

Now that VCFS is able to be diagnosed in younger children, it is important that SLPs are knowledgeable about the syndrome. Knowledge of the effects of VCFS on the development of children’s speech, language, cognition, psychological, and feeding skills are relevant for early intervention. However, knowledge without clinical implications is deficient in functional application. Unfortunately, evidence-based practice literature for interventions with young children with VCFS is lacking. The purpose of this article is to report relevant literature related to early development in children with VCFS with clinical implications to stimulate discussion of best practices for SLPs who are working with young children who have received a VCFS diagnosis and their families.

**EARLY SPEECH CHARACTERISTICS**

**Articulation/Phonology**

Anthel, Marrinan, Kates, Fremont, and Shprintzen (2009) reported that intraoral air pressure was significantly impacted in 75% of children with VCFS, resulting in difficulty producing oral consonants. Golding-Kushner (2005) described types of articulation errors that are demonstrated by children with VCFS, including obligatory errors attributable to velopharyngeal incompetence (VPI) and compensatory errors, the most common of which is the glottal stop. As a result, these children substitute glottal stops, pharyngeal stops and fricatives, and nasal fricatives for phonemes, such as stop-plosives, fricatives, and affricates. The glottal stop is a common error that is reported in young children with VCFS, with observations of this production occurring in babbling in children reportedly as young as 8 months (D’Antonio, Scherer, Miller, Kalbfleisch, & Bartley 2001; Golding-Kushner, 2005).

D’Antonio et al. (2001) profiled the early speech characteristics of a sample of children with VCFS as young as 3 years of age, reporting smaller consonant inventories with commonly produced consonants, including nasals; voiceless, anterior stop /p/; and fricatives /f/ and /s/. Gerdes et al. (1999) also reported common articulation error patterns that included the aforementioned glottal stops as well as decreased pressure on consonants and nasal substitution, which Golding-Kushner (2005) categorized as obligatory errors. These types of errors are qualitatively similar to the errors that children with isolated cleft palate produce, but the degree of deficit is often more significant in children with VCFS (Golding-Kushner, 2005; Scherer et al., 1999). The degree of articulation deficit in children with VCFS does improve in school-age children, which is often attributable to a combination of treatment and surgical management of craniofacial anomalies (Shprintzen, 2005).

Motor speech impairments such as apraxia have recently been reported to occur in higher rates in children with VCFS (Kummer, Lee, Stutz, Maroney, & Brandt, 2007). In a recent comparison of apraxia characteristics in children with VCFS and children with cleft lip and/or palate, Kummer et al. (2007) reported that children with VCFS demonstrated more apraxia characteristics than children with cleft lip and/or palate. Prosodic components of speech, including reduced rate of speech and deficits in rhythm of speech, are often impacted in individuals with apraxia and were reported by Solot et al. (2000). Interventions for apraxia characteristics include motor-based approaches focused on repetition and structure as well as intensive treatment, which is often beneficial for children with VCFS (Bauman-Waengler, 2008; Shprintzen, 2005).

Phonological deficits can co-occur with articulation deficits in young children with VCFS and have been reported in investigations of speech development (Golding-Kushner, 2005; Solot et al., 2000). Solot et al. (2000) documented phonological deficits in addition to articulation deficits in two of 10 participants under the age of 5 with enough speech to assess formally. Golding-Kushner (2005) reported that phonological disorders are not more common in children with VCFS, but if present, require modifications to traditional phonological approaches to intervention. Specifically, intervention for children with VCFS should include increased opportunities for repetition and simplified explanations (Golding-Kushner, 2005).

Significantly diminished speech intelligibility is prevalent in young children with VCFS, resulting in these children needing speech services early and intensively to assist in developing oral speech skills (Solot et al., 2000). The impaired ability of the children to be understood is due in part to deficits in articulation as well as co-occurring phonological disorders. However, it is also attributable to the impact of inadequate velopharyngeal function on resonance.

