

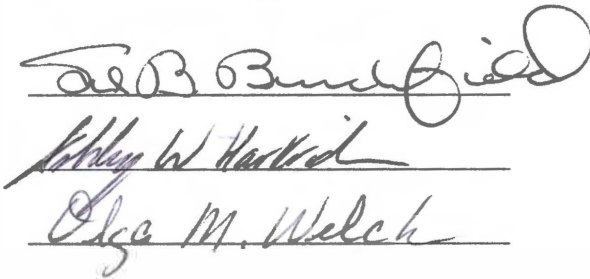
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I am submitting herewith a thesis written by Jill Celeste Fussner entitled "The Determinants of Communication Ability in CHARGE Syndrome." I have examined the final paper copy of this thesis for form and content and recommend that it be accepted in partial fulfillment of the requirements for the degree of Master of Arts, with a major in Audiology.

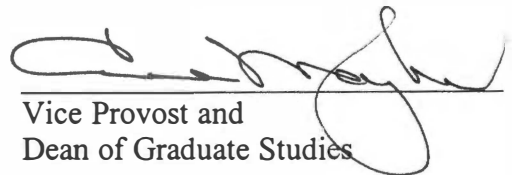


James W. Thelin, Ph.D.  
Major Professor

We have read this thesis  
And recommend its acceptance:



Accepted for the Council:



Vice Provost and  
Dean of Graduate Studies

Thesis  
2003  
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**THE DETERMINANTS OF COMMUNICATION ABILITY  
IN CHARGE SYNDROME**

A Thesis  
Presented for the  
Master of Arts Degree  
The University of Tennessee, Knoxville

Jill Celeste Fussner  
May 2003

## DEDICATION

This thesis is dedicated to my family-

To my parents for planting the seed that has helped me succeed and for supporting me through days of sunshine and rain, and

To my brother who has always been someone to strive to be like. I hope that this may bring me a bit closer to sharing a pair of shoes with you.

## ACKNOWLEDGMENTS

This project is the reflection of the support from so many people – my family, friends, teachers, and all of the CHARGE families.

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I owe a special thanks to all of my friends in my Audiology class – Beth, Stephanie, Danielle, Tommy, Lauren, Jennifer, Brad, Mindy, Amy, and Matt. Each of you has been so wonderful in supporting me through the course of our graduate education. During times of exhaustion and frustration, you lifted me up to get me back on my feet. You have truly become my second family and I love you all.

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## ABSTRACT

Information about the factors that influence the development of communication in CHARGE Syndrome was obtained from parents of 31 children with CHARGE using a questionnaire, personal interview, telephone follow-up interview, and, in some cases, direct observation of the child with CHARGE. CHARGE is a syndrome with multiple congenital anomalies that typically affect physical health and, potentially, all sensory systems. The results of this study indicate that a majority of parents (77%-81%) believe that both physical and sensory problems affected their child's overall development and the development of communication. A majority of the participants had numerous physical anomalies; 100% had vision problems; and 94% had hearing loss.

The 31 participants were divided into two major communication groups: symbolic communication group (n=18) and no symbolic communication group (n=13). The symbolic group was subdivided into groups of participants who used speech as their primary mode of communication (n=11) and those who used sign language (n=6). The no symbolic communication group was subdivided into a gesture group (n=11) and a perlocutionary group (n=2). Comparisons were made between groups and subgroups to determine if there were relations between the prevalence of specific anomalies and the ability to communicate.

The results suggest that development of communication may be compromised by the presence of severe neurologic involvement and severe growth deficiency. There were strong relations between the acquisition of symbolic language and two other factors: the ability to walk independently and total communication training during the first five years

of life. Behavior was initially considered to be a potential factor in the development of communication, but parents unanimously felt that inappropriate behavior was a consequence of a poor ability to communicate.

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# CHAPTER 1

## INTRODUCTION

Hall (1979) described a group of patients with choanal atresia and other congenital anomalies, such as ocular coloboma and small ears. In 1981, Pagon et al. identified a group of patients with anomalies similar to those described by Hall and coined the acronym CHARGE to describe the major features of this genetic association: C = coloboma of eye, H = heart defects, A = atresia choanae, R = retardation of growth and/or development, G = genitourinary anomalies, E = ear anomalies and/or hearing loss. CHARGE has been called an association by some and a syndrome by others; in the present paper it will be referred to as CHARGE. In 1998, Blake et al. revised the diagnostic criteria to include four major features (coloboma, choanal atresia, cranial nerve dysfunction, and the characteristic CHARGE ear), ten minor diagnostic criteria, and thirteen other common findings. For any individual, the number of anomalies that are present may vary greatly, as may the severity of each anomaly. Given this large number of anomalies and variations within anomalies, individuals with CHARGE often have a unique combination of abilities and deficits.

The presence of heart defects, choanal atresia, and/or swallowing problems create survival concerns for infants with CHARGE (Thelin, Hartshorne, & Hartshorne, 1999). Often, the first 2-3 years of life are spent managing these problems. After survival is assured, parental focus then turns to various aspects of development including communication. A number of factors are important in the development of the ability to communicate: a certain degree of physical health, the ability to perceive sensory

information, and the capability for self-expression. Individuals with CHARGE may have deficits in each of these areas – especially sensory deficits. Hearing and vision impairments are often present, and as a result, CHARGE is regarded as a deaf-blind syndrome – however, sensory deficits, such as impairments of smell, touch, and/or balance are also common. CHARGE, therefore, may be more appropriately considered a multi-sensory syndrome. Communication develops as part of an interdependent system with physical development, thus development in one area facilitates the development of the other (Bates, 1979). The purpose of the present study is to evaluate the relation between the development of communication in CHARGE and sensory, behavioral, and physical factors that may influence that development.

## **SENSORY IMPAIRMENTS**

### **Hearing**

In CHARGE, anomalies have been described in all parts of the auditory system (Thelin et al., 1986; Wright et al., 1986). Some form of these anomalies occurs in approximately 90% of cases (Blake et al., 1998). The patterns of external ear anomalies (referred to as characteristic CHARGE ear) and stenosed ear canals are very common, but rarely cause hearing loss (Davenport, Hefner, & Thelin, 1986; Thelin, 1999). Ossicular malformations in the middle ear are a common cause of hearing loss, as is otitis media with effusion that is secondary to eustachian tube dysfunction due to craniofacial anomalies (Edwards, Van Riper, & Kileny, 1995; Shah et al., 1998; Thelin et al., 1986). Cochlear damage and cochlear dysplasia are common causes of significant hearing loss (Dhooge et al., 1998; Edwards, Van Riper, & Kileny, 1995; Franck et al., 2001; Shah et

al., 1998; Thelin et al., 1986). Evidence of auditory nervous system abnormality has been obtained in physiologic studies (Thelin et al., 1986) and anatomic studies (Wright et al., 1986). The most common form of auditory involvement includes ossicular anomalies, middle ear effusion, and cochlear dysfunction that result in significant mixed hearing loss.

Hearing levels in CHARGE range from normal to profound hearing loss. The distribution of better-ear hearing levels reported in four studies is shown in Figure 1 (Dhooge et al., 1998; Edwards, Van Riper, and Kileny, 1995; Shah et al., 1998; Thelin et al., 1986). These data indicate that 15% have normal hearing, 38% have losses in the mild to moderately-severe range, and 47% have losses in the severe-to-profound range. Thus, a majority of individuals with CHARGE have hearing loss that might be expected to have a significant effect on development of communication.

### **Vision**

The most common ocular anomalies in CHARGE are colobomas, or clefts, located in the inferior portion of the iris, retina, macula, or optic nerve. Colobomas occur in 80-90% of cases (Blake et al., 1998). Colobomas of the iris are associated with hypersensitivity to light (Mason, 1997); colobomas of the retina are associated with blind spots, colobomas of the macula and optic nerve also are associated with blind spots in addition to blurry vision (Pagon, 1999). These defects are not correctable with surgery or corrective lenses. Because the blind spot is located in the inferior portion of the eye, images in the upper field of vision may not be seen at all. Thus changes in head position may be required to place images on functioning portions of the retina. In addition, other ocular anomalies may be present as well: cataracts, ptosis, strabismus, retinal detachment,

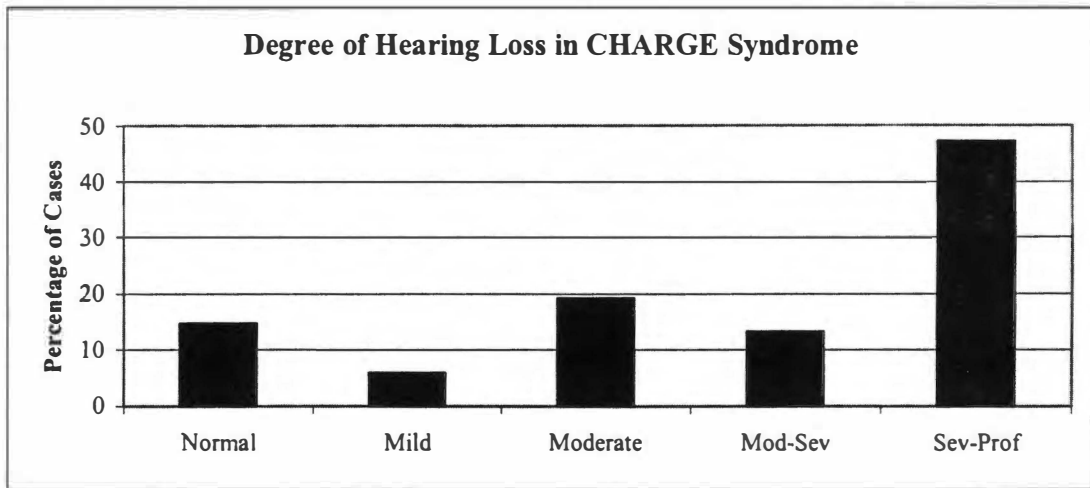


**Figure 1: Degree of Hearing Loss in CHARGE Syndrome**

and/or corneal exposure secondary to facial palsy (Pagon, 1999). With one or more of these visual impairments, focal length may be greatly reduced and vision in face-to-face communication may only be acceptable at close distances. The set of acceptable distances has been called the “communication bubble” (Davenport, 2001). In some cases, visual impairments may make communication using sign language and gestures difficult. Visual impairments may also result in delayed motor development (Pagon, 1999).

### **Other Senses**

Individuals with CHARGE may also have impairments in the other senses - smell, taste, touch, and balance. Cranial Nerve I dysfunction is frequent in CHARGE, thus causing a lack of smell (Blake et al., 1998). A lack or impaired sense of smell may cause feeding problems for children who already have difficulty feeding from swallowing disorders. Malformations of the inner ear may cause anomalies of the semicircular canals (Dhooge et al., 1998), which may result in problems with balance (Wiener-Vacher et al., 1999). Most aspects of touch appear normal in CHARGE. However, by parent reports, children often prefer deep pressure to light touch, a preference that is common in children with autism (Davenport, 2001). Impairments in touch and balance may create difficulties with receptive and expressive communication with gestures and sign language and difficulties with imitative movements and motor skills (Jones, 1988). Parents have reported that their children often have high thresholds for pain; this phenomenon has been associated with inappropriate social interaction (Davenport, 2001). Taste is usually normal in CHARGE, which is important as children with CHARGE often receive information about objects by ‘mouthing’ them (Davenport, 2001). When hearing and

vision, the two primary distance senses, are limited the secondary senses of smell, taste, touch, and balance become the primary resource of sensory stimulation (Jones, 1988). Individuals with CHARGE may have impairments in some or all of these senses.

## **PHYSICAL IMPAIRMENTS**

Blake et al. (1998) have made a comprehensive listing of the physical anomalies in CHARGE and have provided information on the frequency of occurrence for each anomaly. The following is a list of the more common anomalies found in CHARGE:

- |                                                        |          |
|--------------------------------------------------------|----------|
| • Cranial nerve dysfunction                            |          |
| Cranial nerve VII (Facial nerve)                       | 40%+     |
| Cranial nerve IX/X (Glossopharyngeal/ Vagus nerves)    | 70-90%   |
| • Choanal atresia                                      | 50-60%   |
| • Congenital heart defects                             | 70-85%   |
| • Cleft lip and/or palate                              | 20-30%   |
| • Tracheo-esophageal atresia or fistula                | 20%      |
| • Renal anomalies                                      | 40%      |
| • Hypotonia                                            | Frequent |
| • Growth deficiency (short stature)                    | Common   |
| • Laryngomalacia (softness of the laryngeal cartilage) | Frequent |
| • Scoliosis                                            | Common   |

The presence and severity of each anomaly varies greatly across individuals. However, most children with CHARGE have enough serious problems that they are hospitalized many times during the first 2-3 years of life. The average child with CHARGE has more than 20 surgical procedures before 10 years of age (Hefner, 1999a).

## **BEHAVIOR**

Among children with CHARGE, autistic-like behavior has commonly been reported during early childhood, while other behavior problems have been reported later

in life (Blake et al., 1998). There are anecdotal reports of children with CHARGE who have difficulties with sensory integration. Few formal studies have been conducted on behavior; therefore most information has been obtained through parental report.

Carvill (2001) reported that behavior and personality problems are often reported in individuals with hearing impairment. These problems may be related to the lack of communication skills to highlight their distress or anxiety. Personality problems that may be associated with hearing impairment and deafness are unstable and explosive behavior, social and emotional immaturity, lack of tact and empathy, and egocentricity (Denmark, 1966). Carvill (2001) also reported an association between visual impairment and autistic-like behaviors. Sensory deprivation results in behavior problems associated with sensory integration deficits. Impairments in sensory integration may be linked to problems with attention, including unresponsiveness, and regulatory problems, or overreaction to certain situations, increased aggressive or defensive behaviors, decreased activity levels, and poor self-concept (Stephens, 1997).

## **INTELLIGENCE**

Intelligence in children with CHARGE has been a major issue of discussion among parents and professionals. Initially, the “R” in CHARGE included mental retardation because many children did not perform at age-appropriate levels. Parents have objected to classifying their children as mentally retarded because they felt that cognitive development was affected significantly by physical illness and sensory deprivation. An additional consideration is the difficulty in assessing the intelligence of children with multiple anomalies: there are no standardized tests for children with a constellation of

anomalies present in CHARGE. As a result, parents and professionals both recognize that using standardized measures, intelligence is typically underestimated. However, Hartshorne (1999) found that the mean scores for a broad range of abilities were above the number required for a diagnosis of mental retardation.

Despite the discussions among professionals, the issue of intelligence is a sensitive one for parents who are reluctant to underestimate their child's capabilities, even when cognitive development is significantly delayed. In the present study, the decision was made not to ask parents about intelligence because of the difficulty in determining intelligence and because of its emotional content.

## **COMMUNICATION**

Communication is dependent on language, a system that uses rule-governed verbal and/or gestural symbols (Bench, 1992). A language may be described in terms of auditory and visual expressive and receptive abilities (Coplan, 1995). Signed systems and gestures are examples of visual language. Spoken languages are auditory languages with rules for content (semantics), form (phonology, syntax, morphology), and use (pragmatics) (Paul, 2001). To develop language, a nonverbal communication system established through parent/ caregiver interactions must first be in place (Mahoney, 1975).

### **Communication and Hearing Impairment / Deafness**

Individuals who are hearing-impaired or deaf may communicate orally, gesturally, or through a signed system. Dauman et al. (2000) reported on the relationship between mode of communication and degree of hearing loss. Three modes of communication were described: oral, combined oral and gestural (total or bimodal

communication), and purely gestural. Oral communicators typically had a moderate hearing loss, although some with a severe- to-profound hearing loss also communicated orally. Severe-to-profound hearing loss was most common in the oral and gestural group, while the purely gestural group primarily consisted of those with a profound hearing loss.

**Oral Language.** Oral language for the hearing-impaired and deaf may be unisensory or multi-sensory (Paul, 2001). A unisensory approach places emphasis on a primary sense, usually audition, with vision or touch developed later (Paul, 2001). Examples of unisensory methods include auditory-verbal and aural-oral communication. A multi-sensory approach places equal stress on the development of auditory and visual skills (Paul, 2001). This type of approach combines both audition and speechreading. Cued speech, the use of hand positions to complement lip movements, is a type of multi-sensory method (Bench, 1992; Paul, 2001). Oral language for children who are hearing impaired or deaf is delayed when compared to their peers with normal hearing (Davis & Blasdel, 1981; Skarkis & Prutting, 1977).

**Gestured or Signed Language.** Like oral languages, signed languages are also governed by rules. American Sign Language (ASL) is an example of a formal signed system. ASL has rules for content, form, and use that are different from the English language (Paul, 2001). Other formal signed systems may be representative of spoken language. For example, Seeing Essential English, Signing Exact English, and fingerspelling are systems that maintain or approximate the structure of English (Bench, 1992). Gestures may also be used as a form of communication. Gestures may be used to express emotion or they may be used to illustrate spoken words or thoughts (Malandro, Barker, & Barker, 1989).

## **Communication and Deaf-Blindness**

Communication in the deaf-blind population is often not governed by specific rules. While communication may be done through sign in a confined visual field, communication more often is a composite of individual signs, gestures, and/or physical contact (Tedder, Warren, & Sikka, 1993). One study suggested that half of the youth who are deaf-blind do not use symbolic communication (Collins, 1993). This may be a result of the effects of multiple disabilities that prevent language development, or it may result from the lack of availability of language-based communication systems to the deaf-blind (Gothelf & Brown, 1996). Petroff (2001) found a relation between use of language and mobility. He reported that approximately half of youth who are deaf-blind and do not communicate with signed or spoken language also do not walk independently, while the other half of youth who communicate with language also walk independently.

## **Communication in CHARGE**

Communication in CHARGE may be affected by a combination of sensory, physical, and behavior problems. As a result, different modes of communication are used by persons with CHARGE. In a survey of parents of 72 children with CHARGE (Thelin & Stephens, 1996), the following modes of communication were noted:

|                     |     |
|---------------------|-----|
| Speech              | 47% |
| Sign Language       | 38% |
| Own Method          | 50% |
| Communication Board | 7%  |
| Other Methods       | 18% |

The total is greater than 100% because parents were able to report more than one mode of communication. Parents were also asked which problem had the greatest effect on communication. Their responses indicated that a number of factors might contribute to

the development of communication. The disorders that the parents thought had the greatest impact are listed below:

|                                      |     |
|--------------------------------------|-----|
| Hearing Loss                         | 51% |
| Oral motor problems<br>and/or clefts | 22% |
| Retardation                          | 9%  |
| Vision                               | 8%  |
| Overall medical problems             | 5%  |
| Social skills and behavior           | 3%  |
| Facial problems                      | 3%  |

These data suggest that though CHARGE is considered to be a deaf-blind syndrome, hearing loss appears to be of much greater significance than is vision loss in the development of communication. At present there are no data in the literature that describe the effects of multiple disorders on the development of communication.

### **STATEMENT OF THE PROBLEM**

A large percentage of children with CHARGE do not develop normal communication skills. Approximately half of the children with CHARGE do not develop symbolic language, which is similar to the proportion of individuals with deaf-blindness who do not develop symbolic language. In CHARGE, it appears that hearing may be the greatest problem in communication development. Although vision problems also appear to be a factor in the development of communication, there may be other factors that may be more important than vision problems. There are other physical and behavioral factors that may contribute very significantly to the communication difficulties experienced by children with CHARGE. At present there are no studies in which physical, sensory, and

behavioral factors have been evaluated together to determine their relation to communication development.

Information on disorders other than CHARGE has revealed unexpected relationships among physical, behavioral, and sensory factors. For example, there are the findings of increased prevalence of behavioral problems in children with hearing loss and increased prevalence of autistic-like behaviors in children with vision loss. Another finding is the relationship between the ability to use language and mobility in children with deaf-blindness. Since children with CHARGE have multiple physical and sensory anomalies, it is among the most complex of disorders and, as a result, the development of communication may depend complexly on more factors than it does with other disorders.

The purpose of the present study was to perform a comprehensive evaluation of the factors that could potentially influence the development of communication in CHARGE. The methods used to obtain this information were a questionnaire and follow-up interviews with the parents, and in many cases, observation of the child with CHARGE. Parents were asked to make judgments about the disorders that affected overall development and the development of communication, as well as provide factual information about the disorders. The results obtained from the parents were used to determine if there were patterns of anomalies or disorders that were associated with success or failure in the development of communication.

## **CHAPTER 2**

### **REVIEW OF THE LITERATURE**

In 1979, Hall described a group of patients who presented with choanal atresia along with other congenital anomalies, such as cardiac defects, ocular coloboma, small ears, and deafness. In 1981, Pagon et al. described similar anomalies in another group of patients and proposed the term CHARGE association to describe the features. Since that time no consensus has been reached on whether to refer to CHARGE as an association or as a syndrome. The name CHARGE association is used by those who regard the disorder as a nonrandom collection of birth defects of unknown etiology, while the name CHARGE syndrome is used by those who believe the anomalies constitute a specific pattern and are a result of a specific genetic cause (Jones & Dunne, 1988; Thelin, Hartshorne, & Hartshorne, 1999). While an agreement has not been reached on the preferred name, there has been agreement to revise the diagnostic criteria for CHARGE to include four major features (Blake et al., 1998). Those features are coloboma, choanal atresia, cranial nerve dysfunction, and the characteristic CHARGE ear. In addition to these four major anomalies, Blake et al. (1998) have also delineated ten minor criteria and thirteen other common anomalies for diagnosis. Within this wide array of criteria, the number of present anomalies and the severity of each anomaly vary for each individual with CHARGE. Therefore, the specific disabilities and abilities in each case are highly individualistic.

The prevalence of CHARGE syndrome is estimated to be 1:10,000 (Blake et al, 1998). Several patterns of inheritance are possible (Toriello, 1995). However, there are

studies that have suggested increased paternal age (Tellier et al, 1996), chromosomal translocation (Martin, Sheldon, & Gorski, 2001), or neural crest defects (Siebert, Graham, & MacDonald, 1985) as possible causes of CHARGE. Most cases are sporadic, but Brown and Israel (1991) have presented a case of CHARGE in fraternal twins. The recurrence risk for parents with one child with CHARGE is 1-2% (Hefner, 1999b). Children with CHARGE have been shown to have a 70% survival rate to five years of age (Blake, 1999a). The death rate is the highest in the first year of life, which may be attributed to the presence of heart anomalies, choanal atresia, and/or swallowing difficulties (Blake, 1999a; Thelin, Hartshorne, & Hartshorne, 1999). Often, the first 2-3 years of life are spent assuring the survival of the child. Therefore, parental concern with medical issues may result in a lack of attention to communication development until the problems have been managed.

The development of communication is dependent upon the relationship of different factors. Those factors are the ability to perceive sensory information and the ability to be self-expressive, along with a certain degree of physical health. Each of these factors may be affected by CHARGE. The sensory system, in particular, is affected; thus, CHARGE may be considered a multi-sensory deficit disorder.

## **SENSORY IMPAIRMENTS**

### **Hearing**

Anomalies have been described in all parts of the auditory system in CHARGE (Thelin et al., 1986; Wright et al., 1986). The external ear has been called the 'characteristic CHARGE ear' because of the specific patterns of anomalies (Davenport,

Hefner, & Thelin, 1986). The pinna is often short and wide with little to no lobe and is usually cup-shaped. The concha is triangular in shape, the helix often has a snapped off appearance, and there is discontinuity between the anithelix and the antitragus. Furthermore, the pinnas are often asymmetric, low-set, and rotated posteriorly. The external ear canals are usually stenotic, but may be atretic. While external ear anomalies are very common in CHARGE, they rarely result in hearing loss (Davenport, Hefner, & Thelin, 1986; Thelin, 1999). Middle ear anomalies are also common in CHARGE. Ossicular anomalies and otitis media with effusion, secondary to Eustachian tube dysfunction due to craniofacial anomalies, are common causes of hearing loss in CHARGE (Dhooge et al., 1998; Edwards, Van Riper, & Kileny, 1995; Shah et al., 1998; Thelin et al., 1986). Specific middle ear anomalies may include a perforated tympanic membrane, a fused ossicular chain, opacification of the tympanic cavity, and absence of the stapedius muscle (Dhooge et al., 1998; Shah et al., 1998). Recurring otitis media with effusion is a common finding in CHARGE, even at an age when it is uncommon (Dhooge et al., 1998; Thelin et al., 1986). Inner ear malformations are also common causes of hearing loss in CHARGE. An abnormal bony labyrinth, which includes anomalies of the cochlea, the vestibule, and the semicircular canals, is a common finding in CHARGE (Dhooge et al., 1998; Shah et al., 1998). Cochlear malformations range from mild modiolar anomalies to more severe partition defects, such as Mondini's dysplasia. There may also be anomalies of the oval and round windows (Dhooge et al., 1998; Edwards, Van Riper, & Kileny, 1995; Thelin et al., 1986).

The anomalies of the auditory system in CHARGE may result in a conductive or sensorineural hearing loss, but most commonly cause a mixed hearing loss. Thelin et al.

(1986) described a distinctive audiometric configuration found in children with CHARGE. The audiogram was described as wedge-shaped, with flat air conduction thresholds and downward-sloping bone conduction thresholds. While other configurations are common, this shaped audiogram seems unique to CHARGE.

Hearing levels in CHARGE range from normal to profound hearing loss. Figure 1 indicated that 15% have normal hearing, 38% have hearing losses in the mild to moderately-severe range, and 47% have losses in the severe-to-profound range. It would then be expected that those children with a hearing loss in the mild to moderately-severe range might benefit from amplification. However, children with CHARGE present difficulties for appropriate audiological management. Hearing aids are difficult to fit because the earmolds often do not sit well in the malformed conchas and behind-the-ear hearing aids do not fit well on the malformed pinnae (Shah et al., 1998; Thelin et al., 1986). Furthermore, persistent middle ear effusion causes fluctuating hearing loss, which often necessitates intermittent use of hearing aids (Shah et al., 1998). Therefore, hearing loss in any category presents significant difficulty for effective audiological treatment, thus contributing to the difficulty in the development of communication.

### **Vision**

The visual anomalies associated with CHARGE are most often a result of a coloboma. The coloboma may be located in the iris, retina, macula, or optic nerve. The keyhole effect produced by the coloboma may result in sensitivity to light, blurry vision, or blind spots (Pagon, 1999). The blind spots usually affect the upper field of vision, thus creating the effect of wearing a baseball cap. Therefore, children may need to change their head position or turn upside down to make use of the functioning portions of

the retina (Pagon, 1999). Although tinted glasses may be used for photophobia, the visual deficits resultant from colobomas cannot be corrected through surgery or glasses. There are also other visual impairments associated with CHARGE. Facial palsy that is often present in CHARGE can result in a lack of blinking, which then causes the cornea to become overly dry and susceptible to scarring. Artificial tears or gel may alleviate this problem. Other ocular anomalies include strabismus, cataracts, and retinal detachment, all of which may be corrected through surgery (Pagon, 1999).

Decreased visual acuity may result in delayed motor development (Pagon, 1999). Children with a visual impairment have difficulty forming correct concepts of their bodies and conceptually understanding their position in the environment (Stone, 1997). They, therefore, often require specific mobility training. Vision also affects the development of communication. Reynell (1978) observed that a group of preschool children who were blind were delayed by 1 ½ to 2 years in verbal comprehension and expressive language content as compared to their sighted peers, but were later able to achieve at the same level as their peers. However, children with CHARGE, who also have hearing impairment, may not use spoken language. Therefore, visual impairment affects the development of communication by making the use of gestures and sign language difficult (Pagon, 1999). Because the focal lengths may be short, a child with CHARGE may require the communication partner to be close for optimal visual perception. Davenport (2001a) uses the term “communication bubble” to describe this region.

## **Other Senses**

In addition to hearing and visual impairments, CHARGE is associated with impairments of the other sensory modalities – smell, taste, touch, and balance. It is possible that there may be deficits in one or all of these areas. The sense of smell is important, as it is the third distance sense. A lack of smell resulting from cranial nerve I dysfunction may result in feeding problems, as smell is the major part of flavor. Smell is also important in distinguishing different environments and people (Davenport, 2001b). Taste is usually normal in CHARGE, which serves as an important function for ‘mouthing’ objects. This method of object exploration is usually curtailed as communication evolves, but may last longer in children with CHARGE who do not have adequate communication skills (Davenport, 2001b; Kastein, Spaulding, & Scharf, 1980). Most aspects of touch are normal, but children often prefer deep pressure to light touch, a trait often found in autism (Davenport, 2001b). Children may also have increased thresholds for pain, thus resulting in a lack of understanding for why other people react negatively to painful pinches or kicks. Since they cannot feel the pain, they often are unable to empathize with others (Davenport, 2001b). Balance is also often affected in CHARGE as a result of malformations of the vestibule and semicircular canals (Dhooge et al., 1998; Wiener-Vacher, 1999). Balance and mobility are affected by peripheral vestibular dysfunction, ocular anomalies, and hypotonia, all of which may be present in CHARGE. Disordered balance and mobility may cause a delay in motor development (Jones, 1988). As there is a direct connection between movement and learning, the development of communication is also delayed as a result of balance problems (Stone, 1997).

## **PHYSICAL IMPAIRMENTS**

Along with sensory impairments, children with CHARGE present with many significant physical anomalies. Cranial nerve dysfunction is common in CHARGE, particularly dysfunction of the facial, glossopharyngeal, and vagus nerves (Blake et al., 1998). Facial palsy, which may be unilateral or bilateral, often affects feeding, facial expression, and the ability to close the eye completely (Korf, 1999). Anomalies of the glossopharyngeal and vagus nerves result in swallowing and feeding problems, including sucking difficulties, prolonged feeding times, aspiration, nasal reflux, gastroesophageal reflux, and pharyngeal congestion (Abadie, 1999). The consequences of these problems result in adapted diets and feeding procedures for children with CHARGE. In some cases, a gastrostomy tube may be needed. Choanal atresia is also a common finding in CHARGE. The choanae are funnel-shaped openings at the back of the nasal passages that connect the nose and throat. The range of the anomaly extends from unilateral choanal stenosis to bilateral bony choanal atresia (Hall, 1999). Choanal atresia results in respiratory distress and possibly asphyxia, if not treated immediately. Medical management often requires several surgeries to open the nasal passages, while some may need a tracheotomy (Hall, 1999). Congenital heart defects are also usually present in CHARGE. While several defects are possible, the most common are tetralogy of Fallot, double outlet right ventricle, and ventricular septal defect (Lin, 1999). Surgery is often needed early in life, while medications often supplement the procedures. Digoxin, which helps the heart pump stronger, and anti-coagulants, which thin the blood, are common medications (Lin, 1999). Cleft lip and/or palate are also associated with CHARGE. The potential consequences of these anomalies are swallowing, feeding, and speech

difficulties, as well as aspiration and recurrent middle ear infections (Hefner, 1999c). These anomalies require surgical intervention. Esophageal atresia (an esophagus that ends before reaching the stomach) and tracheo-esophageal fistula (a connection between the esophagus and the trachea) may occur in CHARGE (Hefner, 1999d). Both anomalies cause feeding problems and aspiration and must be treated with surgery. Renal anomalies in CHARGE may include solitary kidney, hydronephrosis, renal hypoplasia, and duplex kidney (Williams, 1999a). Such anomalies may be treated with surgery when warranted. Hypotonia, or low muscle tone, is also common in CHARGE, especially in the upper body. With vision impairment, balance problems, and low muscle tone, a child with CHARGE has difficulty sitting alone and may not walk until five or six years of age (Williams, 199b). Growth deficiency is also common in CHARGE. As a consequence of feeding problems, long-term poor nutrition results in growth deficiency and a failure to thrive. Some children with CHARGE may require growth hormone therapy in which daily injections are necessary (Blake, 1999b). In addition to these potential anomalies, there are also many other less frequent anomalies that may occur in CHARGE. The presence of one or more of these anomalies requires extensive medical care and numerous surgeries. Hefner (1999a) stated that the average child with CHARGE is treated by approximately 17 different medical specialists.

## **BEHAVIOR**

In addition to physical anomalies, behavior problems have been reported in children with CHARGE. In childhood, parents have reported that their child demonstrates autistic-like behavior (Blake et al., 1998). Behavior typical to children with

autism may include under-reaction to certain sensory stimuli and failure to use objects functionally (Vinson, 1999). Carvill (2001) and Keeler (1958) reported on the association of visual impairment with autistic-like behavior, such as a lack of communication for interpersonal reasons and rhythmic rocking. Furthermore, hearing impairment may also contribute to autistic-like behaviors, such as sensitivity to loud sounds (Rosenhall et al., 1999). These behaviors may be associated with hearing and visual impairments as a result of sensory deprivation, particularly when the impairments are present in the early childhood years (Carvill, 2001; Rosenhall et al., 1999).

Parents have also reported that as their child enters the school-age years, behavior problems begin to surface (Blake et al., 1998). Sachs (1989) noted that when communication abilities are poor, a child's social interactions and emotional attitudes are affected. Children with CHARGE have difficulty interpreting input from the different sensory systems of the body, which results in sensory integration dysfunction. Children with integration problems may be unresponsive, easily distracted, irritable, difficult to soothe, or hypersensitive to touch. They may also overreact to certain environmental stimuli or lack purpose in their activity (Stephens, 1997).

## **COMMUNICATION**

Communication is primarily expressed through language, a system that may be conveyed auditorily or visually (Coplan, 1995). Prior to the emergence of intentional language communication, an infant develops a nonverbal communication system through interacting with parents or caregivers and the environment (Mahoney, 1975). During this time the infant learns to control cries and babbles and realizes that these sounds can

influence others to do things (Paul, 2001). The basis for turn-taking is also established during this time, as the caregiver will adjust behaviors according to the infant's sounds, cries, and facial expressions (Appell, 1987). By the end of the first year of life with the onset of object permanence and control over sounds, the child begins to produce single words (Paul, 2001). Language develops through phonology, the production of sounds; semantics, the meaning of words; syntax, the grammar of the language; and pragmatics, the use of the language (Paul, 2001). As the child enters into the pre-school years and develops linguistic maturity, conventional and conversational speech acts will be advanced (Paul, 2001). Conventional speech includes getting attention and making a request, while conversational speech consists of turn-taking and topic development. The child will also develop metalinguistic knowledge about acceptable and unacceptable forms of language (Paul, 2001). By the age of four or five years, most children have internalized much of the grammar of a language and have a significant number of metalinguistic skills (Paul, 2001).

### **Communication and Hearing Impairment / Deafness**

Language is dependent on perception; therefore, an impaired sense of speech perception resulting from hearing impairment will result in problems with language acquisition (Bench, 1992). The degree of hearing loss has a significant impact on the use of oral language by the child and the other modes of communication that may be used with or in place of spoken language (Dauman et al., 2000).

**Oral Language.** There are different approaches to helping a child with hearing impairment acquire oral language. An auditory-verbal approach supports the child's

ability to listen and use verbal communication without using speechreading as a cue (Dornan, 1999). On the other hand, an aural-oral method promotes listening, speech, and the use of available speechreading cues (Paul, 2001). Cued speech uses a combination of talking and using different hand shapes and positions to clarify speech sounds that are not visible (Paul, 2001). In order for the child to effectively communicate using cued speech, others must learn the system.

Oral language used by children who are hearing-impaired may only be governed in part by rules as a result of the incomplete information provided by speechreading and the amount of useable hearing (Paul, 2001). Syntactic, semantic, and pragmatic development of children with hearing impairment is similar to that of their normal hearing peers but occurs at a slower pace (Davis & Blasdel, 1975; Skarakis & Prutting, 1977).

**Gestured and Signed Language.** Signed languages are formal languages with rules for grammar, semantics, pragmatics, and intonation (Paul, 2001). Signed languages are kinesthetic in nature and, therefore, require motor functioning (Bench, 1992). Signed systems also rely on vision for communicative interactions and to exchange the many facial expressions involved when signing (Bench, 1992). Some signed systems mimic the English language in structure, while others do not. American Sign Language (ASL) does not follow the grammatical or pragmatic rules of English, while Seeing Essential English has more of an English basis as it uses ASL signs with other signs to convey inflections, such as prefixes and suffixes (Bench, 1992). Signing Exact English is signed in a way that is consistent with how English is spoken (Paul, 2001). Although it is rarely used alone, fingerspelling consists of signs that correspond to each letter in the alphabet.

While signs tend to communicate content, fingerspelling communicates function and can be used to connect signs into sentences (Bench, 1992).

Gestures may be used in conjunction with another communication system or by they may act as the primary means of communication. Gestures may be used to express emotion, such as anxiety (Malandro, Barker, & Barker, 1989). These gestures may be body-focused as in the form of scratching or object-focused like fidgeting with an article of clothing (Malandro, Barker, & Barker, 1989). Gestures may also be used to illustrate spoken words, such as pointing (Malandro, Barker, & Barker, 1989).

### **Communication and Deaf-Blindness**

Communication used by children who are deaf-blind may consist of a number of modes. Petroff (2001) reported that a high percentage of youth who are deaf-blind use gestures, vocalizations, facial expressions, and/or behavior as the primary mode of communication. A small number of youth use sign language as their primary mode (Petroff, 2001). When other disabilities are present in addition to deaf-blindness, the methods of communication are more likely to be at the informal, pre-language level (Tedder, Warden, & Sikka, 1993). It is possible that a person who is deaf-blind and has other severe impairments will use non-symbolic communication throughout life (Siegel-Causey & Downing, 1987).

### **Communication in CHARGE**

As reported by parents of children with CHARGE, hearing loss is regarded by a majority of the parents to be the most significant contributor to their child's

communication abilities (Thelin & Stephens, 1996). While CHARGE is considered a deaf-blind syndrome, parents rated vision at a much lower significance level. Between hearing and vision was a list of different physical and behavioral factors. Therefore, in order to gain a complete view of the determinants of communication ability in CHARGE, it is necessary to evaluate the effects of sensory, physical, and behavioral conditions of the child.

## **CHAPTER 3**

### **METHODS**

#### **PARTICIPANTS**

The participants were children or young adults with a formal medical diagnosis of CHARGE syndrome (or association) and their parents. The median age of the participants was 7 years and the range was one to 27 years of age. One group of participants (n=25) was families that attended the Mid-Atlantic CHARGE Syndrome Conference in Aberdeen, Maryland from June 21-23, 2002 or families that received information about the Mid-Atlantic CHARGE Syndrome Conference but were unable to attend. Another group of participants (n=6) was families who were members of the CHARGE Syndrome Canada organization.

#### **PARENT QUESTIONNAIRE**

The parents completed a nine-page questionnaire on the physical, sensory, and behavioral conditions that affected their child with CHARGE. The questionnaire (Appendix A) was designed to obtain parental judgments of the overall significance of problems in the three major research categories. After parents provided overall judgments, they were asked to identify the specific anomalies or conditions that contributed to the overall judgments. A checklist of specific conditions was provided for the parents. Judgments were made with regards to communication and on the basis of the child's general state. Following each section, the parents were permitted to elaborate on the factors that they checked or they could have described other less frequent CHARGE conditions that were not listed but that they felt were important. After descriptions of the

child's physical, sensory, and behavioral conditions, the parents were asked to provide a description of their child's ability to communicate: mode of communication, communication abilities, the type of communication training/therapy the child received, the child's educational level, and the age appropriateness of that level.

## **EXPERIMENTAL PROTOCOL**

### **American Experimental Group**

The initial phase of the experiment was done in conjunction with the Mid-Atlantic CHARGE Syndrome Conference. Prospective applicants to the conference were mailed information about the study (Appendix B). Interested parties returned the information form by mail or by email. Several weeks before the conference, questionnaires were mailed to the parents along with informed consent forms. The informed consent form (Appendix C) included three parts: consent to provide information on the questionnaire and supporting medical information, consent to meet with the investigator at the conference, and consent to be contacted after the conference to clarify any information obtained in the questionnaire. Parents were asked to return the questionnaire by mail, email, or by bringing it to the conference. In many cases, preliminary analysis of the questionnaires took place prior to the conference; in other cases, parents completed the questionnaire at the conference. At the conference, meetings were arranged with parents to discuss the questionnaires. In cases when further clarification of information was needed or in cases in which the family was unable to attend the conference, parents were contacted after the conference by telephone, mail, or email. Following the completion of the study, parents were given a summary of the study's results.

### **Canadian Experimental Group**

Contact was made with the president of the CHARGE Syndrome Canada organization. Members of the organization were emailed information about the study (Appendix B) and interested parties returned the information form by mail or by email. The questionnaire (Appendix A) and the informed consent form (Appendix C) was then mailed to the interested family. The informed consent form included two parts: consent to provide information on the questionnaire and supporting medical information and consent to be contacted by phone or email to clarify information on the questionnaire. Parents returned the questionnaire and the consent form by mail. Each questionnaire was reviewed prior to parent contact. Following the completion of the study, parents were given a summary of the study's results.

### **DATA ANALYSIS**

Questionnaire data were tabulated in two-page tables with 64 entries in each table. This data format was used to display all of the parent responses except for a few which were edited out (The reasons for editing are described in the RESULTS.). There were four tables of this type:

- A description of the parent responses for all of the participants.
- A comparison of responses for symbolic and no symbolic language groups.
- A comparison of speech and sign language groups.
- A comparison of gesture and perlocutionary groups.

Using the data in these tables, nearly 200 pair-wise comparisons were made to determine if groups could be differentiated on the basis of any of the responses obtained from parents.

## CHAPTER 4

### RESULTS

#### RESEARCH CATEGORIES

The questionnaire used in this study was divided into six major categories:

- Primary Mode of Communication
- Communication Training
- Education
- Physical Health
- Sensory Function
- Behavior

The first three categories were used to describe the participants in regard to communication and development. The other three categories, “Physical Health,” “Sensory Function,” and “Behavior,” were used to obtain information about disorders that could have potentially affected overall development and development of communication.

In each of these categories, the initial two questions to the parents required judgments regarding the effect on overall development and the development of communication on a five-point rating scale. Based on the investigator’s observation of the children in this study and on parent responses, it appeared that parents had a good basis for determining the presence of significant involvement but a poorer basis for judging the degree of involvement – relative to other children with CHARGE. For example, the investigator observed children that she considered to have very significant involvement (rank 4) while the parents judged the involvement to be only significant (rank 3). In other cases, the investigator and the parents’ judgments were reversed

between ranks 3 and 4. However, there were only rare instances in which the investigator and the parent judgments disagreed between those in ranks 1, 2, and 3 (no significance, slight significance, and some significance) versus those in ranks 4 and 5. As a result, the decision was made to reduce the data into two groups: (1) minimal or no significant effects (rankings 0, 1, and 2) and (2) significant effects (rankings 3 and 4). It was felt that this binary ranking scale more accurately reflected the precision of the parents' judgment abilities. In the reporting of the data, the number and percentage of significant effects is provided for each judgment question.

Within each category, following the judgment questions, parents were asked to provide factual information that was related to the disorders in that category. For this type of question, the data that are reported are the number of responses of each type and the percentage of the total number of possible responses.