**Resonance**

Children with VCFS display a velopharynx that often does not function properly to effect closure due to cleft palate, pharyngeal hypotonia, or both, thus leading to pervasive severe hypernasality, nasal emission, and nasal turbulence (Jones, 2006; Shprintzen, 2005). Scherer et al. (1999) documented some degree of hypernasality in all children with VCFS who participated in perceptual assessment. D’Antonio et al. (2001) also reported significant hypernasality with documented lack of mobility of either the soft palate or lateral pharyngeal walls using instrumental assessment. Unintelligibility may result from the use of compensatory articulations secondary to VPI (Golding-Kushner, Weller, & Shprintzen, 1985). These children usually require surgery on
the velopharynx to help it to close fully in order to correct the hypernasality and nasal air emission. However, surgery will not correct misarticulations. SLPs must still teach these children correct articulatory placement, manner, and voicing.

Children with VPI cannot build up intra-oral air pressure because air escapes through the nasal cavity. Therefore, before velopharyngeal surgery, in order to teach the contrast between nasal and oral airflow, the nose can be plugged intermittently to prevent air leakage. Either manually squeezing the child’s nose or using a nose clip can be effective. With the nose plugged, the child will be able to build up the pressure required to produce a high-pressure consonant correctly. The easiest stop consonants to begin training are usually [b, p]; the easiest fricative may be [f] due to the visual nature of placement and early development of these phonemes (Bauman-Waengler, 2008). Keep in mind that while the nose is plugged, the child will not be able to produce nasal consonants, so words such as “mop” should be avoided during these treatment tasks.

Language and Cognition

**Expressive language.** A very common clinical sign of VCFS is language delay. In a longitudinal study in which children with VCFS were assessed at 6, 12, 18, 24, and 30 months, significant language impairments were found in their expressive language (Scherer et al., 1999). At 6 months of age, the VCFS group’s mean age-equivalency score for expressive language was 4 months (33% delay), and at 30 months of age, the group’s mean age-equivalency score was 15 months (50% delay; Scherer et al., 1999). In another study of 112 preschool children (ages 4 months to 6 years) who were tested using the Preschool Language Scale (Zimmerman, Steiner, & Pond, 1992), the children’s total language scores showed that 34% of the children with VCFS were significantly delayed (2 SDs below mean), 46% were mildly delayed (1 SD below mean), and 20% were in the average range (Gerdes, Solot, Wang, McDonald-McGinn, & Zackai, 2001).

Roizen et al. (2007) measured 17 developmental milestones that children typically acquire before age 3 in 88 children with VCFS. When compared with age-matched peers, children with VCFS demonstrated significant delays in expressive language, with the gap in expressive language increasing after the first year of life (Roizen et al., 2007). The gap reported for the emergence of babbling in young children with VCFS and children without VCFS was <1 month. This gap expanded to 9 months when reporting on the production of intelligible, single words other than “mama” and “dada” (Roizen et al., 2007).

The delay in expressive language reported by Roizen et al. (2007) is consistent with delays reported by Swillen et al. (1997), but Swillen et al. reported a more significant expressive language delay in children with intellectual disabilities. Because the diagnosis of VCFS places a child with the syndrome in the “established risk” category, language treatment should begin in early infancy to ensure that children with VCFS develop the prelinguistic skills necessary to support language (Rossetti, 2001). Scherer et al. (1999) demonstrated an abnormal course of development, with the gap widening as the child aged from 6 months of age to 30 months. Therefore, according to the principles of evidence-based practice, it would be efficacious for the EI to work with the family from the time the child is a newborn to try to minimize this effect as much as possible.

**Receptive language.** Although expressive language was the most common area of language impacted based on parent report of verbal milestones for children with VCFS, Roizen et al. (2007) reported that these children’s receptive language scores were found to be predictive of later IQ performance. Because of this predictive nature of receptive language skills, it is critical to monitor and enhance receptive language in young children. Scherer et al. (1999) reported that children with VCFS between 6 and 30 months of age demonstrated significant impairments in receptive as well as expressive language. For receptive language, the VCFS group performed within normal limits at 6 months chronological age, but the mean age-equivalency score at 30 months of age was 18 months (40% delay; Scherer et al., 1999). If receptive language deficits are present, then there is greater likelihood of deficits in cognition that will remain until later childhood (Roizen et al., 2007). Because the degree of deficit in expressive skills will likely be greater than any deficit in receptive skills in young children with VCFS, the inclination to focus exclusively on expressive language development is understandable. However, it appears from the literature that receptive language skills might be more predictive of later cognition, which should provide the EI with ample justification for addressing receptive language development in young children with VCFS.

**Cognition.** Cognitive impairment will be an ongoing concern for families with children with VCFS, although children with VCFS demonstrate strengths in areas of verbal IQ and relative weaknesses in perceptual IQ. In an investigation of full-scale IQ (FSIQ) of a large number of children with VCFS, DeSmedt et al. (2007) reported that 60% of the participants had IQ scores >70 and 40% had IQ scores <70, indicating an intellectual disability. DeSmedt et al. reported that children with an inherited form of VCFS, in which the genetic anomaly is passed from parent to child, had significantly lower IQ scores than children who presented with VCFS but did not have the inherited form of deletion. The educational attainment level of the parents of the children with inherited VCFS appeared to be the causal factor for the children’s decreased FSIQ, and DeSmedt et al. hypothesized that this was due to both biological and environmental influences.

Gerdes et al. (2001) studied the cognitive abilities of children with VCFS between the ages of 4 months and 6 years. They found that in infancy, 32% of the children with VCFS were mildly delayed, and 41% were significantly delayed. In toddlerhood, 20% were mildly delayed, and 58% were significantly delayed. By 6 years of age, 34% of the children with VCFS were diagnosed with mental retardation, and 32% were functioning in the borderline range of intelligence. Niklasson, Rasmussen, Oskarsdottir, and Gillberg (2009) investigated 100 consecutively referred patients with VCFS at a children’s hospital in Sweden and found that 50% were diagnosed with mental retardation. Although these studies may not agree on the rate or
severity of cognitive disabilities in the VCFS population, they all clearly demonstrate that a significant portion of children with VCFS will face some degree of cognitive impairment.

**IQ strengths and weaknesses.** When examining this population’s scores on IQ tests, relative strengths have been reported for verbal IQ compared to perceptual IQ. Verbal strengths have been reported most consistently in the areas of reading, spelling, and phonological processing relative to other areas of cognition (Antshel, Fremont, & Kates, 2008; Majerus, Van der Linden, Braissand, & Eliez, 2007). Reading comprehension skills are less developed than reading fluency (Antshel et al., 2008). Persistent verbal difficulties include difficulties with vocabulary development, syntax, and articulation (Majerus et al., 2007). Majerus et al. (2007) reported difficulties in serial order short-term memory tasks for children with VCFS that might be the core deficit underlying their poor vocabulary development. Therefore, EIs should emphasize receptive language development, particularly related to vocabulary development. Stimulating early receptive skills for developmentally appropriate vocabulary and concepts may then facilitate later development of comprehension and memory skills.

Perceptual deficits are consistently reported as a weakness in children with VCFS. Math skills, attention, and executive function skills have all been reported as challenges for children with VCFS (Antshel et al., 2008). De Smedt et al. (2007) reported that children with VCFS had a significant weakness in comparing numbers, which required the children to compare rather than recognize quantities. Attention and organization can be stimulated in young children by focusing on joint attention to tasks and increasing the complexity of play skills to include a variety of actions within focused play. Children who can become problem solvers during early, expanded play experiences also incorporate sustained attention and planning.