## **RESULTS FOR ALL PARTICIPANTS**

The data for the 25 American participants and the six Canadian participants are shown in Appendix D. The data from these two groups were sufficiently similar, based on visual inspection, that the decision was made to pool the data to form a single experimental group. The data for all participants are shown in Table 1. Table 1 is a listing of the questions that were contained in the questionnaire. The data are discussed below, by category, in the order in which they appear on the table. The discussion provides an overall description of the characteristics of the participants in this study. The results for individual participants are shown in Appendix E.

**Table 1: Profile of All Participants**

| TOTAL (n=31)<br># of Participants (%)                  |         |
|--------------------------------------------------------|---------|
| <b><u>PRIMARY MODE OF COMMUNICATION</u></b>            |         |
| Sig. effect of communication on overall development    | 23 (74) |
| Sig. effect of CHARGE on comm. development             | 27 (87) |
| <b><u>Communication Mode</u></b>                       |         |
| Speech                                                 | 11 (35) |
| Sign Language                                          | 6 (19)  |
| Gesture                                                | 11 (35) |
| Perlocutionary                                         | 2 (6)   |
| <b><u>PHYSICAL HEALTH</u></b>                          |         |
| Sig. effect of physical health on overall development  | 25 (81) |
| Sig. effect of physical health on comm. development    | 24 (77) |
| <b><u>Physical Conditions</u></b>                      |         |
| Choanal atresia                                        | 23 (74) |
| Heart problems                                         | 23 (74) |
| Cleft lip/palate                                       | 10 (32) |
| Swallowing/feeding problem                             | 27 (87) |
| Facial problems                                        | 17 (55) |
| Hypotonia                                              | 25 (81) |
| Growth deficiency                                      | 17 (55) |
| Tracheostomy                                           | 2 (6)   |
| Gastrostomy tube                                       | 18 (58) |
| Neurologic problems (structural)                       | 5 (16)  |
| Neurologic problems (other)                            | 5 (16)  |
| <b><u>SENSORY FUNCTION</u></b>                         |         |
| Sig. effect of sensory function on overall development | 25 (81) |
| Sig. effect of sensory function on comm. development   | 24 (77) |
| <b><u>HEARING</u></b>                                  |         |
| Sig. effect of hearing on comm. development            | 24 (77) |
| <b><u>Type of Hearing Loss</u></b>                     |         |
| Conductive loss                                        | 0 (0)   |
| Sensorineural loss                                     | 11 (35) |
| Mixed loss                                             | 18 (58) |
| <b><u>Degree of Hearing Loss</u></b>                   |         |
| Normal hearing                                         | 2 (6)   |
| Mild loss                                              | 9 (29)  |
| Moderate loss                                          | 5 (16)  |
| Moderately-Severe loss                                 | 6 (19)  |
| Severe-Profound loss                                   | 9 (29)  |
| <b><u>Treatment of Hearing Loss</u></b>                |         |
| Uses hearing aids                                      | 17 (55) |
| Uses cochlear implant                                  | 1 (3)   |
| No treatment                                           | 13 (42) |

**Table 1: continued**

| TOTAL (n=31)<br># of Participants (%)                      |          |
|------------------------------------------------------------|----------|
| <b><u>BALANCE</u></b>                                      |          |
| Sig. effect of balance on<br>comm. development             | 13 (42)  |
| <b><u>Mobility</u></b>                                     |          |
| Walks independently                                        | 19 (61)  |
| Mobile with assistance                                     | 7 (23)   |
| Does not walk (uses wheelchair)                            | 5 (16)   |
| <b><u>VISION</u></b>                                       |          |
| Sig. effect of vision on<br>comm. development              | 13 (42)  |
| Prevalence of vision problems                              | 31 (100) |
| <b><u>SMELL</u></b>                                        |          |
| Sig. effect of smell on<br>comm. development               | 3 (10)   |
| <b><u>TASTE</u></b>                                        |          |
| Sig. effect of taste on<br>comm. development               | 4 (13)   |
| <b><u>BEHAVIOR</u></b>                                     |          |
| Sig. effect of behavior on<br>overall development          | 9 (29)   |
| Sig. effect of behavior on<br>comm. development            | 9 (29)   |
| Declined to respond                                        | 7 (23)   |
| <b><u>Behaviors</u></b>                                    |          |
| Well-adjusted                                              | 16 (52)  |
| Social or emotional immaturity                             | 26 (84)  |
| Unstable or explosive behavior                             | 10 (32)  |
| Problems with attention                                    | 21 (68)  |
| Overreacts to certain situations                           | 13 (42)  |
| Aggressive or defensive behaviors                          | 14 (45)  |
| Rhythmic rocking                                           | 5 (16)   |
| Self-stimulatory behavior                                  | 19 (61)  |
| Sensitivity to loud sounds                                 | 15 (48)  |
| Lack of communication for<br>interpersonal purposes        | 14 (45)  |
| <b><u>COMMUNICATION TRAINING</u></b>                       |          |
| Sig. effect of speech & language<br>on overall development | 29 (93)  |
| <b><u>CURRENT EDUCATIONAL<br/>PLACEMENT</u></b>            |          |
| Sig. effect of communication on<br>educational development | 26 (84)  |
| <b><u>Educational Setting</u></b>                          |          |
| Mainstream                                                 | 4 (13)   |
| Inclusive                                                  | 7 (23)   |
| Self-contained (hearing impaired)                          | 5 (16)   |
| Self-contained (multiple disabilities)                     | 3 (10)   |
| Deaf school                                                | 2 (6)    |
| Early childhood program                                    | 2 (6)    |
| Home school                                                | 5 (16)   |
| Not currently enrolled                                     | 3 (10)   |

### **Primary Mode of Communication**

The section titled “Communication” was re-named “Primary Mode of Communication” to distinguish the type of communication system that was most often used by the child. This section was not placed first on the questionnaire but was placed first in the data tables, as it was central to the purposes of this study. The primary methods were divided during data analysis into symbolic communication (speech and sign language) and no symbolic communication (gesture and perlocutionary-type communication). The perlocutionary-type of communication was not listed specifically on the questionnaire but was described by parents on both the questionnaire and during the interview. The perlocutionary term was chosen as it describes the primitive-like communication used by the children in this category. Such communication behaviors include crying, laughing, and babbling and are typically used by normally-developing infants, aged 0 to 8 months (Paul, 1995).

The other type of communication method that was described by parents on the questionnaire but was not included in data analysis was the use of one’s own signs. This method was not included in the analysis because it was not reported as the child’s primary means of communication. However, a small group of children were reported as using their own signs as a means of secondary communication.

A majority of parents (74%) reported that the development of communication had a significant effect on overall development. Most parents (87%) also reported that CHARGE Syndrome had a significant effect on the development of communication. More than half (58%) of the participants in the present study developed symbolic language, while the other participants (42%) did not use symbolic communication.

## **Physical Health**

A majority of parents rated physical health as having a significant effect on overall development (81%) and on the development of communication (77%). Parents reported many physical conditions, of those that were the most prevalent included choanal atresia, heart problems, swallowing and/or feeding problems, facial problems, hypotonia, growth deficiency, and the use of a gastrostomy tube. Other reported conditions included cleft lip and/or palate, use of a tracheostomy tube, structural neurologic problems (agenesis of corpus callosum, holoprosencephaly, and hydrocephaly), and other neurologic problems (seizures, apraxia, and autism).

## **Sensory Function**

Most parents reported that sensory function had a significant effect on overall development (81%) and on communication development (77%). Sensory function was evaluated by judgment and informational questions for each sense – hearing, vision, smell, taste, and balance.

**Hearing.** The parents reported that 93% of the participants had hearing loss. The parents also reported that 77% had hearing loss that had a significant effect on communication development. Most participants (58%) were reported as having a mixed hearing loss, while 35% had a sensorineural hearing loss and none had a purely conductive hearing loss. All degrees of hearing and hearing loss were reported, including normal hearing, mild hearing loss, moderate hearing loss, moderately-severe hearing loss, and severe-profound hearing loss. Most of the participants used amplification: 17 (55%) wore hearing aids and one participant (3%) had a cochlear implant.

**Balance.** A significant percentage of the participants (42%) were reported to have balance problems that affected the development of communication. In a separate question, the parents were asked to classify the participants into mobility categories: (1) walks independently, (2) mobile with assistance (crawls or uses a walker), and (3) does not walk (uses a wheelchair or stroller). A majority of participants (61%) were able to walk independently, while 23% were mobile with assistance and 16% did not walk.

**Vision.** The parents reported that 100% of the participants had vision problems, but they also reported that only 42% had vision problems that had a significant effect on communication development. The implication is that many of the vision problems had minimal effects on communication development.

**Smell and Taste.** Most of the parents did not consider smell or taste to have a significant effect on communication development. Only 10% of the parents rated smell to have a significant effect on communication, while 13% rated taste to have a significant effect.

### **Behavior**

The parental responses to questions on behavior were not anticipated. Only 29% of parents reported behavior to have a significant effect on overall development and communication development. Surprisingly, 23% of parents initially declined to respond to the judgment questions. During the parent interviews it was found that every parent believed behavior was an outcome of communication ability rather than a determinant. Each parent felt that limited or poor communication skills may have contributed to the types of inappropriate behavior that were included in the questionnaire. Furthermore,

many parents corrected the investigator by indicating that the behaviors should not be labeled as “poor” because they are a reflection of frustration. Since the parents had such strong views on the reasons for inappropriate behavior, it would have been difficult to obtain information from the parents to indicate that communication development was, in fact, negatively affected by poor behavior. For that reason inappropriate behavior was considered as a consequence rather than a determinant of poor communication abilities. Half of the parents (52%) reported that their child was well-adjusted. However, the common types of behaviors that were reported included social or emotional immaturity (84%), problems with attention (71%), and self-stimulatory behavior (61%).

### **Communication Training**

In Table 1, the category “Communication Training” was originally labeled “Speech and Language” on the questionnaire. The title was changed to include categories for total communication (combination of speech-language therapy and sign language instruction), speech language therapy, sign-language instruction, and no training. (A detailed analysis of types of communication training is presented in a later section of the RESULTS.) Almost all of the parents (93%) reported that speech and language had a significant effect on overall development and the development of communication.

### **Education**

Most of the parents (84%) considered communication abilities to have a significant effect on educational development. In the questionnaire, a parent judgment

question was asked but it was found during parent interview that the question was phrased incorrectly. The question asked for a judgment on the degree to which education affected the child's overall educational development. Since this question was worded poorly, it was eliminated from analysis. The types of reported educational placements included eight different settings, ranging from a mainstream environment to no current school enrollment.

## **RESULTS FOR SYMBOLIC AND NO SYMBOLIC LANGUAGE GROUPS**

Based on the information provided by the parents, the participants in the present study were divided into two communication groups: participants who used symbolic language (n=18) and those that did not use symbolic language (n=13). The characteristics of these two groups were compared on each of the questions on the questionnaire in Table 2 using the same format as Table 1. Statistical analyses were performed on data pairs using 2X2 chi square analyses. Formal tests were conducted on the six data pairs that had the greatest chance to be significantly different. The results for two of these comparisons were significant and the other four were not. As a result, all other data pairs were considered to be not significantly different based on visual inspection.

### **Primary Mode of Communication**

Parents of children in the symbolic communication group reported that communication had a significant effect on overall development (67%) and that CHARGE had a significant effect on the development of communication (94%). Similarly, the no symbolic communication group reported that communication had a significant effect on

**Table 2: Symbolic language group and No Symbolic language group**

|                                                        | <b>Symbolic (n=18)<br/># of Participants (%)</b> | <b>No Symbolic (n=13)<br/># of Participants (%)</b> |
|--------------------------------------------------------|--------------------------------------------------|-----------------------------------------------------|
| <b><u>PRIMARY MODE</u></b>                             |                                                  |                                                     |
| <b><u>OF COMMUNICATION</u></b>                         |                                                  |                                                     |
| Sig. effect of communication on overall development    | 12 (67)                                          | 11 (85)                                             |
| Sig. effect of CHARGE on comm. development             | 17 (94)                                          | 10 (77)                                             |
| <b><u>Communication Mode</u></b>                       |                                                  |                                                     |
| Speech                                                 | 11 (61)                                          | 0 (0)                                               |
| Sign Language                                          | 6 (33)                                           | 0 (0)                                               |
| Gesture                                                | 0 (0)                                            | 11 (85)                                             |
| Perlocutionary                                         | 0 (0)                                            | 2 (15)                                              |
| <b><u>PHYSICAL HEALTH</u></b>                          |                                                  |                                                     |
| Sig. effect of physical health on overall development  | 15 (83)                                          | 10 (77)                                             |
| Sig. effect of physical health on comm. development    | 14 (78)                                          | 10 (77)                                             |
| <b><u>Physical Conditions</u></b>                      |                                                  |                                                     |
| Choanal atresia                                        | 13 (72)                                          | 10 (77)                                             |
| Heart problems                                         | 14 (78)                                          | 9 (69)                                              |
| Cleft lip/palate                                       | 6 (33)                                           | 4 (31)                                              |
| Swallowing/feeding problems                            | 16 (89)                                          | 11 (85)                                             |
| Facial problems                                        | 11 (61)                                          | 6 (46)                                              |
| Hypotonia                                              | 12 (67)                                          | 13 (100)                                            |
| Growth deficiency                                      | 7 (39)                                           | 10 (77)                                             |
| Tracheostomy                                           | 1 (5)                                            | 1 (8)                                               |
| Gastrostomy tube                                       | 11 (61)                                          | 7 (54)                                              |
| Neurologic problems (structural)                       | 2 (11)                                           | 3 (23)                                              |
| Neurologic problems (other)                            | 1 (5)                                            | 4 (31)                                              |
| <b><u>SENSORY FUNCTION</u></b>                         |                                                  |                                                     |
| Sig. effect of sensory function on overall development | 15 (83)                                          | 10 (77)                                             |
| Sig. effect of sensory function on comm. development   | 14 (78)                                          | 10 (77)                                             |
| <b><u>HEARING</u></b>                                  |                                                  |                                                     |
| Sig. effect of hearing on comm. development            | 14 (78)                                          | 10 (77)                                             |
| <b><u>Type of Hearing Loss</u></b>                     |                                                  |                                                     |
| Conductive loss                                        | 0 (0)                                            | 0 (0)                                               |
| Sensorineural loss                                     | 5 (28)                                           | 6 (46)                                              |
| Mixed loss                                             | 12 (67)                                          | 6 (46)                                              |
| <b><u>Degree of Hearing Loss</u></b>                   |                                                  |                                                     |
| Normal hearing                                         | 1 (5)                                            | 1 (8)                                               |
| Mild loss                                              | 6 (33)                                           | 3 (23)                                              |
| Moderate loss                                          | 1 (5)                                            | 4 (31)                                              |
| Moderately-severe loss                                 | 4 (22)                                           | 2 (15)                                              |
| Severe-profound loss                                   | 6 (33)                                           | 3 (23)                                              |
| <b><u>Treatment of Hearing Loss</u></b>                |                                                  |                                                     |
| Uses hearing aids                                      | 12 (67)                                          | 5 (38)                                              |
| Uses cochlear implant                                  | 1 (5)                                            | 0 (0)                                               |
| No treatment                                           | 5 (28)                                           | 8 (61)                                              |

**Table 2: continued**

|                                                            | <b>Symbolic (n=18)<br/># of Participants (%)</b> | <b>No Symbolic (n=13)<br/># of Participants (%)</b> |
|------------------------------------------------------------|--------------------------------------------------|-----------------------------------------------------|
| <b><u>BALANCE</u></b>                                      |                                                  |                                                     |
| Sig. effect of balance on<br>comm. development             | 8 (44)                                           | 5 (38)                                              |
| <b><u>Mobility</u></b>                                     |                                                  |                                                     |
| Walks independently                                        | 16 (89)                                          | 3 (23)                                              |
| Mobile with assistance                                     | 2 (11)                                           | 5 (38)                                              |
| Does not walk (uses wheelchair)                            | 0 (0)                                            | 5 (38)                                              |
| <b><u>VISION</u></b>                                       |                                                  |                                                     |
| Sig. effect of vision on<br>comm. development              | 8 (44)                                           | 5 (38)                                              |
| Prevalence of vision problems                              | 18 (100)                                         | 13 (100)                                            |
| <b><u>SMELL</u></b>                                        |                                                  |                                                     |
| Sig. effect of smell on<br>comm. development               | 1 (5)                                            | 1 (8)                                               |
| <b><u>TASTE</u></b>                                        |                                                  |                                                     |
| Sig. effect of taste on<br>comm. development               | 1 (5)                                            | 2 (15)                                              |
| <b><u>BEHAVIOR</u></b>                                     |                                                  |                                                     |
| Sig. effect of behavior on<br>overall development          | 6 (33)                                           | 3 (23)                                              |
| Sig. effect of behavior on<br>comm. development            | 5 (27)                                           | 4 (31)                                              |
| Declined to respond                                        | 5 (27)                                           | 2 (15)                                              |
| <b><u>Behaviors</u></b>                                    |                                                  |                                                     |
| Well-adjusted                                              | 7 (39)                                           | 9 (69)                                              |
| Social or emotional immaturity                             | 15 (83)                                          | 11 (85)                                             |
| Unstable or explosive behavior                             | 4 (22)                                           | 6 (46)                                              |
| Problems with attention                                    | 12 (67)                                          | 10 (77)                                             |
| Overreacts to certain situations                           | 7 (39)                                           | 6 (46)                                              |
| Aggressive or defensive behaviors                          | 9 (50)                                           | 5 (38)                                              |
| Rhythmic rocking                                           | 0 (0)                                            | 5 (38)                                              |
| Self-stimulatory behavior                                  | 9 (50)                                           | 10 (77)                                             |
| Sensitivity to loud sounds                                 | 8 (44)                                           | 7 (54)                                              |
| Lack of communication for<br>interpersonal purposes        | 6 (33)                                           | 9 (69)                                              |
| <b><u>COMMUNICATION TRAINING</u></b>                       |                                                  |                                                     |
| Sig. effect of speech & language<br>on overall development | 15 (83)                                          | 13 (100)                                            |
| <b><u>CURRENT EDUCATIONAL<br/>PLACEMENT</u></b>            |                                                  |                                                     |
| Sig. effect of communication on<br>educational development | 14 (78)                                          | 12 (92)                                             |
| <b><u>Educational Setting</u></b>                          |                                                  |                                                     |
| Mainstream                                                 | 4 (22)                                           | 0 (0)                                               |
| Inclusive                                                  | 5 (28)                                           | 2 (15)                                              |
| Self-contained (hearing impaired)                          | 2 (11)                                           | 3 (23)                                              |
| Self-contained (multiple disabilities)                     | 1 (5)                                            | 2 (15)                                              |
| Deaf school                                                | 2 (11)                                           | 0 (0)                                               |
| Early childhood program                                    | 0 (0)                                            | 2 (15)                                              |
| Home school                                                | 2 (11)                                           | 3 (23)                                              |
| Not currently enrolled                                     | 2 (11)                                           | 1 (8)                                               |

overall development (85%) and that CHARGE had a significant effect on the development of communication (77%). Of the 18 participants with symbolic communication, 11 (61%) used speech and 6 (33%) used sign language. In the no symbolic group, 11 (85%) used gestures, while 2 (15%) used the perlocutionary-type of communication.

There was one participant who required special consideration. Participant A20 was a one-year-old female with a tracheotomy and Passy-Muir valve that permits speech. This infant has developed some expressive language (speech sounds that were age appropriate) and has demonstrated good receptive language skills (able to maintain communicative interactions and to provide appropriate responses to simple requests). This participant was included in the symbolic communication group but was not classified as being in the speech group or sign language group.

### **Physical Health**

A majority of the parents in both the symbolic and no symbolic groups reported that physical health had a significant effect on overall development (83% and 77%, respectively) and on communication development (78% and 77%). For most of the conditions included in the questionnaire, the frequency of involvement was not significantly different for the two groups in any of the conditions related to the effects of physical health on communication. However, there were a few conditions in which there were differences between the two groups. While the majority of participants had hypotonia, the no symbolic group was more affected (100%) than the symbolic group (67%). On the basis of these data, it is unclear if the difference between the two groups is

significant, but since the majority of participants in each group had hypotonia, it is not a differentiator between groups. Growth deficiency was also more common in the no symbolic group. The use of a gastrostomy tube, however, was more prevalent in the symbolic group, therefore showing that its use did not prevent the acquisition of symbolic language.

### **Sensory Function**

Sensory function was recorded as having a significant effect on overall development (symbolic group: 83% and no symbolic group: 77%) and on the development of communication (78% and 77%) in both groups. Preliminary analysis revealed that the two groups did not differ on the percentage of involvement for vision, smell, or taste, however, there was the possibility that the groups differed on the bases of hearing and balance.

**Hearing.** A majority of the parents in the symbolic group and no symbolic group (77% and 78%) reported hearing to have a significant effect on the development of communication. Though the percentages of participants in each groups who had hearing loss was similar, it was hypothesized that the degree of hearing loss may have been greater in the no symbolic group. A Mann-Whitney test was performed to test this hypothesis using the frequency of occurrence data for the five different degrees of hearing loss for the two groups. The results revealed that the two groups did not differ in degree of hearing loss ( $p=.704$ ).

A second consideration was that there may have been less use of amplification in the no symbolic group. This hypothesis was tested using a 2X2 chi square test comparing

use and non-use of amplification for the two groups. The difference between these groups was not significant ( $p=.079$ ). These results suggest that the symbolic and no symbolic groups did not differ with respect to use of amplification.

**Balance.** For both the symbolic and no symbolic groups, the parents reported that a substantial percentage of the participants had balance problems that had a significant effect on the development of communication. However, the percentage that had balance problems that affected communication in each group was similar (44% for the symbolic group and 38% for the no symbolic group). As a result, these two groups were not differentiated on the basis of balance problems.