**Developmental milestones.** There is limited literature describing developmental milestones in very young children with VCFS, which we have attempted to summarize relative to language and cognition (Antshel et al., 2008; Roizen et al., 2007). Roizen et al. (2007) obtained retrospective data for developmental milestones from parents of children with VCFS that did not indicate that expressive language impairment was predictive of cognitive function. Children with IQ scores >70 reportedly produced their first words at 18 months, whereas children with lower IQ scores spoke their first words at 32 months (Swillen et al., 1997). There is consistency in reports of expressive language delay in young children, but the relationship between expressive language acquisition and cognition is less clear.

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**Psychological Comorbidity**

**VCFS and Attention Deficit Hyperactivity Disorder (ADHD)**

Children with VCFS have long been known to be at risk for developing psychiatric disorders. ADHD, a condition defined by inattentive and hyperactive or impulsive symptoms, is the most common psychiatric condition in this population, affecting between one third and one half of all children with VCFS (Antshel et al., 2007b; Gothelf et al., 2004). Mood disorders, anxiety disorders, and oppositional defiant disorders also occur with increased frequency in the VCFS population (Gothelf et al., 2004). In many cases, these comorbid disorders may not be diagnosed until later in childhood or early adolescence. However, the symptoms often begin to manifest years before the diagnosis is actually made. Children who are eventually diagnosed with ADHD often display the characteristics of impulsivity, overactivity, and developmentally inappropriate levels of attention in early childhood (Jones, Daley, Hutchings, Bywater, & Eames, 2007). General guidance for EIs working with children who exhibit deficits consistent with ADHD include providing support to caregivers and providing interventions that yield positive results in the short term (McGoye, Eckert, & DuPaul, 2002).

One program that has been used to train parents of preschoolers with behaviors consistent with ADHD is the Incredible Years (IY) program. This program teaches parents to (a) follow a child’s lead in order to establish a positive relationship, (b) give children clear instructions and set clear boundaries, and (c) provide children with positive reinforcement for appropriate behaviors (Jones et al., 2007). EIs can adapt these techniques by educating caregivers on developmental expectations for attention, modeling simpler instructions, and providing guidance on establishing clear boundaries during treatment sessions. Parent feedback has shown that training programs that include education on how to provide positive reinforcement are beneficial (Erhardt & Baker, 1990; Jones et al., 2007).

**VCFS and Autism Spectrum Disorder (ASD)**

More recently, several studies investigated the comorbidity of ASD in children with VCFS. One investigation found that 40% of children with VCFS were comorbid for ASD, and of that 40%, 94% had co-occurring psychiatric disorders (Antshel et al., 2007a). A later investigation found a prevalence of 23% comorbidity of VCFS and ASD (Niklasson et al., 2009). Although the rate of comorbidity of VCFS and ASD varied by investigations, both found that a significant number of children with VCFS also had ASD. The high rate of comorbidity between VCFS and ASD found in these investigations justifies screening all children with VCFS for ASD as early as possible, with prompt referrals for ASD assessment if any diagnostic indicators are found.

Kates et al. (2007) described differences in behavior in children with VCFS in isolation and VCFS with comorbid ASD. Children with VCFS + ASD demonstrated impairments in nonverbal social interactions and pretend play in combination with ritualistic, repetitive behaviors and motor stereotypes (Kates et al., 2007). In contrast, children with VCFS had deficits in social interaction consistent with significant early language and speech impairments, including decreased verbal social interactions and initiations as well as deficits in gesture use and specialized interests,
but no ritualistic repetitive behaviors or motor stereotypes. In addition, children diagnosed with VCFS and ASD will have a very different treatment plan than children with VCFS in isolation. The EI should consider the specific additional challenges that children and families will face with a comorbid diagnosis. The treatment plan will need to reflect the communication, social, and behavioral challenges that children with ASD often experience. However, children with comorbid VCFS and ASD still require intervention that prioritizes functional communication and oral speech development.

FEEDING/SWALLOWING DIFFICULTIES

Feeding and swallowing difficulties are common and complex in infants and toddlers with VCFS. Etiologies include airway compromise, vascular anomalies, palatal and pharyngeal anomalies, slow gastric emptying, hypotonia, chronic constipation, and irritable temperament. The complexity and gravity of these medical conditions can be overwhelming to the EI, but appropriate referrals and a team approach to diagnosis and treatment enhances management.

Failure-to-thrive is a pervasive problem in the VCFS population, as it has been found in 74% of infants with VCFS (Shprintzen & Golding-Kushner, 2008). The most common cause of early feeding problems is upper airway compromise (Shprintzen, 2005). Factors contributing to upper airway obstruction are laryngomalacia, laryngeal webbing, vocal fold paralysis, hypertrophic tonsils, hypotonia, and vascular obstructions. Accurate diagnosis of the upper airway obstruction requires imaging such as fiberoptic endoscopic evaluations of swallowing and videofluoroscopic swallow studies. Therefore, EIs must be prepared to refer children who fail to thrive to appropriate professionals for diagnosis.

Cardiac Problems and Feeding

Cardiac anomalies are also prevalent in children with VCFS, appearing in 71% of cases (Shprintzen & Golding-Kushner, 2008). The types of cardiac anomalies vary, with many patients having more than one anomaly. Cardiac problems may cause children to become easily fatigued, negatively impacting sucking and swallowing among infants, which in turn can limit their nutritional intake (Shprintzen & Golding-Kushner, 2008). Cardiac anomalies can also result in arteries or veins wrapping around the trachea or esophagus; if tracheal patency is compromised, the lower airway will be obstructed (Shprintzen, 2005). If the esophagus is impinged on, then bolus transport will be affected. If the esophagus is pinched too tightly, infants will experience persistent emesis (Shprintzen, 2005). This should not be confused with gastroesophageal reflux (GER). In the case of a pinched esophagus, the bolus never reaches the stomach and instead pools in the esophagus above the constricture until enough pressure develops that reverse peristalsis projects the bolus back up (Shprintzen & Golding-Kushner, 2008). It is important that this emesis not be labeled or treated as GER as this could delay the infant receiving proper treatment for the vascular anomaly (Shprintzen, 2005). For infants with VCFS who experience frequent “spitting up,” the EI should not assume that the child will outgrow it but instead should refer the child for imaging studies so that the cause can be diagnosed, following which the EI should coordinate treatment with the cleft palate team.

Cleft Palate and Feeding

If the child with VCFS has a cleft palate, the family will have specific feeding concerns. Most infants with cleft palate learn to feed efficiently rather quickly with proper positioning and special bottles. An infant with a cleft palate should be fed in a more upright position in order to lessen the amount of nasal regurgitation (Kummer, 2008). Feeding may also be eased by appropriate positioning and type of bottle. Positioning the bottle with the nipple under a shelf of bone will give the infant’s tongue a hard surface to compress the nipple against (Kummer, 2008). It may be easier for an infant with a cleft palate to extract milk from a nipple with a large crosscut hole in the tip to allow the milk to flow more readily (Shprintzen, 2005). Bottles that minimize the need for suction are recommended, including squeeze bottles like the Mead Johnson cleft lip and palate feeder or the Haberman feeder (Glass & Wolf, 1999). If the bottle has soft sides, the caregiver can gently squeeze the bottle as the child attempts to suck to help express the milk/formula; the Haberman feeder has a one-way valve enabling efficient flow and compression (see Glass & Wolf, 1999, for a complete review of feeding management).

VPI is present in children with VCFS both with and without a cleft palate. Children with VPI can present with nasal regurgitation during swallowing of liquids and insufficient sucking during feeding (Devriendt, Rommel, & Casteels, 2005). The presence of nasal regurgitation is a sign of possible VPI (Rommel, Davidson, Cain, Hebbard, & Omari, 2008). The presence of VPI can impact speech production as well as feeding and swallowing.