In the section on balance, questions were asked specifically on mobility. The results indicate that the symbolic and no symbolic groups differ significantly on the basis of mobility. Of the 18 participants in the symbolic group, 89% walked independently, 11% were mobile with assistance, and there were none who did not walk. Of the 13 participants in the no symbolic group, 23% walked independently, while 38% were mobile with assistance and 38% did not walk. A 2X2 chi square test was used to evaluate the difference between groups of those participants who walked independently and those participants who did not walk or who required assistance. The difference between the two groups was significant ( $p=.00045$ ). This result suggests that there is a significant relation between the ability to walk independently and the development of symbolic communication.

## **Behavior**

In the parent interviews, the parents indicated that behavior was an outgrowth of the inability to communicate. Based on this report, it was expected that participants in the no symbolic group would have more behavior problems than those who had symbolic communication. However, similar results for each type of behavior were reported for both the symbolic communication group and the no symbolic communication group. These findings provide some evidence to suggest that inappropriate behavior may not be related to poor communication ability.

## **Communication Training**

Information reported by the parents on the questionnaire and during the interview revealed the importance of the type of communication training received and the ages at which the training was received. Table 3 displays the data for each participant for the type of language training that was received and the age at which language intervention was initiated. The table also shows the duration of treatment for each participant. There are a number of observations that can be made about the data in Table 3:

- **Symbolic: Speech Group**  
In this group, language training began in the first or second year of life and was sustained. Of the twelve participants in the group, eleven received total communication training for some period of time.
- **Symbolic: Sign Language Group**  
For five of the six participants in this group, communication training began in the first or second year of life and was sustained. All of these participants also received total communication training for some period of time.
- **No Symbolic: Gesture Group**  
Of the eleven participants in this group, five began receiving communication training in the first or second year of life but symbolic language did not develop,

**Table 3: Ages at which participants received communication training:** blank space=no intervention; SP=speech-language therapy; SI=sign language instruction; TC=total communication training (SP and SI together); shaded areas=years beyond the participant's current age; \* =American Sign Language, <sup>†</sup>=Signed English, 'Langue Signe Quebec.

| Participant Number          | Participant Age (Years) |                  |                  |                  |                  |                  |                  |                  |                  |
|-----------------------------|-------------------------|------------------|------------------|------------------|------------------|------------------|------------------|------------------|------------------|
|                             | 0                       | 1                | 2                | 3                | 4                | 5                | 6                | 7                | >7               |
| <b>Symbolic:</b>            |                         |                  |                  |                  |                  |                  |                  |                  |                  |
| <b>Speech Group</b>         |                         |                  |                  |                  |                  |                  |                  |                  |                  |
| A12 ( 4 F)                  |                         | TC*              | TC*              | SP               | SP               |                  |                  |                  |                  |
| A13 (11 F)                  | TC <sup>†</sup>         | TC <sup>†</sup>  | TC <sup>†</sup>  | TC <sup>†</sup>  | TC <sup>†</sup>  | TC <sup>†</sup>  | TC <sup>†</sup>  | TC <sup>†</sup>  | TC <sup>†</sup>  |
| A14 ( 7 F)                  | SP                      | SP               | SP               | SP               | SP               | SP               | SP               | SP               |                  |
| A15 ( 7 F)                  | SP                      | TC*              | TC*              | SP               | SP               | SP               | SP               | SP               |                  |
| A16 (10M)                   | TC*                     | TC*              | TC*              | TC*              | SP               | SP               | SP               | SP               | SP               |
| A17 ( 8 F)                  | TC*                     | TC*              | TC*              | TC*              | TC*              | SP               | SP               | SP               | SP               |
| A18 ( 5 F)                  | TC*                     | TC*              | TC*              | TC*              | TC*              | TC*              |                  |                  |                  |
| A19 ( 7M)                   | SP                      | TC*              | TC*              | SP               | SP               | SP               | SP               | SP               |                  |
| C1 ( 4M)                    |                         |                  | TC               | TC               | TC               |                  |                  |                  |                  |
| C2 (12 F)                   | SP                      | SP               | TC*              | TC*              | TC*              | TC*              | SP               | SP               | SP               |
| C3 ( 4 F)                   | SI                      | TC*              | TC*              | TC*              | TC*              |                  |                  |                  |                  |
| A20 ( 1 F)**                | TC*                     | TC*              |                  |                  |                  |                  |                  |                  |                  |
| <b>Symbolic:</b>            |                         |                  |                  |                  |                  |                  |                  |                  |                  |
| <b>Sign Language Group</b>  |                         |                  |                  |                  |                  |                  |                  |                  |                  |
| A21 (27 F)                  |                         |                  |                  | TC*              | TC*              | SI               | SI               | SI               | SI               |
| A22 (11M)                   |                         |                  | SI               | TC*              | TC*              | TC*              | TC*              | SI               | SI               |
| A23 ( 9 F)                  | TC* <sup>†</sup>        | TC* <sup>†</sup> | TC* <sup>†</sup> | TC* <sup>†</sup> | TC* <sup>†</sup> | TC* <sup>†</sup> | TC* <sup>†</sup> | TC* <sup>†</sup> | TC* <sup>†</sup> |
| A24 ( 8 M)                  | SI*                     | SI*              | SI*              | SI*              | SI*              | SI*              | SI*              | SI*              | SI*              |
| A25 ( 6 F)                  |                         | SI*              | SI*              | TC*              | TC*              | TC*              | TC*              |                  |                  |
| C4 ( 5 M)                   |                         | SP               | SP               | SP               | SP               | TC*              |                  |                  |                  |
| <b>No Symbolic:</b>         |                         |                  |                  |                  |                  |                  |                  |                  |                  |
| <b>Gesture Group</b>        |                         |                  |                  |                  |                  |                  |                  |                  |                  |
| A1 ( 8 F)                   |                         |                  |                  |                  | SP               | SP               | SP               | TC*              | TC*              |
| A2 ( 8 F)                   |                         |                  |                  |                  | SP               | SP               | SP               | SP               | SP               |
| A3 ( 4 F)                   |                         |                  | SI*              | TC*              | TC*              |                  |                  |                  |                  |
| A4 ( 4M)                    | TC?                     | TC?              | TC?              | TC?              | TC?              |                  |                  |                  |                  |
| A5 ( 3 F)                   |                         | SP               | TC?              | TC?              |                  |                  |                  |                  |                  |
| A6 ( 2M)                    |                         | TC*              | TC*              |                  |                  |                  |                  |                  |                  |
| A7 ( 9M)                    |                         |                  |                  |                  |                  | SP               | TC*              | TC*              | TC*              |
| A8 ( 3 F)                   |                         |                  |                  |                  |                  |                  |                  |                  |                  |
| A9 (11M)                    |                         | SP               | SP               | SP               | SP               | SP               | SP               | SP               | SP               |
| C5 (11 F)                   |                         |                  |                  |                  | SP               | SP               | SP               | SP               | SP               |
| C6 ( 7 M)                   | SP                      | SP               | SP               | SP               | TC*              | TC*              | TC*              | TC*              |                  |
| <b>No Symbolic:</b>         |                         |                  |                  |                  |                  |                  |                  |                  |                  |
| <b>Perlocutionary Group</b> |                         |                  |                  |                  |                  |                  |                  |                  |                  |
| A10 ( 5 M)                  |                         | SP               | SP               | SP               | SP               | SP               |                  |                  |                  |
| A11 ( 2 M)                  | SI*                     | SI*              | SI*              |                  |                  |                  |                  |                  |                  |

regardless of the type of training that was implemented. The other six participants began receiving intervention after the second year of life or not at all and also did not develop symbolic language.

- No Symbolic: Perlocutionary Group

For these two participants, intervention began early and was sustained. However, each participant (as will be discussed later) had neurologic conditions that were probably sufficiently disabling, that they were unable to take advantage of the communication training to develop symbolic language.

The hypothesis was made that there was a relationship between the development of symbolic language and communication training in the first five years of life. Those participants who received any total communication training in the first five years of life were compared to those who did not using a 2X2 chi square test. The results of this analysis indicate that there is a significant relationship between the development of symbolic language and intervention with total communication training in the first five years of life ( $p=.006$ ).

## **RESULTS FOR THE SPEECH AND SIGN LANGUAGE GROUPS**

In Table 2, factors were established as possible determinants in the development of symbolic language. In Table 4, the participants in the symbolic group were further divided into speech ( $n=11$ ) or sign language ( $n=6$ ) groups to determine if there were factors that were associated with the development of these modes of communication. Comparisons were made on all of the questions on the questionnaire. There were only a few comparisons of interest between these two groups.

**Table 4: Speech group and Sign Language group**

|                                                        | Speech (n=11)<br># of Participants (%) | Sign Language (n=6)<br># of Participants (%) |
|--------------------------------------------------------|----------------------------------------|----------------------------------------------|
| <b><u>PRIMARY MODE OF COMMUNICATION</u></b>            |                                        |                                              |
| Sig. effect of communication on overall development    | 6 (55)                                 | 5 (83)                                       |
| Sig. effect of CHARGE on comm. development             | 10 (91)                                | 6 (100)                                      |
| <b><u>PHYSICAL HEALTH</u></b>                          |                                        |                                              |
| Sig. effect of physical health on overall development  | 8 (73)                                 | 6 (100)                                      |
| Sig. effect of physical health on comm. development    | 7 (64)                                 | 6 (100)                                      |
| <b><u>Physical Conditions</u></b>                      |                                        |                                              |
| Choanal atresia                                        | 9 (82)                                 | 3 (50)                                       |
| Heart problems                                         | 8 (73)                                 | 5 (83)                                       |
| Cleft lip/palate                                       | 3 (27)                                 | 3 (50)                                       |
| Swallowing/feeding problems                            | 10 (91)                                | 5 (83)                                       |
| Facial problems                                        | 5 (45)                                 | 6 (100)                                      |
| Hypotonia                                              | 7 (64)                                 | 5 (83)                                       |
| Growth deficiency                                      | 3 (27)                                 | 4 (67)                                       |
| Tracheostomy                                           | 0 (0)                                  | 0 (0)                                        |
| Gastrostomy tube                                       | 7 (64)                                 | 3 (50)                                       |
| Neurologic problems (structural)                       | 1 (9)                                  | 1 (17)                                       |
| Neurologic problems (other)                            | 0 (0)                                  | 1 (17)                                       |
| <b><u>SENSORY FUNCTION</u></b>                         |                                        |                                              |
| Sig. effect of sensory function on overall development | 9 (82)                                 | 6 (100)                                      |
| Sig. effect of sensory function on comm. development   | 9 (82)                                 | 5 (83)                                       |
| <b><u>HEARING</u></b>                                  |                                        |                                              |
| Sig. effect of hearing on comm. development            | 9 (82)                                 | 5 (83)                                       |
| <b><u>Type of Hearing Loss</u></b>                     |                                        |                                              |
| Conductive loss                                        | 0 (0)                                  | 0 (0)                                        |
| Sensorineural loss                                     | 4 (36)                                 | 1 (17)                                       |
| Mixed loss                                             | 7 (64)                                 | 5 (83)                                       |
| <b><u>Degree of Hearing Loss</u></b>                   |                                        |                                              |
| Normal hearing                                         | 0 (0)                                  | 0 (0)                                        |
| Mild loss                                              | 5 (45)                                 | 1 (17)                                       |
| Moderate loss                                          | 1 (9)                                  | 0 (0)                                        |
| Moderately-severe loss                                 | 2 (18)                                 | 2 (33)                                       |
| Severe-profound loss                                   | 3 (27)                                 | 3 (50)                                       |
| <b><u>Treatment of Hearing Loss</u></b>                |                                        |                                              |
| Uses hearing aids                                      | 9 (82)                                 | 3 (50)                                       |
| Uses cochlear implant                                  | 1 (9)                                  | 0 (0)                                        |
| No treatment                                           | 1 (9)                                  | 3 (50)                                       |

**Table 4: continued**

|                                                            | <b>Speech (n=11)<br/># of Participants (%)</b> | <b>Sign Language (n=6)<br/># of Participants (%)</b> |
|------------------------------------------------------------|------------------------------------------------|------------------------------------------------------|
| <b><u>BALANCE</u></b>                                      |                                                |                                                      |
| Sig. effect of balance on<br>comm. development             | 4 (36)                                         | 4 (67)                                               |
| <b><u>Mobility</u></b>                                     |                                                |                                                      |
| Walks independently                                        | 10 (91)                                        | 6 (100)                                              |
| Mobile with assistance                                     | 1 (9)                                          | 0 (0)                                                |
| Does not walk (uses wheelchair)                            | 0 (0)                                          | 0 (0)                                                |
| <b><u>VISION</u></b>                                       |                                                |                                                      |
| Sig. effect of vision on<br>Comm. development              | 3 (27)                                         | 5 (83)                                               |
| Prevalence of vision problems                              | 11 (100)                                       | 6 (100)                                              |
| <b><u>SMELL</u></b>                                        |                                                |                                                      |
| Sig. effect of smell on<br>comm. development               | 0 (0)                                          | 1 (17)                                               |
| <b><u>TASTE</u></b>                                        |                                                |                                                      |
| Sig. effect of taste on<br>comm. development               | 0 (0)                                          | 1 (17)                                               |
| <b><u>BEHAVIOR</u></b>                                     |                                                |                                                      |
| Sig. effect of behavior on<br>overall development          | 4 (36)                                         | 2 (33)                                               |
| Sig. effect of behavior on<br>comm. development            | 2 (18)                                         | 3 (50)                                               |
| Declined to respond                                        | 2 (18)                                         | 3 (50)                                               |
| <b><u>Behaviors</u></b>                                    |                                                |                                                      |
| Well-adjusted                                              | 4 (36)                                         | 3 (50)                                               |
| Social or emotional immaturity                             | 10 (91)                                        | 5 (83)                                               |
| Unstable or explosive behavior                             | 3 (27)                                         | 1 (17)                                               |
| Problems with attention                                    | 8 (73)                                         | 4 (67)                                               |
| Overreacts to certain situations                           | 4 (36)                                         | 3 (50)                                               |
| Aggressive or defensive behaviors                          | 5 (45)                                         | 4 (67)                                               |
| Rhythmic rocking                                           | 0 (0)                                          | 0 (0)                                                |
| Self-stimulatory behavior                                  | 6 (55)                                         | 3 (50)                                               |
| Sensitivity to loud sounds                                 | 6 (55)                                         | 2 (33)                                               |
| Lack of communication for<br>interpersonal purposes        | 4 (36)                                         | 2 (33)                                               |
| <b><u>COMMUNICATION TRAINING</u></b>                       |                                                |                                                      |
| Sig. effect of speech & language<br>on overall development | 9 (82)                                         | 6 (100)                                              |
| <b><u>CURRENT EDUCATIONAL<br/>PLACEMENT</u></b>            |                                                |                                                      |
| Sig. effect of communication on<br>educational development | 7 (64)                                         | 6 (100)                                              |
| <b><u>Educational Setting</u></b>                          |                                                |                                                      |
| Mainstream                                                 | 4 (36)                                         | 0 (0)                                                |
| Inclusive                                                  | 4 (36)                                         | 1 (17)                                               |
| Self-contained (hearing impaired)                          | 0 (0)                                          | 2 (33)                                               |
| Self-contained (multiple disabilities)                     | 1 (9)                                          | 0 (0)                                                |
| Deaf school                                                | 1 (9)                                          | 1 (17)                                               |
| Early childhood program                                    | 0 (0)                                          | 0 (0)                                                |
| Home school                                                | 1 (9)                                          | 1 (17)                                               |
| Not currently enrolled                                     | 0 (0)                                          | 1 (17)                                               |

## **Physical Health**

The frequency of occurrence of many of the physical disorders that were on the questionnaire was similar for participants who used speech and those who used sign language. A difference was noted in the occurrence of facial problems, which were present in all (100%) of the participants in the sign language group but in 45% in the speech group.

## **Hearing Loss and Treatment of Hearing Loss**

One hypothesis at the outset of this study was that the participants who used sign language would have worse hearing than those participants who spoke. A Mann-Whitney test was performed to compare the five different degrees of hearing loss between the two groups. Results of the test were not significant ( $p=.204$ ) suggesting that degree of hearing loss was not a primary factor in determining whether speech or sign language was the primary mode of communication.

Another consideration in distinguishing the two groups was use of amplification. Of the eleven participants who were in the speech group, nine wore hearing aids, one used a cochlear implant, and one did not use any type of amplification because his hearing loss was in the mild range. Thus, all were receiving adequate auditory input to develop spoken language. In the sign language group, three wore hearing aids, none used a cochlear implant, and three did not use any amplification. One of the three participants who did not use amplification (Participant A21) was a twenty-seven year-old female who had been diagnosed with apraxia. This participant had a mild hearing loss and did not use amplification due to good functional hearing. Sign language was the mode of

communication used by the participant because of oral-motor problems associated with apraxia. The other two participants who did not use amplification had previously tried to use amplification but had rejected wearing hearing aids. One of the participants in the sign language group (Participant A22) who did use amplification was not aided for speech purposes but was instead amplified to hear environmental noises. The other two participants in the sign language group who used amplification had an unaided moderately-severe hearing loss.

Based on this analysis, it was then hypothesized that it was not use of amplification that distinguished the two groups but was instead effectiveness of the amplification used. The children that were able to be amplified adequately and who accepted wearing amplification were able to develop speech, while the others used sign language. In the sign language group, the results suggest that two of the six participants appeared to be amplified appropriately and may possibly transition to speech, but the other four participants were not receiving sufficient benefit from amplification.

## **RESULTS FOR THE PERLOCUTIONARY GROUP**

This group contained only two participants. The data for these participants is shown in Table 5. One participant (A10) was reported to have significant structural neurologic damage (holoprosencephaly) and severe growth deficiency (age: 5 years; weight: 17 lbs.). The other participant (A11) was reported to have been missing portions of his brain and did not have vocal folds. These severe conditions alone were probably the cause of the failure to develop even a gesture system for means of communication.

**Table 5: Gesture group and Perlocutionary group**

|                                                        | <b>Gesture (n=11)<br/># of Participants (%)</b> | <b>Perlocutionary (n=2)<br/># of Participants (%)</b> |
|--------------------------------------------------------|-------------------------------------------------|-------------------------------------------------------|
| <b><u>PRIMARY MODE<br/>OF COMMUNICATION</u></b>        |                                                 |                                                       |
| Sig. effect of communication on overall development    | 10 (91)                                         | 1 (50)                                                |
| Sig. effect of CHARGE on comm. development             | 9 (82)                                          | 1 (50)                                                |
| <b><u>PHYSICAL HEALTH</u></b>                          |                                                 |                                                       |
| Sig. effect of physical health on overall development  | 8 (73)                                          | 2 (100)                                               |
| Sig. effect of physical health on comm. development    | 8 (73)                                          | 2 (100)                                               |
| <b><u>Physical Conditions</u></b>                      |                                                 |                                                       |
| Choanal atresia                                        | 8 (73)                                          | 2 (100)                                               |
| Heart problems                                         | 7 (64)                                          | 2 (100)                                               |
| Cleft lip/palate                                       | 2 (18)                                          | 2 (100)                                               |
| Swallowing/feeding problems                            | 9 (82)                                          | 2 (100)                                               |
| Facial problems                                        | 5 (45)                                          | 1 (50)                                                |
| Hypotonia                                              | 11 (100)                                        | 2 (100)                                               |
| Growth deficiency                                      | 9 (82)                                          | 1 (50)                                                |
| Tracheostomy                                           | 0 (0)                                           | 1 (50)                                                |
| Gastrostomy tube                                       | 6 (55)                                          | 1 (50)                                                |
| Neurologic problems (structural)                       | 1 (9)                                           | 2 (100)                                               |
| Neurologic problems (other)                            | 4 (36)                                          | 0 (0)                                                 |
| <b><u>SENSORY FUNCTION</u></b>                         |                                                 |                                                       |
| Sig. effect of sensory function on overall development | 8 (73)                                          | 2 (100)                                               |
| Sig. effect of sensory function on comm. development   | 8 (73)                                          | 2 (100)                                               |
| <b><u>HEARING</u></b>                                  |                                                 |                                                       |
| Sig. effect of hearing on comm. development            | 9 (82)                                          | 1 (50)                                                |
| <b><u>Type of Hearing Loss</u></b>                     |                                                 |                                                       |
| Conductive loss                                        | 0 (0)                                           | 0 (0)                                                 |
| Sensorineural loss                                     | 5 (45)                                          | 1 (50)                                                |
| Mixed loss                                             | 5 (45)                                          | 1 (50)                                                |
| <b><u>Degree of Hearing Loss</u></b>                   |                                                 |                                                       |
| Normal hearing                                         | 1 (9)                                           | 0 (0)                                                 |
| Mild loss                                              | 2 (18)                                          | 1 (50)                                                |
| Moderate loss                                          | 3 (27)                                          | 1 (50)                                                |
| Moderately-severe loss                                 | 2 (18)                                          | 0 (0)                                                 |
| Severe-profound loss                                   | 3 (27)                                          | 0 (0)                                                 |
| <b><u>Treatment of Hearing Loss</u></b>                |                                                 |                                                       |
| Uses hearing aids                                      | 5 (45)                                          | 0 (0)                                                 |
| Uses cochlear implant                                  | 0 (0)                                           | 0 (0)                                                 |
| No treatment                                           | 6 (55)                                          | 2 (100)                                               |