Hypotonia and Feeding

Children with VCFS often have hypotonia, which can affect not only the swallow and bolus transport, but the entire gastric tract (Jones, 2006; Shprintzen, 2005). Constipation is also a common problem in children with VCFS due to anal anomalies (Shprintzen, 2005). These two factors can lead to slow gastric emptying, which is a condition in which food remains in the stomach longer before passing to the intestines, resulting in decreased appetite. Because children with VCFS grow at slower rates than the normal population, concern often arises over increasing their food intake in order to promote growth. However, increasing their food intake may cause gastric discomfort, increase vomiting, and exacerbate constipation. SLPs should be prepared to consult with medically based feeding teams in order to support families. Family education will likely focus on the feeding patterns of children with VCFS and how to provide adequate nutrition that promotes gastric
health and supports a growth rate consistent with VCFS in consultation with the pediatrician and dietician.

**CLINICAL RECOMMENDATIONS FOR FEEDING**

Rommel et al. (2008) reported on three children ages 20 months to 3 years with VCFS who had significant dysphagia, or swallowing difficulty. Two of the three children presented with nasal regurgitation following swallows not because of velopharyngeal or palatal dysfunction; rather, their deficits were attributed to dysphagia occurring at later stages of the swallow related to dysfunction of the upper esophageal sphincter. The specific type of dysfunction experienced, and the treatment recommendations offered, varied with each participant. One participant responded to botulinum toxin A injections that relaxed the upper esophageal sphincter; one participant required only behavioral adjustments and dietary changes, including the introduction of foods with stronger flavor to accommodate some hyposensitivity in the pharynx and spasms that occurred in the upper esophageal sphincter; and the final participant required intervention for both the upper esophageal dysfunction as well as oral deficits such as diminished tongue movement during swallowing. Each of the three participants required an instrumental swallow evaluation. Swallowing is extremely complex, and the EI should be comfortable referring young children for instrumental assessments with feeding teams or SLPs who specialize in pediatric dysphagia.

Babies who do not feel well are often unhappy. The numerous medical complications that infants with VCFS face can cause them to be irritable much of the time. This irritable temperament coupled with gastrointestinal issues often results in poor appetite. However, a well-intentioned parent may try to force the infant to eat more, thereby increasing the infant’s gastrointestinal discomfort and causing him or her to resist feeding, which leads to parental frustration. This cycle results in stressful meal times to which both parent and child react negatively. It is the EI’s responsibility to help families break this cycle by teaching feeding techniques that are appropriate for infants with VCFS. The EI should work with families to establish a feeding pattern that better meets the child’s needs, such as adjusting the amount per feeding and the frequency of feedings.

**RECOMMENDATIONS FOR COMMUNICATION ASSESSMENT AND INTERVENTION**

**Assessment**

The unique developmental profile of children with VCFS will require communication assessments of the children’s speech, language, and early interactions. Standardized tools used to describe early speech and language development include the Preschool Language Scale—3 (Zimmerman et al., 1992), the Peabody Picture Vocabulary Test—3 (Dunn & Dunn, 1997), and the Goldman-Fristoe Test of Articulation—2 (Goldman & Fristoe, 2000). All three speech-language assessments have normative data available for children under 3 years of age (Gerdes et al., 1999; Solot et al., 2000). However, normative data may be insufficient to discern the child’s specific needs and his or her individual strengths to build on during intervention. Criterion-based assessments, such as the Rossetti Infant Toddler Language Scale (1990), enable an analysis of specific behavioral skills across a broad area of communication-related development, including expressive and receptive language, play, and gestural use. For individuals with VCFS who likely have limited expressive language, a criterion-based assessment would allow the SLP to determine the child’s strengths and needs in turn-taking, imitation, and use of gestures such as pointing to meet communication needs.

In addition, parent report of children’s specific behavioral strengths and challenges can be used when observation of behaviors cannot be obtained. Particularly for children with VCFS, examination of the child’s overall communication development is critical for determining intervention.