**Table 5: continued**

|                                                            | <b>Gesture (n=11)<br/># of Participants (%)</b> | <b>Perlocutionary (n=2)<br/># of Participants (%)</b> |
|------------------------------------------------------------|-------------------------------------------------|-------------------------------------------------------|
| <b><u>BALANCE</u></b>                                      |                                                 |                                                       |
| Sig. effect of balance on<br>comm. development             | 5 (45)                                          | 0 (0)                                                 |
| <b><u>Mobility</u></b>                                     |                                                 |                                                       |
| Walks independently                                        | 3 (27)                                          | 0 (0)                                                 |
| Mobile with assistance                                     | 4 (36)                                          | 1 (50)                                                |
| Does not walk (uses wheelchair)                            | 4 (36)                                          | 1 (50)                                                |
| <b><u>VISION</u></b>                                       |                                                 |                                                       |
| Sig. effect of vision on<br>Comm. development              | 4 (36)                                          | 1 (50)                                                |
| Prevalence of vision problems                              | 11 (100)                                        | 2 (100)                                               |
| <b><u>SMELL</u></b>                                        |                                                 |                                                       |
| Sig. effect of smell on<br>comm. development               | 1 (9)                                           | 0 (0)                                                 |
| <b><u>TASTE</u></b>                                        |                                                 |                                                       |
| Sig. effect of taste on<br>comm. development               | 2 (18)                                          | 0 (0)                                                 |
| <b><u>BEHAVIOR</u></b>                                     |                                                 |                                                       |
| Sig. effect of behavior on<br>overall development          | 2 (18)                                          | 1 (50)                                                |
| Sig. effect of behavior on<br>comm. development            | 3 (27)                                          | 1 (50)                                                |
| Declined to respond                                        | 2 (18)                                          | 0 (0)                                                 |
| <b><u>Behaviors</u></b>                                    |                                                 |                                                       |
| Well-adjusted                                              | 8 (73)                                          | 1 (50)                                                |
| Social or emotional immaturity                             | 9 (82)                                          | 2 (100)                                               |
| Unstable or explosive behavior                             | 5 (45)                                          | 1 (50)                                                |
| Problems with attention                                    | 8 (73)                                          | 2 (100)                                               |
| Overreacts to certain situations                           | 5 (45)                                          | 1 (50)                                                |
| Aggressive or defensive behaviors                          | 4 (36)                                          | 1 (50)                                                |
| Rhythmic rocking                                           | 4 (36)                                          | 1 (50)                                                |
| Self-stimulatory behavior                                  | 9 (82)                                          | 1 (50)                                                |
| Sensitivity to loud sounds                                 | 6 (55)                                          | 1 (50)                                                |
| Lack of communication for<br>interpersonal purposes        | 7 (64)                                          | 1 (50)                                                |
| <b><u>COMMUNICATION TRAINING</u></b>                       |                                                 |                                                       |
| Sig. effect of speech & language<br>on overall development | 11 (100)                                        | 2 (100)                                               |
| <b><u>CURRENT EDUCATIONAL<br/>PLACEMENT</u></b>            |                                                 |                                                       |
| Sig. effect of communication on<br>educational development | 10 (91)                                         | 2 (100)                                               |
| <b><u>Educational Setting</u></b>                          |                                                 |                                                       |
| Mainstream                                                 | 0 (0)                                           | 0 (0)                                                 |
| Inclusive                                                  | 2 (18)                                          | 0 (0)                                                 |
| Self-contained (hearing impaired)                          | 3 (27)                                          | 0 (0)                                                 |
| Self-contained (multiple disabilities)                     | 2 (18)                                          | 0 (0)                                                 |
| Deaf school                                                | 0 (0)                                           | 0 (0)                                                 |
| Early childhood Program                                    | 2 (18)                                          | 0 (0)                                                 |
| Home school                                                | 1 (9)                                           | 2 (100)                                               |
| Not currently enrolled                                     | 1 (9)                                           | 0 (0)                                                 |

## **RESULTS FOR THE GESTURE GROUP**

The gesture communication group represented 35% of all participants. The data for the gesture group is in Table 5. This group required special consideration, in the attempt to determine why they did not develop symbolic language. Each participant was evaluated separately using the selected factors that were likely to be related to the development of symbolic language: neurologic status, severe growth deficiency, success with amplification, communication training, and mobility. It was not known if any of these factors affected communication development; however, there was the possibility that one or more could have critically affected the development process.

While growth deficiency was not a differentiator between any of the communication groups, the presence of severe growth deficiency was reported by some parents during the interview. Since none of the participants with symbolic language were reported to have severe growth deficiency, it was hypothesized that the severity of growth deficiency may have been a possible determinant in the acquisition of symbolic language, and was therefore part of the gesture group analysis. Data for each participant in the gesture group is shown in Table 6.

Of the eleven participants in the gesture group, five participants (A1, A8, A9, C5, C6) had four potential reasons for not developing symbolic language and four participants (A2, A3, A5, A7) had three potential reasons for not developing symbolic language. It was then hypothesized that one single factor may not have contributed to the inability to develop symbolic communication, but instead was possibly caused by some type of interaction of factors. It is less clear why the remaining two participants (A4, A6) did not have symbolic communication. One participant (A4) was possibly was not

**Table 6: Gesture communication group: shaded areas=possible contributory factors to the use of gesture.**

| <b>Participant<br/>(age, sex)</b> | <b>Neurologic<br/>Status</b>      | <b>Growth<br/>Deficiency</b>   | <b>Success w/<br/>Amplification<br/>(HL)</b>                                                 | <b>Speech-<br/>Language<br/>Therapy</b>             | <b>Sign<br/>Language<br/>Instruction</b>            | <b>Mobility</b>                         |
|-----------------------------------|-----------------------------------|--------------------------------|----------------------------------------------------------------------------------------------|-----------------------------------------------------|-----------------------------------------------------|-----------------------------------------|
| A1<br>(8F)                        | Unknown                           | Yes                            | No<br>amplification<br>(Profound)                                                            | Began at 4<br>yrs. of age<br>(stopped at<br>7 yrs.) | Began at 7<br>yrs. of age<br>(stopped at 8<br>yrs.) | Walks<br>with<br>assistance             |
| A2<br>(8F)                        | Seizures                          | Yes                            | No<br>amplification<br>(Mild)                                                                | Began at 4<br>yrs. of age                           | None                                                | Walks<br>independ.                      |
| A3<br>(4F)                        | Agenesis<br>of corpus<br>callosum | Severe<br>(weight: 19<br>lbs.) | No<br>amplification<br>(Mild)                                                                | Began at 3<br>yrs. of age                           | Began at 2<br>yrs. of age                           | Does not<br>walk or<br>crawl            |
| A4<br>(4M)                        | Unknown                           | No                             | Binaural amp.<br>(fit @ 10 mos.<br>– possible<br>underamp.)<br>(Severe)                      | Began at 8<br>mos. of age                           | Sporadic                                            | Walks<br>independ.                      |
| A5<br>(3F)                        | Unknown                           | Severe<br>(weight: 20<br>lbs.) | Binaural amp.<br>(intermittent<br>use - problems<br>with ME)<br>(Sev-Prof)                   | Began at 1<br>yr. of age                            | Began at 2<br>yrs. of age                           | Does not<br>walk or<br>crawl            |
| A6<br>(2M)                        | Unknown                           | Yes                            | Binaural amp.<br>(fit @ 5 mos.)                                                              | Began at 8<br>mos. of age                           | Began at 1<br>yr. of age                            | Walks<br>with<br>assistance             |
| A7<br>(9M)                        | Unknown                           | No                             | No amp.<br>(Profound)                                                                        | Began at 5<br>yrs. of age                           | Began at 6<br>yrs. of age                           | Walks<br>independ.                      |
| A8<br>(3F)                        | Seizures                          | Yes                            | No amp.<br>(Normal)                                                                          | None                                                | None                                                | Does not<br>walk or<br>crawl            |
| A9<br>(11M)                       | Enlarged<br>ventricles            | Yes                            | No amp.<br>(Moderate)                                                                        | Began at 1<br>yr. of age                            | None                                                | Does not<br>walk;<br>scoots on<br>floor |
| C5<br>(11F)                       | Unknown                           | Yes                            | Binaural amp.<br>(fit @ 11 yrs.)<br>(Moderate)                                               | Began at 4<br>yrs. of age                           | Began at 11<br>yrs. of age                          | Walks<br>with<br>assistance             |
| C6<br>(7M)                        | Diagnosis of<br>autism            | Yes                            | Unilateral amp.<br>(fit @ 3 yrs. –<br>problems w/<br>ME and fit on<br>pinna)<br>(Mod-Severe) | Began in<br>infancy                                 | Began at 4<br>yrs. of age                           | Walks<br>with<br>assistance             |

receiving adequate amplification for his hearing loss and did not receive consistent sign language instruction. In addition, this participant had never had a neurologic examination, a finding that was common in many of the participants in the gesture group. With the status of his neurologic function being unknown along with the other factors of amplification and sporadic sign language instruction, it is impossible to know what is causing his inability to acquire symbolic language. The other participant (A6) did not walk independently but was able to walk with assistance and had unknown neurologic status. None of the other factors were of concern. It is, therefore, a possibility that this participant is in a transition between the no symbolic and symbolic group due to his young age.

## CHAPTER 5

### DISCUSSION

The majority of parents (77%-81%) reported that both physical and sensory problems affected their child's overall development and the development of communication. While no formal communication evaluations were performed as a part of this study, it is the investigator's opinion that the parents' judgments were correct. Almost all of the children did not seem to have expressive communication that was commensurate with their chronological age.

Of the 31 participants in this study, 18 (58%) developed symbolic language, while the remaining 13 (42%) did not. This distribution is similar to the study conducted by Collins (1993), who reported that approximately half of the children who are deaf-blind do not use symbolic communication. In this regard, CHARGE syndrome may be similar to other deaf-blind disorders.

All of the participants had significant physical, sensory, and behavioral involvement. The most common problems that were reported among the participants in the present study are listed below:

|                                  |      |
|----------------------------------|------|
| • Vision problems                | 100% |
| • Hearing loss                   | 94%  |
| • Swallowing/Feeding problems    | 87%  |
| • Social or emotional immaturity | 84%  |
| • Hypotonia                      | 81%  |
| • Choanal atresia                | 74%  |
| • Heart problems                 | 74%  |
| • Problems with attention        | 68%  |
| • Self-stimulatory behavior      | 61%  |
| • Gastrostomy tube               | 58%  |
| • Facial problems                | 55%  |
| • Growth deficiency              | 55%  |

Despite the presence of these conditions, none of the conditions were significant in differentiating those participants who developed symbolic language and those who did not. Similarly, there were no differences between participants with symbolic language and those with no symbolic language in the four diagnostic criteria for CHARGE: coloboma, choanal atresia, cranial nerve dysfunction, and characteristic CHARGE ear (Blake et al., 1998). Therefore, although many disorders were present in the experimental sample, most physical and sensory disorders were not determinants in the acquisition of symbolic language. Bates (1979) reported that communication development and physical development facilitate each other. The present findings suggest, that for CHARGE, communication development occurs in the presence of many physical disorders. However, it may be that, in CHARGE, communication development may be affected primarily by specific disorders.

### **DETERMINANTS OF COMMUNICATION ABILITY**

There were a number of factors identified in this study that may have contributed to the development of symbolic language. First, the presence of major neurological damage may be a critical factor in the development of symbolic communication. This was true for both of the participants in the perlocutionary group (A10, A11) and possibly for several participants in the gesture group (A2, A3, A8, A9, C6).

A second factor that may have affected the development of symbolic communication was severe growth deficiency. This condition was present in one of the participants (A10) in the perlocutionary group and two of the participants in the gesture group (A3 and A5). The severity of growth deficiency in these participants resulted in

their inability to walk or crawl and may have also affected their ability to acquire symbolic language.

A third factor was the ability to walk independently. This finding is similar to that of Petroff (2001), who found that approximately half of the children with deaf-blindness who did not have symbolic language did not walk independently while those who did have symbolic language were able to walk independently. While the inability to walk may be indicative of possible neurological involvement, it is hypothesized that, in CHARGE, mobility may provide a child with the ability to overcome the effects of sensory deficits. Colobomas, the major cause of visual problems in CHARGE, create a void in the visual field that cannot be corrected by surgery or corrective lenses. Mobility may give a child the opportunity to position himself or herself so that images are placed in visual fields that are acceptable, thus at least partially overcoming a visual deficit. Mobility may also serve the same function in overcoming hearing loss by moving to a location that increases loudness or improves the signal-to-background-noise ratio. Davenport (2001) has used the term “communication bubble” to denote the areas around a child in which communication is possible or optimal. Davenport has stressed the need for talkers to position themselves in the communication bubble, but the results of the present study suggest that the greatest advantage may be derived when the child is able to position himself or herself in the unique location(s) that optimize communication. This phenomenon was observed directly by the investigator in her conversations with children with CHARGE.

The fourth factor in acquiring symbolic language that was identified in this study was the type of communication training used, as well as the age at which the training

began and the consistency, or duration, of the training. The present data indicate that the probability of developing symbolic language in children with CHARGE is greatest with total communication training that is undertaken very early in life and continues until approximately age seven. These results need to be confirmed on other children with CHARGE, but the present results provide unexpectedly strong evidence that total communication appears to provide children with CHARGE the best opportunity for developing symbolic communication. One possible explanation for these results is that total communication provides auditory and visual information to children who have deficits in both of these sensory modalities. Typically, it is difficult to specify the type and degree of hearing and vision losses experienced by children with CHARGE early in life. In most cases, visual deficits cannot be corrected and the hearing losses in CHARGE are some of the most difficult to overcome with amplification (Shah et al., 1998; Thelin et al., 1986). Total communication may allow a child to receive language information in the best modality for that child. Or, if the child is able to receive partial information in both the auditory and visual channels, the total communication approach may provide valuable redundancy of language information.

A fifth factor that was identified is success with amplification for the development of speech. Unexpectedly, it was found that success with amplification was not significantly related to the degree of hearing loss. The factors related to success were not determined in the present study, but possible factors include neurologic status and issues related to hearing aid usage with ear anomalies, active middle-ear disease, tactile defensiveness, and the inability to hold the head upright.

## **BEHAVIOR**

In the present study, questions were not asked to parents about intelligence because of the complexity of the issue and because of its emotional aspects. The parents' responses to the investigator suggest that they may also have special concerns and emotional involvement with issues related to behavior. Parents had the firm conviction that much of the inappropriate behavior exhibited by their child with CHARGE was the result of frustration from the inability to communicate well. If this were true, then it would have been expected that the children with no symbolic language would have been reported to have more inappropriate behaviors than those with symbolic language. The present data did not support the hypothesis that inappropriate behavior is related to the acquisition of symbolic language. While hearing impairment has been associated with behavior and personality problems and blindness with autistic-like behaviors (Carvill, 2001), it is unclear how the multiple deficits in CHARGE affect behavior and communication.

## **FUTURE RESEARCH**

The primary methods used in this research for most of the participants in the study included contacting parents, meeting with the parents during a conference, and collecting data prior to and after the conference. This combination of methods proved to be useful for acquiring information on the very wide range of topics addressed in the present study. This approach used in the present study permitted the assessment of many combinations of factors that previously had been studied in isolation. Future research of this type may

be of value in establishing a comprehensive model of the complex problems in CHARGE related to communication or other aspects of the disorder.

A major finding of the present study was that many factors that were anticipated to affect the development of communication such as multiple sensory deficits and severe physical problems that are present in CHARGE were, in fact, not related to success in the acquisition of symbolic language. However, there were a number of factors of importance or potential importance to communication that were identified, that were not anticipated, and that are incompletely understood. These factors include neurologic status, physical growth, behavior, mobility, and communication training. Knowledge of the contributions of these factors may assist in understanding the development of symbolic communication for those who use gestures alone and also in the improvement of communication abilities among those who have symbolic language that is not age appropriate.

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## **APPENDICES**

**APPENDIX A**  
**Parent Questionnaire**

## “The Determinants of Communication Ability in CHARGE Syndrome”

### PARENT QUESTIONNAIRE

Name of Child: \_\_\_\_\_ Age: \_\_\_\_\_  
Race of Child: \_\_\_\_\_ Gender: M F  
Name of Parent(s): \_\_\_\_\_  
Address: \_\_\_\_\_  
Phone: \_\_\_\_\_ Email: \_\_\_\_\_

The purpose of the present study is to evaluate the factors that may influence the development of communication in children with CHARGE. In this questionnaire, you will be asked to provide judgments and information about your child's physical health, sensory function, and behavior. Your answers will help us determine the factors that affect your child's ability to develop communication skills.

There are six sections below. In each section (Physical Health, Sensory Function, Behavior, Communication, Speech and Language, and Education), you will be asked to make a judgment about your child's overall development and another judgment about your child's development of communication. Following those judgments, you will be asked to provide specific information about conditions that your child may have.

#### I. PHYSICAL HEALTH

Children with CHARGE often have medical and physical problems that affect them in many ways. Do you think your child's physical health has affected his/her **overall development** or ability to function? Please indicate the degree to which you think physical and medical problems have affected your child's development.

- ☐ Very significant effect
- ☐ Significant effect
- ☐ Some effect
- ☐ Slight effect
- ☐ No effect

Do you think your child's physical health has affected his/her **development of communication**? Please indicate the degree to which you think physical and medical problems have affected your child's development.

- ☐ Very significant effect
- ☐ Significant effect
- ☐ Some effect
- ☐ Slight effect
- ☐ No effect

Please indicate below the medical conditions your child has and indicate if you think that the condition affected communication.

☐ Choanal Atresia (narrowed nasal passages/difficulty breathing) \_\_\_\_\_

☐ Heart Problems \_\_\_\_\_

☐ Cleft Lip and/or Palate \_\_\_\_\_

☐ Swallowing/Feeding Problems \_\_\_\_\_

☐ Facial Problems \_\_\_\_\_

☐ Hypotonia (low muscle tone) \_\_\_\_\_

☐ Growth Deficiency \_\_\_\_\_

☐ Other \_\_\_\_\_

## II. SENSORY FUNCTION

In addition to physical and medical problems, sensory function may also affect development. Do you think your child's sensory function has affected his/her **overall development**? Please indicate the degree to which you think problems with hearing, vision, smell, taste, touch, and balance have combined to affect your child's development.

- ☐ Very significant effect
- ☐ Significant effect
- ☐ Some effect
- ☐ Slight effect
- ☐ No effect

Do you think your child's sensory function has affected his/her **development of communication**? Please indicate the degree to which you think problems with hearing, vision, smell, taste, touch, and balance have combined to affect your child's development of communication.

- ☐ Very significant effect
- ☐ Significant effect
- ☐ Some effect
- ☐ Slight effect
- ☐ No effect

If your child has developmental delays and communication difficulties resulting from sensory problems, list the sensory problems (hearing, vision, smell, taste, balance) in order that are most significant:

First \_\_\_\_\_  
Second \_\_\_\_\_  
Third \_\_\_\_\_

Please provide information about each of the specific senses below and how each as affected your child's communication.

**Hearing:** Please indicate the degree to which you think your child's hearing has affected his/her development of communication.

- ☐ Very significant effect
- ☐ Significant effect
- ☐ Some effect
- ☐ Slight effect
- ☐ No effect

Degree and type of hearing loss: \_\_\_\_\_

Does your child wear hearing aids? If so, since what age and how often are they worn? \_\_\_\_\_

What type of hearing aids does your child wear? Does it fit over the ear or in the ear? Please include the make and model. \_\_\_\_\_

Describe the benefit from the hearing aids. Do you feel that your child hears normally with his/her hearing aids? \_\_\_\_\_

Have ear infections affected your child's ability to use hearing aids? \_\_\_\_\_

Does your child use an assistive listening device (i.e. FM system)? If so, what type of device is it? \_\_\_\_\_

Does your child have a cochlear implant? If so, what type of implant is it and when was it implanted? Describe the benefit from the implant. \_\_\_\_\_

**Vision** : Please indicate the degree to which you think your child's vision has affected his/her development of communication.

- ☐ Very significant effect
- ☐ Significant effect
- ☐ Some effect
- ☐ Slight effect
- ☐ No effect

**Does your child have colobomas? Where?**

Does your child have other visual problems? Describe. \_\_\_\_\_

How does your child's visual problem affect his/her ability to perceive? Do you have to be close to the child for him/her to see you? Does your child hang upside down? \_\_\_\_\_

**Smell** : Please indicate the degree to which you think your child's smell has affected his/her development of communication.

- ☐ Very significant effect
- ☐ Significant effect
- ☐ Some effect
- ☐ Slight effect
- ☐ No effect

Is your child's sense of smell normal? \_\_\_\_\_

**If your child's smell is abnormal, how do you know?**

If your child's smell is abnormal, does it affect feeding? \_\_\_\_\_

**Taste** : Please indicate the degree to which you think your child's taste has affected his/her development of communication.

- ☐ Very significant effect
- ☐ Significant effect
- ☐ Some effect
- ☐ Slight effect
- ☐ No effect

Is your child's sense of taste normal? \_\_\_\_\_

If your child's taste is abnormal, how do you know? \_\_\_\_\_

Does your child often put objects in his/her mouth to determine their shape? \_\_\_\_\_

**Balance** : Please indicate the degree to which you think your child's balance has affected his/her development of communication.

- ☐ Very significant effect
- ☐ Significant effect
- ☐ Some effect
- ☐ Slight effect
- ☐ No effect

Describe your child's mobility.

Does your child walk? If so, at what age did he/she begin to walk?

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### III. BEHAVIOR

Children with CHARGE have been reported to have problems with behavior. It is possible that these problems may contribute to difficulties with communication and development. Do you think your child's behavior has affected his/her **overall development**? Please indicate the degree to which you think behavior has affected your child's development.

- ☐ Very significant effect
- ☐ Significant effect
- ☐ Some effect
- ☐ Slight effect
- ☐ No effect

Do you think your child's behavior has affected his/her **development of communication**? Please indicate the degree to which you think behavior has affected your child's communication.