Articulation assessment of children who are producing verbal language will be enhanced by observation and transcription of articulation errors. D’Antonio et al. (2001) described the frequency of articulation errors in children with VCFS and categorized the errors as developmental or compensatory. Developmental errors included errors such as phonological errors; compensatory errors included glottal stops and pharyngeal fricative substitutions. Scherer et al. (1999) also transcribed glottal stops from videotaped interactions with children with VCFS and cautioned that untrained clinicians might record a glottal stop followed by a vowel production as a vowel-only production. This error results in underestimating the prevalence of compensatory errors and fails to recognize the syllable structure (consonant–vowel) that the child is producing. Videotaping observational samples assists clinicians in checking the reliability of live transcription. Descriptions of articulation errors based on speech samples enable clinicians to recognize phonological patterns of errors, compensatory errors, and phonemic inventories of young children with VCFS.

Assessment of VPI requires instrumental assessment from clinicians who are experienced in working with children with a cleft palate. Due to the prevalence of cleft palate in children with VCFS, the child will likely be under the care of a cleft palate team that will monitor his or her velopharyngeal function. If the child is not being followed by a cleft palate team, the child can be referred to a tertiary care hospital with a velopharyngeal clinic for evaluation and treatment recommendations. The most common instrumental assessments used to assess resonance include nasendoscopy and videofluoroscopy (Lam et al., 2006). These assessments are used to determine the pattern and percentage of velopharyngeal closure and to aid in treatment planning.

**Intervention**

Early intervention teams collaborate on intervention recommendations, which include family priorities as well as the child’s communication needs that were identified during assessment. For SLPs who are providing direct services to the
family, intervention strategies should facilitate the integration of communication into daily activities, such as feeding or play; enhance daily routines; and strengthen relationships between the caregiver and child (Trivette & Dunst, 2005). Although there is not a VCFS-specific methodology for early intervention services, a review of literature specific to VCFS as well as cleft palate early intervention literature can provide some general recommendations for SLPs. The intervention frameworks often integrate areas of speech, language, and social development because development in one area positively impacts another; for example, early vocabulary is linked to intelligibility (Hardin-Jones, Chapman, & Scherer, 2006).

The enhanced milieu (EM) model has been used to facilitate early vocabulary growth by focusing on enhancing the early environment of young children (Hancock & Kaiser, 2006; Hardin-Jones et al., 2006; Hemmeter & Kaiser, 1994). The EM model encourages environmental manipulation to facilitate verbal communication, for example, placing a preferred item out of the child’s reach to encourage the child to request the item (Hemmeter & Kaiser, 1994). Specific teaching strategies are centered on responding to the child during interactions and prompting the child to use functional verbal behavior with different techniques, including modeling followed by a request for verbal imitation or simply a time delay (Hancock & Kaiser, 2006; Hemmeter & Kaiser, 1994; Kaderavek, 2011). A child needs to verbally imitate to participate in the imitation activities, but caregivers can implement this model of intervention in the context of daily activities with children who have a very limited vocabulary (Hancock & Kaiser, 2006). For a child with VCFS, an EM model offers social interaction opportunities that are structured and predictable because of caregiver supports.

EM has been discussed with phonological enhancement (PE) designed for young children with cleft lip and palate (Scherer & Kaiser, 2010). The PE portion is specifically speech recasting in which the caregiver or clinician repeats a child’s utterance immediately following the child’s production with adult pronunciation while maintaining the same grammatical structure (Scherer & Kaiser, 2010; Yoder, Camarata, & Gardner, 2005). Speech recasts enable phonological information in a way that children can process the specific phonological differences between their production and the adult’s immediate reproduction (Yoder et al., 2005). PE continues to be embedded into the EM framework (Scherer & Kaiser, 2010).