- ☐ Very significant effect
- ☐ Significant effect
- ☐ Some effect
- ☐ Slight effect
- ☐ No effect

Do you have problems with your child's behavior? Can you describe them in your own words? \_\_\_\_\_

Do others have problems with your child's behavior? Please describe them. \_\_\_\_\_

Has the child had a behavioral evaluation? Is so, what were the results? \_\_\_\_\_

Do any of the following apply to your child?

- ☐ Well-adjusted
- ☐ Social or emotional immaturity
- ☐ Unstable or explosive behavior
- ☐ Problems with attention
- ☐ Overreacts to certain situations
- ☐ Aggressive or defensive behaviors
- ☐ Rhythmic rocking
- ☐ Self-stimulatory behavior
- ☐ Sensitivity to loud sounds
- ☐ Lack of communication for interpersonal purposes
- ☐ Other \_\_\_\_\_

#### IV. COMMUNICATION

Children with CHARGE often have difficulties communicating. Please indicate the degree to which your child's communication has affected his/her **overall development**.

- ☐ Very significant effect
- ☐ Significant effect
- ☐ Some effect
- ☐ Slight effect
- ☐ No effect

Do you think your child's communication has been affected by CHARGE syndrome?  
Please indicate the degree to which your child's communication has been affected by CHARGE.

- ☐ Very significant effect
- ☐ Significant effect
- ☐ Some effect
- ☐ Slight effect
- ☐ No effect

Indicate your child's primary modes of communication; rank order them 1, 2, etc. Add any information about their communication that you feel is important.

\_\_\_\_\_ Speech \_\_\_\_\_  
\_\_\_\_\_ Sign Language (such as ASL) \_\_\_\_\_  
\_\_\_\_\_ Own Signs \_\_\_\_\_  
\_\_\_\_\_ Gestures \_\_\_\_\_  
\_\_\_\_\_ Communication Board \_\_\_\_\_  
\_\_\_\_\_ Other \_\_\_\_\_

If your child uses speech, please describe. (i.e. single words, phrases, continuous discourse) \_\_\_\_\_  
\_\_\_\_\_

Is your child's communication age-appropriate, or similar to children of the same age who do not have CHARGE? \_\_\_\_\_

Do you have difficulty understanding your child's communication? Please describe. \_\_\_\_\_  
\_\_\_\_\_

Do others have difficulty understanding your child's communication? Please describe. \_\_\_\_\_  
\_\_\_\_\_

If you have further information regarding your child's communication, please include that information in the comments section below.

\_\_\_\_\_  
\_\_\_\_\_  
\_\_\_\_\_  
\_\_\_\_\_  
\_\_\_\_\_  
\_\_\_\_\_

## V. SPEECH AND LANGUAGE

Please indicate the degree to which you think speech and language has affected your child's **overall development**.

- ☐ Very significant effect
- ☐ Significant effect
- ☐ Some effect
- ☐ Slight effect
- ☐ No effect

Please indicate the degree to which you think speech and language has affected your child's **communication** abilities.

- ☐ Very significant effect
- ☐ Significant effect
- ☐ Some effect
- ☐ Slight effect
- ☐ No effect

Has your child received speech and language therapy? If so, at what age did the therapy begin and how much therapy was received? \_\_\_\_\_

Has your child received instruction in sign language? Is so, at what age did the instruction begin and how much instruction was received? \_\_\_\_\_

## VI. EDUCATION

Do you think that the problems due to CHARGE syndrome affected your child's overall educational development? Please indicate the degree to which education has affected your child's **overall educational development**?

- ☐ Very significant effect
- ☐ Significant effect
- ☐ Some effect
- ☐ Slight effect
- ☐ No effect

The ability to communicate may affect a child's educational level. Please indicate the degree to which you think **communication** has affected your child's educational development.

- ☐ Very significant effect
- ☐ Significant effect
- ☐ Some effect
- ☐ Slight effect
- ☐ No effect

Is your child enrolled in school? If so, what type of educational setting is it? \_\_\_\_\_

Is your child's educational performance similar to children of the same age who do not have CHARGE? \_\_\_\_\_

#### COMMENTS

If you have any additional information about any of the preceding topics, please include that information below. \_\_\_\_\_

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\_\_\_\_\_  
Parent Signature

\_\_\_\_\_  
Date

Thank you for your time in completing this questionnaire - your participation is greatly appreciated. At the completion of this study, you will receive a summary of the results of the study. If you have any questions regarding the questions asked in this questionnaire or about this study, please contact Dr. James Thelin at (865) 974-1796 or [jthelin@utk.edu](mailto:jthelin@utk.edu) or Jill Fussner at [jfussner03@aol.com](mailto:jfussner03@aol.com).

**APPENDIX B**  
**Information Form**

## American Information Form

This research study is being conducted during the Mid-Atlantic CHARGE Syndrome Conference by the University of Tennessee.

The study is completely independent of the Mid-Atlantic CHARGE Syndrome Program. Attendance to the conference is not dependent on your participation in this research project.

### Communication Abilities of Children with CHARGE A Study to Be Conducted at the Mid-Atlantic CHARGE Conference \_\_\_\_\_o0o\_\_\_\_\_

Jill Fussner, Master's Student in Audiology, and James W. Thelin, Ph.D., Associate Professor of Audiology  
Department of Audiology and Speech Pathology, University of Tennessee, Knoxville, TN 37966-0740

We will be attending the Mid-Atlantic CHARGE Conference and we would like the opportunity to study your child's ability to communicate. The purpose of this note is to determine whether you are interested in participating. Participation would involve the following three activities:

- 1) completing a mailed questionnaire about medical and behavioral factors that might affect communication,
- 2) meeting with us at the conference for about 15 minutes to discuss the questionnaire that you completed, and
- 3) permitting us to contact you by phone or email after the conference to clarify issues not covered by the questionnaire or at our meeting.

We are conducting this because there is very little information on communication in CHARGE . Parents and educators need to know what can be done to maximize the communication abilities of children with CHARGE. It is our intent to present the results at the 2003 CHARGE Conference and at other professional meetings as well as to publish the results. A summary of the results will be sent to each participating family.

***Participation in this study is voluntary. Also, if you agree to participate, you may withdraw from the study at any time. This study will be conducted in accordance with the policy of the University of Tennessee for the use of human subjects. This policy requires that confidentiality of information be maintained.***

If you are interested in participating, please respond by completing the information below and emailing it to Jill Fussner at [Jfussner03@aol.com](mailto:Jfussner03@aol.com) or mailing it to Jill Fussner, University of Tennessee, Department of Audiology and Speech Pathology, 457 South Stadium Hall, Knoxville, TN 37996-0740. If you have any special questions, contact James Thelin at (865) 974-1796.

Parent(s):

Child's Name:

Child's Age:

Address:

Telephone Number:

We appreciate your consideration in taking part in this study and we look forward to meeting each of you in June.--- Jill Fussner and Dr. James Thelin

***If you would like to participate, but are registering late –  
please contact us when you receive this – even though you haven't registered.***

## Canadian Information Form

### Communication Abilities of Children with CHARGE Syndrome

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Jill Fussner, Master's Student in Audiology & James W. Thelin, Ph.D., Associate Professor of  
Audiology  
Department of Audiology and Speech Pathology  
University of Tennessee, Knoxville, TN 37966-0740

We are currently conducting a research study and would like the opportunity to study your child's ability to communicate. The purpose of this note is to determine whether you are interested in participating. Participation would involve the following three activities: completing a mailed questionnaire about medical and behavioral factors that might affect communication, and permitting us to contact you by phone or email to clarify issues not covered by the questionnaire.

We are conducting this study because there is very little information on communication in CHARGE. Parents and educators need to know what can be done to maximize the communication abilities of children with CHARGE. It is our intent to present the results at the 2003 CHARGE Conference and at other professional meetings as well as to publish the results. A summary of the results will be sent to each participating family.

***Participation in this study is voluntary. Also, if you agree to participate, you may withdraw from the study at any time. This study will be conducted in accordance with the policy of the University of Tennessee for the use of human subjects. This policy requires that confidentiality of information be maintained.***

If you are interested in participating, please respond by completing the information below and emailing it to Jill Fussner at [Jfussner03@aol.com](mailto:Jfussner03@aol.com) or mailing it to Jill Fussner, University of Tennessee, Department of Audiology and Speech Pathology, 457 South Stadium Hall, Knoxville, TN 37996-0740. If you have any special questions, contact James Thelin at (865) 974-1796.

Parent(s):

Child's Name:

Child's Age:

Address:

Telephone Number:

We appreciate your consideration in taking part in this study and we look forward to talking with each of you soon.

--- Jill Fussner and Dr. James Thelin

## **APPENDIX C**

### **Parental Informed Consent Form**

## American Consent Form

### PARENTAL INFORMED CONSENT

for

#### The Determinants of Communication Ability in CHARGE Syndrome: A Study to be Conducted at the Mid-Atlantic CHARGE Syndrome Conference

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Jill Fussner, Master's Student in Audiology, and James W. Thelin, Ph.D., Associate Professor of Audiology  
Department of Audiology and Speech Pathology, University of Tennessee, Knoxville, TN 37966-0740,  
Tel: 865-974-1796

We would like the opportunity to study your child's ability to communicate. Participation would involve the following activities:

- 1) completing the questionnaire the attached questionnaire about medical, behavioral, and sensory factors that might have affected your child's ability to communicate (and returning the questionnaire and this consent form),
- 2) meeting with us at the conference for about 15-20 minutes so that we can speak with you about your child and the questionnaire you completed, and
- 3) permitting us to contact you by phone or email to clarify issues not covered by the questionnaire.

We are conducting this study because there is very little information on communication in CHARGE. Parents and educators need to know what can be done to maximize the communication abilities of children with CHARGE. It is our intent to share our results with you personally, to present the results at the 2003 CHARGE Conference in Cleveland, OH, and at other professional meetings as well as to publish the results. A summary of the results will be sent to each participating family.

***There are no known risks for you or your child in this study and participation in this study is voluntary. This study will be conducted in accordance with the policies of the University of Tennessee for the use of human subjects. These policies require that confidentiality of information be maintained.***

***If you agree to participate, you may withdraw from the study at any time with no penalty to you or your child. If you or your child withdraw from the study, any data that have been collected will be destroyed.***

If you have any question about the study or the procedures, you may contact the principal investigator, Jill C. Fussner ([Jfussner03@aol.com](mailto:Jfussner03@aol.com)) or the faculty advisor, James W. Thelin ([jthelin@utk.edu](mailto:jthelin@utk.edu)) at the address and telephone number at the top of this form. If you have any questions about your rights or your child's rights as a participant, you may contact the Compliance Section of the Office of Research at 865-974-3466.

I have read and understand the above information. I have received a copy of this form. I agree to participate and to allow my child to participate in this study.

Parent signature \_\_\_\_\_

Date \_\_\_\_\_

Parent signature \_\_\_\_\_

Date \_\_\_\_\_

## Canadian Consent Form

### PARENTAL INFORMED CONSENT for The Determinants of Communication Ability in CHARGE Syndrome \_\_\_\_\_o0o\_\_\_\_\_

Jill Fussner, Master's Student in Audiology, and James W. Thelin, Ph.D., Associate Professor of Audiology  
Department of Audiology and Speech Pathology, University of Tennessee, Knoxville, TN 37966-0740,  
Tel: 865-974-1796

We would like the opportunity to study your child's ability to communicate. Participation would involve the following activities:

- 1) completing the questionnaire the attached questionnaire about medical, behavioral, and sensory factors that might have affected your child's ability to communicate (and returning the questionnaire and this consent form), and
- 2) permitting us to contact you by phone or email to clarify issues not covered by the questionnaire.

We are conducting this study because there is very little information on communication in CHARGE. Parents and educators need to know what can be done to maximize the communication abilities of children with CHARGE. It is our intent to share our results with you personally, to present the results at the 2003 CHARGE Conference in Cleveland, OH, and at other professional meetings as well as to publish the results. A summary of the results will be sent to each participating family.

***There are no known risks for you or your child in this study and participation in this study is voluntary. This study will be conducted in accordance with the policies of the University of Tennessee for the use of human subjects. These policies require that confidentiality of information be maintained.***

***If you agree to participate, you may withdraw from the study at any time with no penalty to you or your child. If you or your child withdraw from the study, any data that have been collected will be destroyed.***

If you have any question about the study or the procedures, you may contact the principal investigator, Jill C. Fussner ([Jfussner03@aol.com](mailto:Jfussner03@aol.com)) or the faculty advisor, James W. Thelin ([jthelin@utk.edu](mailto:jthelin@utk.edu)) at the address and telephone number at the top of this form. If you have any questions about your rights or your child's rights as a participant, you may contact the Compliance Section of the Office of Research at 865-974-3466.

I have read and understand the above information. I have received a copy of this form. I agree to participate and to allow my child to participate in this study.

Parent signature \_\_\_\_\_

Date \_\_\_\_\_

Parent signature \_\_\_\_\_

Date \_\_\_\_\_

**APPENDIX D**

**American and Canadian Participant Data**

|                                                            | American Group (n=25)<br># of Participants (%) | Canadian Group (n=6)<br># of Participants (%) | TOTAL (n=31) |
|------------------------------------------------------------|------------------------------------------------|-----------------------------------------------|--------------|
| <b>PRIMARY MODE<br/>OF COMMUNICATION</b>                   |                                                |                                               |              |
| Sig. effect of communication on<br>overall development     | 20 (80)                                        | 3 (50)                                        | 23 (74)      |
| Sig. effect of CHARGE on<br>comm. development              | 21 (84)                                        | 6 (100)                                       | 27 (87)      |
| <u>Communication Mode</u>                                  |                                                |                                               |              |
| Speech                                                     | 8 (32)                                         | 3 (50)                                        | 11 (35)      |
| Sign Language                                              | 5 (20)                                         | 1 (17)                                        | 6 (19)       |
| Gesture                                                    | 9 (36)                                         | 2 (33)                                        | 11 (35)      |
| Perlocutionary                                             | 2 (8)                                          | 0 (0)                                         | 2 (6)        |
| <b>PHYSICAL HEALTH</b>                                     |                                                |                                               |              |
| Sig. effect of physical health<br>on overall development   | 21 (84)                                        | 4 (67)                                        | 25 (81)      |
| Sig. effect of physical health<br>on comm. development     | 19 (76)                                        | 5 (83)                                        | 24 (77)      |
| <u>Physical Conditions</u>                                 |                                                |                                               |              |
| Choanal atresia                                            | 18 (72)                                        | 5 (83)                                        | 23 (74)      |
| Heart problems                                             | 18 (72)                                        | 5 (83)                                        | 23 (74)      |
| Cleft lip/palate                                           | 7 (28)                                         | 3 (50)                                        | 10 (32)      |
| Swallowing/feeding problems                                | 22 (88)                                        | 5 (83)                                        | 27 (87)      |
| Facial problems                                            | 14 (56)                                        | 3 (50)                                        | 17 (55)      |
| Hypotonia                                                  | 21 (84)                                        | 4 (67)                                        | 25 (81)      |
| Growth deficiency                                          | 14 (56)                                        | 3 (50)                                        | 17 (55)      |
| Tracheostomy                                               | 2 (8)                                          | 0 (0)                                         | 2 (6)        |
| Gastrostomy tube                                           | 14 (56)                                        | 4 (67)                                        | 18 (58)      |
| Neurologic problems (structural)                           | 4 (16)                                         | 1 (17)                                        | 5 (16)       |
| Neurologic problems (other)                                | 4 (16)                                         | 1 (17)                                        | 5 (16)       |
| <b>SENSORY FUNCTION</b>                                    |                                                |                                               |              |
| Sig. effect of sensory function<br>on overall development  | 20 (80)                                        | 5 (83)                                        | 25 (81)      |
| Sig. effect of sensory function on<br>on comm. development | 19 (76)                                        | 5 (83)                                        | 24 (77)      |
| <b>HEARING</b>                                             |                                                |                                               |              |
| Sig. effect of hearing on<br>comm. development             | 19 (76)                                        | 5 (83)                                        | 24 (77)      |
| <u>Type of Hearing Loss</u>                                |                                                |                                               |              |
| Conductive loss                                            | 0 (0)                                          | 0 (0)                                         | 0 (0)        |
| Sensorineural loss                                         | 9 (36)                                         | 2 (33)                                        | 11 (35)      |
| Mixed loss                                                 | 14 (56)                                        | 4 (67)                                        | 18 (58)      |
| <u>Degree of Hearing Loss</u>                              |                                                |                                               |              |
| Normal hearing                                             | 2 (8)                                          | 0 (0)                                         | 2 (6)        |
| Mild loss                                                  | 8 (32)                                         | 1 (17)                                        | 9 (29)       |
| Moderate loss                                              | 3 (12)                                         | 2 (33)                                        | 5 (16)       |
| Moderately-Severe loss                                     | 4 (16)                                         | 2 (33)                                        | 6 (19)       |
| Severe-Profound loss                                       | 8 (32)                                         | 1 (17)                                        | 9 (29)       |
| <u>Treatment of Hearing Loss</u>                           |                                                |                                               |              |
| Uses Hearing aids                                          | 12 (48)                                        | 5 (83)                                        | 17 (55)      |
| Uses cochlear implant                                      | 1 (4)                                          | 0 (0)                                         | 1 (3)        |

|                                                            | American Group (n=25)<br># of Participants (%) | Canadian Group (n=6)<br># of Participants (%) | TOTAL (n=31) |
|------------------------------------------------------------|------------------------------------------------|-----------------------------------------------|--------------|
| <b><u>BALANCE</u></b>                                      |                                                |                                               |              |
| Sig. effect of balance on<br>comm. development             | 10 (40)                                        | 3 (50)                                        | 13 (42)      |
| <b><u>Mobility</u></b>                                     |                                                |                                               |              |
| Walks independently                                        | 15 (60)                                        | 4 (67)                                        | 19 (61)      |
| Mobile with assistance                                     | 5 (20)                                         | 2 (33)                                        | 7 (23)       |
| Does not walk (uses wheelchair)                            | 5 (20)                                         | 0 (0)                                         | 5 (16)       |
| <b><u>VISION</u></b>                                       |                                                |                                               |              |
| Sig. effect of vision on<br>comm. development              | 10 (40)                                        | 3 (50)                                        | 13 (42)      |
| Prevalence of vision problems                              | 25 (100)                                       | 6 (100)                                       | 31 (100)     |
| <b><u>SMELL</u></b>                                        |                                                |                                               |              |
| Sig. effect of smell on<br>comm. development               | 3 (12)                                         | 0 (0)                                         | 3 (10)       |
| <b><u>TASTE</u></b>                                        |                                                |                                               |              |
| Sig. effect of taste on<br>comm. development               | 2 (8)                                          | 2 (33)                                        | 4 (13)       |
| <b><u>BEHAVIOR</u></b>                                     |                                                |                                               |              |
| Sig. effect of behavior on<br>overall development          | 7 (28)                                         | 2 (33)                                        | 9 (29)       |
| Sig. effect of behavior on<br>comm. development            | 8 (32)                                         | 1 (17)                                        | 9 (29)       |
| Declined to respond                                        | 5 (20)                                         | 2 (33)                                        | 7 (23)       |
| <b><u>Behaviors</u></b>                                    |                                                |                                               |              |
| Well-adjusted                                              | 14 (56)                                        | 2 (33)                                        | 16 (52)      |
| Social or emotional immaturity                             | 20 (80)                                        | 6 (100)                                       | 26 (84)      |
| Unstable or explosive behavior                             | 6 (24)                                         | 4 (67)                                        | 10 (32)      |
| Problems with attention                                    | 17 (68)                                        | 4 (67)                                        | 21 (68)      |
| Overreacts to certain situations                           | 11 (44)                                        | 2 (33)                                        | 13 (42)      |
| Aggressive or defensive behaviors                          | 11 (44)                                        | 3 (50)                                        | 14 (45)      |
| Rhythmic rocking                                           | 4 (16)                                         | 1 (17)                                        | 5 (16)       |
| Self-stimulatory behavior                                  | 14 (56)                                        | 5 (83)                                        | 19 (61)      |
| Sensitivity to loud sounds                                 | 10 (40)                                        | 5 (83)                                        | 15 (48)      |
| Lack of communication for<br>interpersonal purposes        | 10 (40)                                        | 4 (67)                                        | 14 (45)      |
| <b><u>COMMUNICATION TRAINING</u></b>                       |                                                |                                               |              |
| Sig. effect of speech & language<br>on overall development | 24 (96)                                        | 5 (83)                                        | 29 (93)      |
| <b><u>CURRENT EDUCATIONAL<br/>PLACEMENT</u></b>            |                                                |                                               |              |
| Sig. effect of communication on<br>educational development | 22 (88)                                        | 4 (67)                                        | 26 (84)      |
| <b><u>Educational Setting</u></b>                          |                                                |                                               |              |
| Mainstream                                                 | 4 (16)                                         | 0 (0)                                         | 4 (13)       |
| Inclusive                                                  | 3 (12)                                         | 4 (67)                                        | 7 (23)       |
| Self-contained (hearing impaired)                          | 5 (20)                                         | 0 (0)                                         | 5 (16)       |
| Self-contained (multiple disabilities)                     | 1 (4)                                          | 2 (33)                                        | 3 (10)       |
| Deaf school                                                | 2 (8)                                          | 0 (0)                                         | 2 (6)        |
| Early childhood program                                    | 2 (8)                                          | 0 (0)                                         | 2 (6)        |
| Home school                                                | 5 (20)                                         | 0 (0)                                         | 5 (16)       |
| Not currently enrolled                                     | 3 (12)                                         | 0 (0)                                         | 3 (10)       |

## **APPENDIX E**

### **Individual Participant Data**

**\*\*** On the individual data tables, the “A” in the participant number signifies that the participant was in the American experimental group, while the “C” represents a participant in the Canadian experimental group.