DeThorne, Johnson, Walder, and Mahurin-Smith (2009) described evidence-based strategies to use with young children who lack speech imitation in order to facilitate early development of expressive language skills. Strategies recommended to EIIs included providing augmentative and alternative communication, minimizing pressure to speak, imitating the child’s actions and verbal productions (except for glottal productions in the VCFS population), using an exaggerated intonation and slow tempo when modeling expressive language, providing feedback with alternative senses such as visual and tactile feedback, and focusing on function rather than emphasizing nonspeech articulator movements. Gerdes et al. (2001) recommended using a total communication program combining manual signs with verbal productions when working with young children with VCFS. Although Shprintzen (2005) suggested that with a focus on oral speech, manual sign is often unnecessary in young children with VCFS, others argue that total communication programs offer a bridge until the child’s speech becomes more intelligible or until his or her verbal imitation skills improve (Gerdes et al., 2001). Teaching manual signs offers the opportunity to teach nonverbal imitation through sign imitation, which often precedes verbal imitation skills developmentally (Rossetti, 2001).

Oral speech instruction should focus on targeting oral consonants and diminishing the use of glottal stop productions. Establishing placement for pressure consonants such as stop-possives will assist in speech intelligibility and also enable evaluation of the velopharyngeal function as soon as the child is old enough for instrumental assessment of the velopharyngeal port (Hardin-Jones et al., 2006). Scherer, D’Antonio, and McGahey (2008) taught parents to use focused stimulation to target oral speech production in young children with cleft palate. Scherer et al. reported that children in the program increased their speech accuracy consonant productions while decreasing their use of glottal stops. In the focused stimulation program, parents modeled specific words multiple times during play interactions, expanded the child’s production if the child produced a targeted word, and provided corrective feedback if the child attempted a targeted word (Scherer et al., 2008). Modeling, expansion, and corrective feedback focused on developing oral consonants enable parents to stimulate oral communication in children with limited verbal productions.

Focused stimulation and EM techniques occur during interactions between the caregiver or clinician and the child. Children will sustain joint attention, enabling caregivers and clinicians to encourage increased length of interactions, thereby improving attention. Facilitating these early positive social experiences is beneficial to children’s socioemotional development. Gerdes et al. (2001) recommended promoting socioemotional skills in children with VCFS by increasing compliance and increasing social interaction with others, particularly peers. In a discussion of guidelines for educational interventions with young children with VCFS, Swil-len (2001) recommended a total communication program in part because teaching children to use these alternative means to both initiate and respond to others may assist them in establishing turn-taking and basic social interactions. SLPs can encourage positive socioemotional development by improving sustained joint attention, promoting positive social experiences, and encouraging the development of peer relationships, simultaneously improving communication, which is our focus. For more significant early psychological concerns, SLPs should refer families to social workers, pediatricians, or psychologists to meet their medical and psychological needs.

**CONCLUSION**

Velocardiofacial syndrome is a common, complex disorder that is caused by a chromosomal deletion. Children affected...
with this syndrome require a multidisciplinary treatment plan that addresses the multitude of symptoms that affect nearly every part and system of their bodies. The EI should be prepared to address any articulation disorder, language delay, cognitive impairment, psychiatric disorder, and feeding/swallowing difficulties with which children may present either through direct service, consultation, or, in the case of psychiatric concerns, referral. The treatment plan for each child with VCFS will vary according to the severity of each of these challenges and the family’s priorities. The SLP should plan to assess the child’s communication, use clinical judgment to prioritize goals when constructing a treatment plan, and commit him- or herself to working with the child and family through the many years of treatment the child will likely require. Families of children with VCFS face a lengthy course of intervention including treatment, surgery, and medical treatment, and they deserve the assurance of a knowledgeable SLP who can help them give their child the best start in life possible.

REFERENCES


Contact author: Leah I. Fullman, Pediatric Speech Language Pathologist, Children’s Rehabilitation Center, 3004 West Faidley Avenue, Grand Island, NE 68803. E-mail: leah.fullman@gmail.com.