**\*\*** On the data tables, a “1” signifies the presence of a condition, while the “0” signifies that a condition was not present.

| Participant Number                                     | A1 | A2 | A3 | A4 | A5 | A6 | A7 |
|--------------------------------------------------------|----|----|----|----|----|----|----|
| Gender                                                 | F  | F  | F  | M  | F  | M  | M  |
| Age                                                    | 8  | 8  | 4  | 4  | 3  | 2  | 9  |
| Race                                                   | W  | W  | W  | W  | W  | W  | W  |
| Parents interview: personal                            | 1  | 1  | 1  | 1  | 1  | 1  | 1  |
| Parents interview: telephone/email                     | 1  | 1  | 1  | 1  | 0  | 1  | 1  |
| Child observed                                         | 1  | 1  | 1  | 1  | 1  | 1  | 1  |
| <b>PRIMARY MODE</b>                                    |    |    |    |    |    |    |    |
| <b>OF COMMUNICATION</b>                                |    |    |    |    |    |    |    |
| Sig. effect of communication on overall development    | 1  | 1  | 1  | 1  | 1  | 1  | 1  |
| Sig. effect of CHARGE on comm. development             | 1  | 1  | 0  | 1  | 1  | 1  | 1  |
| <b>Communication Mode</b>                              |    |    |    |    |    |    |    |
| Speech                                                 | 0  | 0  | 0  | 0  | 0  | 0  | 0  |
| Sign Language                                          | 0  | 0  | 0  | 0  | 0  | 0  | 0  |
| Gesture                                                | 1  | 1  | 1  | 1  | 1  | 1  | 1  |
| Perlocutionary                                         | 0  | 0  | 0  | 0  | 0  | 0  | 0  |
| <b>PHYSICAL HEALTH</b>                                 |    |    |    |    |    |    |    |
| Sig. effect of physical health on overall development  | 1  | 1  | 0  | 0  | 1  | 1  | 1  |
| Sig. effect of physical health on comm. development    | 1  | 0  | 0  | 0  | 1  | 1  | 1  |
| <b>Physical Conditions</b>                             |    |    |    |    |    |    |    |
| Choanal atresia                                        | 1  | 1  | 0  | 1  | 1  | 0  | 1  |
| Heart problems                                         | 1  | 0  | 0  | 1  | 1  | 0  | 1  |
| Cleft lip/palate                                       | 0  | 0  | 0  | 0  | 0  | 0  | 0  |
| Swallowing/feeding problems                            | 1  | 0  | 0  | 1  | 1  | 1  | 1  |
| Facial problems                                        | 1  | 0  | 1  | 0  | 0  | 0  | 1  |
| Hypotonia                                              | 1  | 1  | 1  | 1  | 1  | 1  | 1  |
| Growth deficiency                                      | 1  | 1  | 1  | 0  | 1  | 1  | 0  |
| Tracheostomy                                           | 0  | 0  | 0  | 0  | 0  | 0  | 0  |
| Gastrostomy tube                                       | 1  | 0  | 0  | 1  | 1  | 0  | 1  |
| Neurologic problems (structural)                       | 0  | 0  | 1  | 0  | 0  | 0  | 0  |
| Neurologic problems (other)                            | 0  | 1  | 0  | 0  | 0  | 0  | 0  |
| <b>SENSORY FUNCTION</b>                                |    |    |    |    |    |    |    |
| Sig. effect of sensory function on overall development | 1  | 0  | 0  | 1  | 1  | 1  | 1  |
| Sig. effect of sensory function on comm. development   | 1  | 0  | 0  | 1  | 1  | 1  | 1  |
| <b>HEARING</b>                                         |    |    |    |    |    |    |    |
| Sig. effect of hearing on comm. development            | 1  | 1  | 1  | 1  | 1  | 1  | 1  |
| <b>Type of Hearing Loss</b>                            |    |    |    |    |    |    |    |
| Conductive loss                                        | 0  | 0  | 0  | 0  | 0  | 0  | 0  |
| Sensorineural loss                                     | 0  | 1  | 1  | 1  | 0  | 1  | 0  |
| Mixed loss                                             | 1  | 0  | 0  | 0  | 1  | 0  | 1  |
| <b>Degree of Hearing Loss</b>                          |    |    |    |    |    |    |    |
| Normal hearing                                         | 0  | 0  | 0  | 0  | 0  | 0  | 0  |
| Mild loss                                              | 0  | 1  | 1  | 0  | 0  | 0  | 0  |
| Moderate loss                                          | 0  | 0  | 0  | 0  | 0  | 1  | 0  |
| Moderately-severe loss                                 | 0  | 0  | 0  | 1  | 0  | 0  | 0  |
| Severe-profound loss                                   | 1  | 0  | 0  | 0  | 1  | 0  | 1  |
| <b>Treatment of Hearing Loss</b>                       |    |    |    |    |    |    |    |
| Uses hearing aids                                      | 0  | 0  | 0  | 1  | 1  | 1  | 0  |
| Uses cochlear implant                                  | 0  | 0  | 0  | 0  | 0  | 0  | 0  |

| Participant Number                                      | A1 | A2 | A3 | A4 | A5 | A6 | A7 |
|---------------------------------------------------------|----|----|----|----|----|----|----|
| <b><u>BALANCE</u></b>                                   |    |    |    |    |    |    |    |
| Sig. effect of balance on comm. development             | 1  | 0  | 0  | 1  | 1  | 0  | 1  |
| <b><u>Mobility</u></b>                                  |    |    |    |    |    |    |    |
| Walks independently                                     | 0  | 1  | 0  | 1  | 0  | 0  | 1  |
| Mobile with assistance                                  | 1  | 0  | 0  | 0  | 0  | 1  | 0  |
| Does not walk (uses wheelchair)                         | 0  | 0  | 1  | 0  | 1  | 0  | 0  |
| <b><u>VISION</u></b>                                    |    |    |    |    |    |    |    |
| Sig. effect of vision on comm. development              | 1  | 0  | 0  | 0  | 1  | 0  | 0  |
| Prevalence of vision problems                           | 1  | 1  | 1  | 1  | 1  | 1  | 1  |
| <b><u>SMELL</u></b>                                     |    |    |    |    |    |    |    |
| Sig. effect of smell on comm. development               | 1  | 0  | 0  | 0  | 0  | 0  | 0  |
| <b><u>TASTE</u></b>                                     |    |    |    |    |    |    |    |
| Sig. effect of taste on comm. development               | 1  | 0  | 0  | 0  | 0  | 0  | 0  |
| <b><u>BEHAVIOR</u></b>                                  |    |    |    |    |    |    |    |
| Sig. effect of behavior on overall development          | 0  | 0  | 0  | 0  | 0  | 0  | 1  |
| Sig. effect of behavior on comm. development            | 0  | 0  | 0  | 0  | 0  | 1  | 1  |
| Declined to respond                                     | 1  | 0  | 0  | 0  | 0  | 0  | 0  |
| <b><u>Behaviors</u></b>                                 |    |    |    |    |    |    |    |
| Well-adjusted                                           | 1  | 1  | 1  | 1  | 0  | 1  | 0  |
| Social or emotional immaturity                          | 1  | 1  | 1  | 1  | 1  | 1  | 1  |
| Unstable or explosive behavior                          | 0  | 1  | 0  | 1  | 0  | 0  | 1  |
| Problems with attention                                 | 0  | 1  | 1  | 0  | 0  | 1  | 1  |
| Overreacts to certain situations                        | 1  | 0  | 0  | 1  | 1  | 0  | 1  |
| Aggressive or defensive behaviors                       | 0  | 1  | 0  | 0  | 0  | 0  | 1  |
| Rhythmic rocking                                        | 0  | 0  | 0  | 0  | 0  | 0  | 1  |
| Self-stimulatory behavior                               | 0  | 1  | 1  | 1  | 1  | 1  | 1  |
| Sensitivity to loud sounds                              | 0  | 1  | 1  | 0  | 0  | 0  | 0  |
| Lack of communication for interpersonal purposes        | 0  | 1  | 1  | 1  | 1  | 0  | 1  |
| <b><u>COMMUNICATION TRAINING</u></b>                    |    |    |    |    |    |    |    |
| Sig. effect of speech & language on overall development | 1  | 1  | 1  | 1  | 1  | 1  | 1  |
| <b><u>CURRENT EDUCATIONAL PLACEMENT</u></b>             |    |    |    |    |    |    |    |
| Sig. effect of communication on educational development | 1  | 1  | 0  | 1  | 1  | 1  | 1  |
| <b><u>Educational Setting</u></b>                       |    |    |    |    |    |    |    |
| Mainstream                                              | 0  | 0  | 0  | 0  | 0  | 0  | 0  |
| Inclusive                                               | 0  | 0  | 0  | 1  | 0  | 0  | 0  |
| Self-contained (hearing impaired)                       | 0  | 0  | 1  | 0  | 0  | 0  | 1  |
| Self-contained (multiple disabilities)                  | 0  | 1  | 0  | 0  | 0  | 0  | 0  |
| Deaf school                                             | 0  | 0  | 0  | 0  | 0  | 0  | 0  |
| Early childhood program                                 | 0  | 0  | 0  | 0  | 1  | 1  | 0  |
| Home school                                             | 1  | 0  | 0  | 0  | 0  | 0  | 0  |
| Not currently enrolled                                  | 0  | 0  | 0  | 0  | 0  | 0  | 0  |

| Participant Number                                     | A8 | A9 | A10 | A11 | A12 | A13 | A14 |
|--------------------------------------------------------|----|----|-----|-----|-----|-----|-----|
| Gender                                                 | F  | M  | M   | M   | F   | F   | F   |
| Age                                                    | 3  | 11 | 5   | 2   | 4   | 11  | 7   |
| Race                                                   | W  | W  | W   | H   | W   | W   | W   |
| Parent interview: personal                             | 1  | 1  | 1   | 1   | 1   | 1   | 0   |
| Parent interview: telephone/email                      | 0  | 1  | 1   | 1   | 1   | 1   | 1   |
| Child observed                                         | 0  | 1  | 1   | 1   | 1   | 1   | 0   |
| <b>PRIMARY MODE</b>                                    |    |    |     |     |     |     |     |
| <b>OF COMMUNICATION</b>                                |    |    |     |     |     |     |     |
| Sig. effect of communication on overall development    | 0  | 1  | 0   | 1   | 0   | 1   | 1   |
| Sig. effect of CHARGE on comm. development             | 0  | 1  | 0   | 1   | 0   | 1   | 1   |
| <b>Communication Mode</b>                              |    |    |     |     |     |     |     |
| Speech                                                 | 0  | 0  | 0   | 0   | 1   | 1   | 1   |
| Sign Language                                          | 0  | 0  | 0   | 0   | 0   | 0   | 0   |
| Gesture                                                | 1  | 1  | 0   | 0   | 0   | 0   | 0   |
| Perlocutionary                                         | 0  | 0  | 1   | 1   | 0   | 0   | 0   |
| <b>PHYSICAL HEALTH</b>                                 |    |    |     |     |     |     |     |
| Sig. effect of physical health on overall development  | 0  | 1  | 1   | 1   | 1   | 1   | 1   |
| Sig. effect of physical health on comm. development    | 1  | 1  | 1   | 1   | 1   | 0   | 1   |
| <b>Physical Conditions</b>                             |    |    |     |     |     |     |     |
| Choanal atresia                                        | 0  | 1  | 1   | 1   | 1   | 1   | 1   |
| Heart problems                                         | 0  | 1  | 1   | 1   | 0   | 1   | 1   |
| Cleft lip/palate                                       | 0  | 0  | 1   | 1   | 0   | 1   | 1   |
| Swallowing/feeding problems                            | 1  | 1  | 1   | 1   | 1   | 1   | 1   |
| Facial problems                                        | 0  | 1  | 0   | 1   | 0   | 1   | 0   |
| Hypotonia                                              | 1  | 1  | 1   | 1   | 1   | 1   | 0   |
| Growth deficiency                                      | 1  | 1  | 1   | 0   | 1   | 0   | 0   |
| Tracheostomy                                           | 0  | 0  | 0   | 1   | 0   | 0   | 0   |
| Gastrostomy tube                                       | 0  | 0  | 0   | 1   | 1   | 1   | 1   |
| Neurologic problems (structural)                       | 0  | 0  | 1   | 1   | 0   | 0   | 0   |
| Neurologic problems (other)                            | 1  | 1  | 0   | 0   | 0   | 0   | 0   |
| <b>SENSORY FUNCTION</b>                                |    |    |     |     |     |     |     |
| Sig. effect of sensory function on overall development | 0  | 1  | 1   | 1   | 1   | 1   | 1   |
| Sig. effect of sensory function on comm. development   | 0  | 1  | 1   | 1   | 1   | 1   | 1   |
| <b>HEARING</b>                                         |    |    |     |     |     |     |     |
| Sig. effect of hearing on comm. development            | 0  | 0  | 0   | 1   | 0   | 1   | 1   |
| <b>Type of Hearing Loss</b>                            |    |    |     |     |     |     |     |
| Conductive loss                                        | 0  | 0  | 0   | 0   | 0   | 0   | 0   |
| Sensorineural loss                                     | 0  | 0  | 1   | 0   | 0   | 0   | 1   |
| Mixed loss                                             | 0  | 1  | 0   | 1   | 1   | 1   | 0   |
| <b>Degree of Hearing Loss</b>                          |    |    |     |     |     |     |     |
| Normal hearing                                         | 1  | 0  | 0   | 0   | 0   | 0   | 0   |
| Mild loss                                              | 0  | 0  | 1   | 0   | 1   | 0   | 0   |
| Moderate loss                                          | 0  | 1  | 0   | 1   | 0   | 0   | 0   |
| Moderately-severe loss                                 | 0  | 0  | 0   | 0   | 0   | 1   | 0   |
| Severe-profound loss                                   | 0  | 0  | 0   | 0   | 0   | 0   | 1   |
| <b>Treatment of Hearing Loss</b>                       |    |    |     |     |     |     |     |
| Uses hearing aids                                      | 0  | 0  | 0   | 0   | 1   | 1   | 0   |
| Uses cochlear implant                                  | 0  | 0  | 0   | 0   | 0   | 0   | 1   |

| Participant Number                                      | A8 | A9 | A10 | A11 | A12 | A13 | A14 |
|---------------------------------------------------------|----|----|-----|-----|-----|-----|-----|
| <b><u>BALANCE</u></b>                                   |    |    |     |     |     |     |     |
| Sig. effect of balance on comm. development             | 0  | 0  | 0   | 0   | 0   | 1   | 0   |
| <b><u>Mobility</u></b>                                  |    |    |     |     |     |     |     |
| Walks independently                                     | 0  | 0  | 0   | 0   | 1   | 1   | 1   |
| Mobile with assistance                                  | 0  | 0  | 0   | 1   | 0   | 0   | 0   |
| Does not walk (uses wheelchair)                         | 1  | 1  | 1   | 0   | 0   | 0   | 0   |
| <b><u>VISION</u></b>                                    |    |    |     |     |     |     |     |
| Sig. effect of vision on Ccomm. development             | 0  | 0  | 0   | 1   | 0   | 1   | 0   |
| Prevalence of vision problems                           | 1  | 1  | 1   | 1   | 1   | 1   | 1   |
| <b><u>SMELL</u></b>                                     |    |    |     |     |     |     |     |
| Sig. effect of smell on comm. development               | 0  | 0  | 0   | 0   | 0   | 0   | 0   |
| <b><u>TASTE</u></b>                                     |    |    |     |     |     |     |     |
| Sig. effect of taste on comm. development               | 0  | 0  | 0   | 0   | 0   | 0   | 0   |
| <b><u>BEHAVIOR</u></b>                                  |    |    |     |     |     |     |     |
| Sig. effect of behavior on overall development          | 0  | 0  | 0   | 1   | 0   | 1   | 0   |
| Sig. effect of behavior on comm. development            | 0  | 0  | 0   | 1   | 0   | 1   | 0   |
| Declined to respond                                     | 0  | 0  | 0   | 0   | 0   | 0   | 0   |
| <b><u>Behaviors</u></b>                                 |    |    |     |     |     |     |     |
| Well-adjusted                                           | 1  | 1  | 1   | 0   | 0   | 0   | 0   |
| Social or emotional immaturity                          | 0  | 0  | 1   | 1   | 1   | 1   | 1   |
| Unstable or explosive behavior                          | 0  | 0  | 0   | 1   | 0   | 1   | 0   |
| Problems with attention                                 | 1  | 1  | 1   | 1   | 0   | 1   | 1   |
| Overreacts to certain situations                        | 0  | 0  | 0   | 1   | 0   | 1   | 0   |
| Aggressive or defensive behaviors                       | 0  | 0  | 0   | 1   | 0   | 0   | 1   |
| Rhythmic rocking                                        | 1  | 1  | 0   | 1   | 0   | 0   | 0   |
| Self-stimulatory behavior                               | 0  | 1  | 0   | 1   | 1   | 1   | 1   |
| Sensitivity to loud sounds                              | 1  | 1  | 1   | 0   | 1   | 1   | 0   |
| Lack of communication for interpersonal purposes        | 0  | 0  | 0   | 1   | 0   | 0   | 0   |
| <b><u>COMMUNICATION TRAINING</u></b>                    |    |    |     |     |     |     |     |
| Sig. effect of speech & language on overall development | 1  | 1  | 1   | 1   | 0   | 1   | 1   |
| <b><u>CURRENT EDUCATIONAL PLACEMENT</u></b>             |    |    |     |     |     |     |     |
| Sig. effect of communication on educational development | 1  | 1  | 1   | 1   | 0   | 1   | 1   |
| <b><u>Educational Setting</u></b>                       |    |    |     |     |     |     |     |
| Mainstream                                              | 0  | 0  | 0   | 0   | 0   | 0   | 1   |
| Inclusive                                               | 0  | 0  | 0   | 0   | 1   | 0   | 0   |
| Self-contained (hearing impaired)                       | 0  | 1  | 0   | 0   | 0   | 0   | 0   |
| Self-contained (multiple disabilities)                  | 0  | 0  | 0   | 0   | 0   | 0   | 0   |
| Deaf school                                             | 0  | 0  | 0   | 0   | 0   | 1   | 0   |
| Early childhood program                                 | 0  | 0  | 0   | 0   | 0   | 0   | 0   |
| Home school                                             | 0  | 0  | 1   | 1   | 0   | 0   | 0   |
| Not currently enrolled                                  | 1  | 0  | 0   | 0   | 0   | 0   | 0   |

| Participant Number                                     | A15 | A16 | A17 | A18 | A19 | A20 | A21 |
|--------------------------------------------------------|-----|-----|-----|-----|-----|-----|-----|
| Gender                                                 | F   | M   | F   | F   | M   | F   | F   |
| Age                                                    | 7   | 10  | 8   | 5   | 7   | 1   | 27  |
| Race                                                   | W   | W   | W   | W   | W   | W   | W   |
| Parent interview: personal                             | 1   | 1   | 1   | 1   | 1   | 1   | 0   |
| Parent interview: telephone/email                      | 1   | 1   | 1   | 1   | 1   | 1   | 1   |
| Child observed                                         | 1   | 1   | 1   | 1   | 0   | 1   | 0   |
| <b>PRIMARY MODE</b>                                    |     |     |     |     |     |     |     |
| <b>OF COMMUNICATION</b>                                |     |     |     |     |     |     |     |
| Sig. effect of communication on overall development    | 0   | 1   | 1   | 1   | 1   | 1   | 1   |
| Sig. effect of CHARGE on comm. development             | 1   | 1   | 1   | 1   | 1   | 1   | 1   |
| <b>Communication Mode</b>                              |     |     |     |     |     |     |     |
| Speech                                                 | 1   | 1   | 1   | 1   | 1   | 1*  | 0   |
| Sign Language                                          | 0   | 0   | 0   | 0   | 0   | 0   | 1   |
| Gesture                                                | 0   | 0   | 0   | 0   | 0   | 0   | 0   |
| Perlocutionary                                         | 0   | 0   | 0   | 0   | 0   | 0   | 0   |
| <b>PHYSICAL HEALTH</b>                                 |     |     |     |     |     |     |     |
| Sig. effect of physical health on overall development  | 1   | 1   | 0   | 1   | 1   | 1   | 1   |
| Sig. effect of physical health on comm. development    | 1   | 1   | 0   | 1   | 0   | 1   | 1   |
| <b>Physical Conditions</b>                             |     |     |     |     |     |     |     |
| Choanal atresia                                        | 0   | 1   | 0   | 1   | 1   | 1   | 0   |
| Heart problems                                         | 0   | 1   | 0   | 1   | 1   | 1   | 1   |
| Cleft lip/palate                                       | 0   | 0   | 1   | 0   | 0   | 0   | 0   |
| Swallowing/feeding problems                            | 1   | 1   | 1   | 1   | 1   | 1   | 0   |
| Facial problems                                        | 1   | 1   | 0   | 0   | 1   | 0   | 1   |
| Hypotonia                                              | 1   | 1   | 0   | 1   | 0   | 0   | 1   |
| Growth deficiency                                      | 0   | 1   | 0   | 0   | 0   | 0   | 1   |
| Tracheostomy                                           | 0   | 0   | 0   | 0   | 0   | 1   | 0   |
| Gastrostomy tube                                       | 1   | 0   | 0   | 1   | 1   | 1   | 0   |
| Neurologic problems (structural)                       | 0   | 0   | 0   | 0   | 0   | 0   | 0   |
| Neurologic problems (other)                            | 0   | 0   | 0   | 0   | 0   | 0   | 1   |
| <b>SENSORY FUNCTION</b>                                |     |     |     |     |     |     |     |
| Sig. effect of sensory function on overall development | 1   | 1   | 1   | 1   | 0   | 0   | 1   |
| Sig. effect of sensory function on comm. development   | 1   | 1   | 1   | 1   | 0   | 0   | 0   |
| <b>HEARING</b>                                         |     |     |     |     |     |     |     |
| Sig. effect of hearing on comm. development            | 1   | 1   | 1   | 1   | 1   | 0   | 0   |
| <b>Type of Hearing Loss</b>                            |     |     |     |     |     |     |     |
| Conductive loss                                        | 0   | 0   | 0   | 0   | 0   | 0   | 0   |
| Sensorineural loss                                     | 1   | 0   | 0   | 0   | 1   | 0   | 0   |
| Mixed loss                                             | 0   | 1   | 1   | 1   | 0   | 0   | 1   |
| <b>Degree of Hearing Loss</b>                          |     |     |     |     |     |     |     |
| Normal hearing                                         | 0   | 0   | 0   | 0   | 0   | 1   | 0   |
| Mild loss                                              | 0   | 1   | 1   | 1   | 0   | 0   | 1   |
| Moderate loss                                          | 0   | 0   | 0   | 0   | 0   | 0   | 0   |
| Moderately-severe loss                                 | 1   | 0   | 0   | 0   | 0   | 0   | 0   |
| Severe-profound loss                                   | 0   | 0   | 0   | 0   | 1   | 0   | 0   |
| <b>Treatment of Hearing Loss</b>                       |     |     |     |     |     |     |     |
| Uses hearing aids                                      | 1   | 1   | 1   | 1   | 1   | 0   | 0   |
| Uses cochlear implant                                  | 0   | 0   | 0   | 0   | 0   | 0   | 0   |

| Participant Number                                      | A15 | A16 | A17 | A18 | A19 | A20 | A21 |
|---------------------------------------------------------|-----|-----|-----|-----|-----|-----|-----|
| <b><u>BALANCE</u></b>                                   |     |     |     |     |     |     |     |
| Sig. effect of balance on comm. development             | 0   | 1   | 0   | 1   | 0   | 0   | 0   |
| <b><u>Mobility</u></b>                                  |     |     |     |     |     |     |     |
| Walks independently                                     | 1   | 1   | 1   | 0   | 1   | 0   | 1   |
| Mobile with assistance                                  | 0   | 0   | 0   | 1   | 0   | 1   | 0   |
| Does not walk (uses wheelchair)                         | 0   | 0   | 0   | 0   | 0   | 0   | 0   |
| <b><u>VISION</u></b>                                    |     |     |     |     |     |     |     |
| Sig. effect of vision on Ccmm. development              | 0   | 0   | 1   | 1   | 0   | 0   | 1   |
| Prevalence of vision problems                           | 1   | 1   | 1   | 1   | 1   | 1   | 1   |
| <b><u>SMELL</u></b>                                     |     |     |     |     |     |     |     |
| Sig. effect of smell on comm. development               | 0   | 0   | 0   | 0   | 0   | 0   | 0   |
| <b><u>TASTE</u></b>                                     |     |     |     |     |     |     |     |
| Sig. effect of taste on comm. development               | 0   | 0   | 0   | 0   | 0   | 0   | 0   |
| <b><u>BEHAVIOR</u></b>                                  |     |     |     |     |     |     |     |
| Sig. effect of behavior on overall development          | 0   | 1   | 1   | 0   | 0   | 0   | 0   |
| Sig. effect of behavior on comm. development            | 0   | 1   | 0   | 0   | 0   | 0   | 0   |
| Declined to respond                                     | 0   | 0   | 0   | 1   | 1   | 0   | 1   |
| <b><u>Behaviors</u></b>                                 |     |     |     |     |     |     |     |
| Well-adjusted                                           | 1   | 0   | 0   | 1   | 1   | 0   | 0   |
| Social or emotional immaturity                          | 0   | 1   | 1   | 1   | 1   | 0   | 1   |
| Unstable or explosive behavior                          | 0   | 0   | 1   | 0   | 0   | 0   | 0   |
| Problems with attention                                 | 0   | 1   | 1   | 1   | 0   | 0   | 1   |
| Overreacts to certain situations                        | 0   | 1   | 1   | 1   | 0   | 0   | 1   |
| Aggressive or defensive behaviors                       | 0   | 1   | 1   | 1   | 0   | 0   | 1   |
| Rhythmic rocking                                        | 0   | 0   | 0   | 0   | 0   | 0   | 0   |
| Self-stimulatory behavior                               | 0   | 0   | 0   | 0   | 1   | 0   | 1   |
| Sensitivity to loud sounds                              | 0   | 0   | 1   | 0   | 1   | 0   | 1   |
| Lack of communication for interpersonal purposes        | 0   | 0   | 1   | 1   | 1   | 0   | 0   |
| <b><u>COMMUNICATION TRAINING</u></b>                    |     |     |     |     |     |     |     |
| Sig. effect of speech & language on overall development | 1   | 1   | 1   | 1   | 1   | 1   | 1   |
| <b><u>CURRENT EDUCATIONAL PLACEMENT</u></b>             |     |     |     |     |     |     |     |
| Sig. effect of communication on educational development | 0   | 1   | 1   | 1   | 1   | 1   | 1   |
| <b><u>Educational Setting</u></b>                       |     |     |     |     |     |     |     |
| Mainstream                                              | 0   | 1   | 1   | 1   | 0   | 0   | 0   |
| Inclusive                                               | 0   | 0   | 0   | 0   | 1   | 0   | 0   |
| Self-contained (hearing impaired)                       | 0   | 0   | 0   | 0   | 0   | 0   | 0   |
| Self-contained (multiple disabilities)                  | 0   | 0   | 0   | 0   | 0   | 0   | 0   |
| Deaf school                                             | 0   | 0   | 0   | 0   | 0   | 0   | 0   |
| Early childhood program                                 | 0   | 0   | 0   | 0   | 0   | 0   | 0   |
| Home school                                             | 1   | 0   | 0   | 0   | 0   | 0   | 0   |
| Not currently enrolled                                  | 0   | 0   | 0   | 0   | 0   | 1   | 1   |

| Participant Number                                     | A22 | A23 | A24 | A25 |
|--------------------------------------------------------|-----|-----|-----|-----|
| Gender                                                 | M   | F   | M   | F   |
| Age                                                    | 11  | 9   | 8   | 6   |
| Race                                                   | W   | W   | W   | W   |
| Parent interview: personal                             | 1   | 1   | 1   | 1   |
| Parent interview: telephone/email                      | 1   | 1   | 1   | 1   |
| Child observed                                         | 0   | 1   | 1   | 1   |
| <b>PRIMARY MODE OF COMMUNICATION</b>                   |     |     |     |     |
| Sig. effect of communication on overall development    | 1   | 1   | 0   | 1   |
| Sig. effect of CHARGE on comm. development             | 1   | 1   | 1   | 1   |
| <b>Communication Mode</b>                              |     |     |     |     |
| Speech                                                 | 0   | 0   | 0   | 0   |
| Sign Language                                          | 1   | 1   | 1   | 1   |
| Gesture                                                | 0   | 0   | 0   | 0   |
| Perlocutionary                                         | 0   | 0   | 0   | 0   |
| <b>PHYSICAL HEALTH</b>                                 |     |     |     |     |
| Sig. effect of physical health on overall development  | 1   | 1   | 1   | 1   |
| Sig. effect of physical health on comm. development    | 1   | 1   | 1   | 1   |
| <b>Physical Conditions</b>                             |     |     |     |     |
| Choanal atresia                                        | 1   | 1   | 1   | 0   |
| Heart problems                                         | 1   | 1   | 1   | 1   |
| Cleft lip/palate                                       | 1   | 0   | 0   | 1   |
| Swallowing/feeding problems                            | 1   | 1   | 1   | 1   |
| Facial problems                                        | 1   | 1   | 1   | 1   |
| Hypotonia                                              | 1   | 1   | 1   | 1   |
| Growth deficiency                                      | 1   | 1   | 0   | 1   |
| Tracheostomy                                           | 0   | 0   | 0   | 0   |
| Gastrostomy tube                                       | 0   | 0   | 1   | 1   |
| Neurologic problems (structural)                       | 0   | 0   | 1   | 0   |
| Neurologic problems (other)                            | 0   | 0   | 0   | 0   |
| <b>SENSORY FUNCTION</b>                                |     |     |     |     |
| Sig. effect of sensory function on overall development | 1   | 1   | 1   | 1   |
| Sig. effect of sensory function on comm. development   | 1   | 1   | 1   | 1   |
| <b>HEARING</b>                                         |     |     |     |     |
| Sig. effect of hearing on comm. development            | 1   | 1   | 1   | 1   |
| <b>Type of Hearing Loss</b>                            |     |     |     |     |
| Conductive loss                                        | 0   | 0   | 0   | 0   |
| Sensorineural loss                                     | 0   | 0   | 1   | 0   |
| Mixed loss                                             | 1   | 1   | 0   | 1   |
| <b>Degree of Hearing Loss</b>                          |     |     |     |     |
| Normal hearing                                         | 0   | 0   | 0   | 0   |
| Mild loss                                              | 0   | 0   | 0   | 0   |
| Moderate loss                                          | 0   | 0   | 0   | 0   |
| Moderately-severe loss                                 | 0   | 0   | 0   | 1   |
| Severe-profound loss                                   | 1   | 1   | 1   | 0   |
| <b>Treatment of Hearing Loss</b>                       |     |     |     |     |
| Uses hearing aids                                      | 1   | 0   | 0   | 1   |
| Uses cochlear implant                                  | 0   | 0   | 0   | 0   |

| Participant Number                                      | A22 | A23 | A24 | A25 |
|---------------------------------------------------------|-----|-----|-----|-----|
| <b><u>BALANCE</u></b>                                   |     |     |     |     |
| Sig. effect of balance on comm. development             | 0   | 1   | 1   | 1   |
| <b><u>Mobility</u></b>                                  |     |     |     |     |
| Walks independently                                     | 1   | 1   | 1   | 1   |
| Mobile with assistance                                  | 0   | 0   | 0   | 0   |
| Does not walk (uses wheelchair)                         | 0   | 0   | 0   | 0   |
| <b><u>VISION</u></b>                                    |     |     |     |     |
| Sig. effect of vision on Cmm. development               | 1   | 0   | 1   | 1   |
| Prevalence of vision problems                           | 1   | 1   | 1   | 1   |
| <b><u>SMELL</u></b>                                     |     |     |     |     |
| Sig. effect of smell on comm. development               | 1   | 0   | 0   | 1   |
| <b><u>TASTE</u></b>                                     |     |     |     |     |
| Sig. effect of taste on comm. development               | 1   | 0   | 0   | 0   |
| <b><u>BEHAVIOR</u></b>                                  |     |     |     |     |
| Sig. effect of behavior on overall development          | 0   | 1   | 0   | 1   |
| Sig. effect of behavior on comm. development            | 1   | 1   | 0   | 1   |
| Declined to respond                                     | 0   | 0   | 1   | 0   |
| <b><u>Behaviors</u></b>                                 |     |     |     |     |
| Well-adjusted                                           | 1   | 1   | 1   | 0   |
| Social or emotional immaturity                          | 0   | 1   | 1   | 1   |
| Unstable or explosive behavior                          | 0   | 0   | 0   | 0   |
| Problems with attention                                 | 1   | 1   | 0   | 1   |
| Overreacts to certain situations                        | 0   | 1   | 0   | 0   |
| Aggressive or defensive behaviors                       | 1   | 1   | 0   | 1   |
| Rhythmic rocking                                        | 0   | 0   | 0   | 0   |
| Self-stimulatory behavior                               | 0   | 0   | 0   | 1   |
| Sensitivity to loud sounds                              | 0   | 0   | 0   | 0   |
| Lack of communication for interpersonal purposes        | 1   | 0   | 0   | 0   |
| <b><u>COMMUNICATION TRAINING</u></b>                    |     |     |     |     |
| Sig. effect of speech & language on overall development | 1   | 1   | 1   | 1   |
| <b><u>CURRENT EDUCATIONAL PLACEMENT</u></b>             |     |     |     |     |
| Sig. effect of communication on educational development | 1   | 1   | 1   | 1   |
| <b><u>Educational Setting</u></b>                       |     |     |     |     |
| Mainstream                                              | 0   | 0   | 0   | 0   |
| Inclusive                                               | 0   | 0   | 0   | 0   |
| Self-contained (hearing impaired)                       | 1   | 0   | 0   | 1   |
| Self-contained (multiple disabilities)                  | 0   | 0   | 0   | 0   |
| Deaf school                                             | 0   | 1   | 0   | 0   |
| Early childhood program                                 | 0   | 0   | 0   | 0   |
| Home school                                             | 0   | 0   | 1   | 0   |
| Not currently enrolled                                  | 0   | 0   | 0   | 0   |

| <b>Participant Number</b>                              | <b>C1</b> | <b>C2</b> | <b>C3</b> | <b>C4</b> | <b>C5</b> | <b>C6</b> |
|--------------------------------------------------------|-----------|-----------|-----------|-----------|-----------|-----------|
| <b>Gender</b>                                          | M         | F         | F         | M         | F         | M         |
| <b>Age</b>                                             | 4         | 12        | 4         | 5         | 11        | 7         |
| <b>Race</b>                                            | W         | W         | W         | W         | W         | W         |
| <b>Parent interview: personal</b>                      | 0         | 0         | 0         | 0         | 0         | 0         |
| <b>Parent interview: telephone/email</b>               | 1         | 1         | 1         | 1         | 1         | 1         |
| <b>Child observed</b>                                  | 0         | 0         | 0         | 0         | 0         | 0         |
| <b><u>PRIMARY MODE OF COMMUNICATION</u></b>            |           |           |           |           |           |           |
| Sig. effect of communication on overall development    | 0         | 0         | 0         | 1         | 1         | 1         |
| Sig. effect of CHARGE on comm. development             | 1         | 1         | 1         | 1         | 1         | 1         |
| <b><u>Communication Mode</u></b>                       |           |           |           |           |           |           |
| Speech                                                 | 1         | 1         | 1         | 0         | 0         | 0         |
| Sign Language                                          | 0         | 0         | 0         | 1         | 0         | 0         |
| Gesture                                                | 0         | 0         | 0         | 0         | 1         | 1         |
| Perlocutionary                                         | 0         | 0         | 0         | 0         | 0         | 0         |
| <b><u>PHYSICAL HEALTH</u></b>                          |           |           |           |           |           |           |
| Sig. effect of physical health on overall development  | 1         | 0         | 0         | 1         | 1         | 1         |
| Sig. effect of physical health on comm. development    | 1         | 0         | 1         | 1         | 1         | 1         |
| <b><u>Physical Conditions</u></b>                      |           |           |           |           |           |           |
| Choanal atresia                                        | 1         | 1         | 1         | 0         | 1         | 1         |
| Heart problems                                         | 1         | 1         | 1         | 0         | 1         | 1         |
| Cleft lip/palate                                       | 0         | 0         | 0         | 1         | 1         | 1         |
| Swallowing/feeding problems                            | 1         | 0         | 1         | 1         | 1         | 1         |
| Facial problems                                        | 0         | 0         | 1         | 1         | 0         | 1         |
| Hypotonia                                              | 1         | 1         | 0         | 0         | 1         | 1         |
| Growth deficiency                                      | 1         | 0         | 0         | 0         | 1         | 1         |
| Tracheostomy                                           | 0         | 0         | 0         | 0         | 0         | 0         |
| Gastrostomy tube                                       | 0         | 0         | 1         | 1         | 1         | 1         |
| Neurologic problems (structural)                       | 0         | 1         | 0         | 0         | 0         | 0         |
| Neurologic problems (other)                            | 0         | 0         | 0         | 0         | 0         | 1         |
| <b><u>SENSORY FUNCTION</u></b>                         |           |           |           |           |           |           |
| Sig. effect of sensory function on overall development | 1         | 0         | 1         | 1         | 1         | 1         |
| Sig. effect of sensory function on comm. development   | 1         | 0         | 1         | 1         | 1         | 1         |
| <b><u>HEARING</u></b>                                  |           |           |           |           |           |           |
| Sig. effect of hearing on comm. development            | 1         | 1         | 0         | 1         | 1         | 1         |
| <b><u>Type of Hearing Loss</u></b>                     |           |           |           |           |           |           |
| Conductive loss                                        | 0         | 0         | 0         | 0         | 0         | 0         |
| Sensorineural loss                                     | 0         | 1         | 0         | 0         | 1         | 0         |
| Mixed loss                                             | 1         | 0         | 1         | 1         | 0         | 1         |
| <b><u>Degree of Hearing Loss</u></b>                   |           |           |           |           |           |           |
| Normal hearing                                         | 0         | 0         | 0         | 0         | 0         | 0         |
| Mild loss                                              | 0         | 1         | 0         | 0         | 0         | 0         |
| Moderate loss                                          | 1         | 0         | 0         | 0         | 1         | 0         |
| Moderately-severe loss                                 | 0         | 0         | 0         | 1         | 0         | 1         |
| Severe-profound loss                                   | 0         | 0         | 1         | 0         | 0         | 0         |
| <b><u>Treatment of Hearing Loss</u></b>                |           |           |           |           |           |           |
| Uses hearing aids                                      | 0         | 1         | 1         | 1         | 1         | 1         |
| Uses cochlear implant                                  | 0         | 0         | 0         | 0         | 0         | 0         |

| Participant Number                                      | C1 | C2 | C3 | C4 | C5 | C6 |
|---------------------------------------------------------|----|----|----|----|----|----|
| <b><u>BALANCE</u></b>                                   |    |    |    |    |    |    |
| Sig. effect of balance on comm. development             | 1  | 0  | 0  | 1  | 1  | 0  |
| <b><u>Mobility</u></b>                                  |    |    |    |    |    |    |
| Walks independently                                     | 1  | 1  | 1  | 1  | 0  | 0  |
| Mobile with assistance                                  | 0  | 0  | 0  | 0  | 1  | 1  |
| Does not walk (uses wheelchair)                         | 0  | 0  | 0  | 0  | 0  | 0  |
| <b><u>VISION</u></b>                                    |    |    |    |    |    |    |
| Sig. effect of vision on Cmm. development               | 0  | 0  | 0  | 1  | 1  | 1  |
| Prevalence of vision problems                           | 1  | 1  | 1  | 1  | 1  | 1  |
| <b><u>SMELL</u></b>                                     |    |    |    |    |    |    |
| Sig. effect of smell on comm. development               | 0  | 0  | 0  | 0  | 0  | 0  |
| <b><u>TASTE</u></b>                                     |    |    |    |    |    |    |
| Sig. effect of taste on comm. development               | 0  | 0  | 0  | 1  | 1  | 0  |
| <b><u>BEHAVIOR</u></b>                                  |    |    |    |    |    |    |
| Sig. effect of behavior on overall development          | 0  | 1  | 0  | 0  | 1  | 0  |
| Sig. effect of behavior on comm. development            | 0  | 0  | 0  | 0  | 1  | 0  |
| Declined to respond                                     | 0  | 0  | 0  | 1  | 0  | 1  |
| <b><u>Behaviors</u></b>                                 |    |    |    |    |    |    |
| Well-adjusted                                           | 1  | 0  | 0  | 0  | 1  | 0  |
| Social or emotional immaturity                          | 1  | 1  | 1  | 1  | 1  | 1  |
| Unstable or explosive behavior                          | 0  | 0  | 1  | 1  | 1  | 1  |
| Problems with attention                                 | 0  | 1  | 1  | 0  | 1  | 1  |
| Overreacts to certain situations                        | 0  | 0  | 0  | 1  | 0  | 1  |
| Aggressive or defensive behaviors                       | 0  | 0  | 1  | 0  | 1  | 1  |
| Rhythmic rocking                                        | 0  | 0  | 0  | 0  | 0  | 1  |
| Self-stimulatory behavior                               | 0  | 1  | 1  | 1  | 1  | 1  |
| Sensitivity to loud sounds                              | 1  | 1  | 0  | 1  | 1  | 1  |
| Lack of communication for interpersonal purposes        | 0  | 1  | 0  | 1  | 1  | 1  |
| <b><u>COMMUNICATION TRAINING</u></b>                    |    |    |    |    |    |    |
| Sig. effect of speech & language on overall development | 1  | 1  | 0  | 1  | 1  | 1  |
| <b><u>CURRENT EDUCATIONAL PLACEMENT</u></b>             |    |    |    |    |    |    |
| Sig. effect of communication on educational development | 1  | 0  | 0  | 1  | 1  | 1  |
| <b><u>Educational Setting</u></b>                       |    |    |    |    |    |    |
| Mainstream                                              | 0  | 0  | 0  | 0  | 0  | 0  |
| Inclusive                                               | 1  | 0  | 1  | 1  | 0  | 1  |
| Self-contained (hearing impaired)                       | 0  | 0  | 0  | 0  | 0  | 0  |
| Self-contained (multiple disabilities)                  | 0  | 1  | 0  | 0  | 1  | 0  |
| Deaf school                                             | 0  | 0  | 0  | 0  | 0  | 0  |
| Early childhood Program                                 | 0  | 0  | 0  | 0  | 0  | 0  |
| Home school                                             | 0  | 0  | 0  | 0  | 0  | 0  |
| Not currently enrolled                                  | 0  | 0  | 0  | 0  | 0  | 0  |

## VITA

Jill Celeste Fussner was born on August 3, 1978 in St. Petersburg, Florida. She was raised in Seminole, Florida and is a graduate of Seminole High School. She attended the University of Florida in Gainesville and graduated in May of 2000 with a Bachelor of Arts degree in Communication Sciences and Disorders. Jill will graduate in May of 2003 from the University of Tennessee, Knoxville with a Master of Arts degree in Audiology.