I, Katherine Snell, hereby submit this work as part of the requirements for the degree of:
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Receptive and Expressive Language Outcomes of Children with Cochlear Implants and CHARGE Syndrome

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Receptive and Expressive Language Outcomes of Children with Cochlear Implants and CHARGE Syndrome

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ABSTRACT

This study examined the receptive and expressive language benefits of cochlear implantation of children with CHARGE syndrome. CHARGE syndrome involves multiple congenital abnormalities including deafness and blindness. Four children exhibiting co-occurring cochlear implants and CHARGE syndrome were evaluated to determine the amount of language gained since implantation. Three of the four children made gains in receptive communication skills. Expressive communication post implantation increased in most cases, but the children did not necessarily develop functional verbal communication. Due to limitations of the study, it is not known whether the increase in receptive and expressive communication is due to therapy intervention, growth and development, or the cochlear implant. This study assisted in collecting baseline data for a prospective study.
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CHAPTER I.
INTRODUCTION

Communication in children with CHARGE syndrome is often delayed in comparison to their typically developing peers. One of the leading reasons for this delay in communication is the hearing loss commonly associated with CHARGE syndrome. Many research studies show that having a hearing loss has a direct effect on an individual’s communication development (Carney & Moeller, 1998). Children with permanent hearing loss that is rated moderate to severe/profound are at risk for deficits in the areas of language, speech, academia, and social-emotional development (Joint Committee on Infant Hearing, 2007). According to the 2007 position statement by the Joint Committee on Infant Hearing, research studies have shown that children who are identified as having a hearing loss through early detection using newborn infant hearing screenings perform 20-40 percent higher in areas of communication such as articulation and intelligibility. The Joint Committee on Infant Hearing has mandated that all children who are identified as having a hearing loss receive treatment at no later than 6 months of age and parents/caregivers be informed of the benefits of hearing aids, assistive devices, and cochlear implants in order to determine the most appropriate course of treatment. The Joint Committee on Infant Hearing also states that children should be considered for cochlear implantation regardless of the presence of a developmental disability. This statement was created because research is beginning to demonstrate that cochlear implants are beneficial to language development and allow deaf children with a developmental disability to connect with the hearing world.
Because hearing and language are so closely related, the question has arisen as to how large of an impact hearing loss has on a child with CHARGE syndrome’s communication, and how much of the language deficit is due to developmental delay or other common deficits associated with the syndrome.

It is important to determine the impact of hearing loss on communication in children with developmental disabilities because approximately 30-40% of children with sensorineural hearing loss also have an additional disability (Holden-Pitt & Diaz, 1998; Wiley, Meinzen-Derr, Choo, 2004). Cochlear implants (CI) have become a common course of treatment for hearing loss in children with severe to profound deafness. More recently, a larger number of children with developmental disabilities and hearing loss have received a cochlear implant as a course of treatment (Wiley, Jahnke, Meinzen-Derr, Choo, 2005). Little is known about the true outcomes of CI and language development in children with developmental disabilities. There have been several studies researching the advantages of CI with typically developing children, but few as of yet that determine advantages in language acquisition and functioning for children with developmental disabilities and CI. Because CI is becoming such a remarkable treatment option for children with hearing loss, it is crucial for research to be done in the areas of communication and language acquisition to document the effectiveness of cochlear implants as treatment.

CHARGE syndrome

CHARGE syndrome (also called CHARGE Association) was first discovered as a group of non-random anomalies (Hall, 1979). It was later named as a syndrome in 1981 by Pagon, Graham, Zonana, and Yong. CHARGE syndrome was originally named as an
acronym for the characteristic anomalies associated with the syndrome: coloboma of the eye, heart defect, atresia choanae, retarded growth and development, genital hypoplasia, and ear anomalies with or without hearing loss (Blake and Prasad, 2006).

The criterion for diagnosing CHARGE syndrome was reassessed in 1998 and remains current to date (Blake and Prasad, 2006). CHARGE syndrome is now diagnosed by a medical doctor based on criteria of four major categories (categories that are uncommon in syndromes other than CHARGE). The categories include: ocular coloboma, choanal atresia/stenosis, cranial nerve anomalies, and ear abnormalities. Ten minor diagnostic categories also exist. These are categories that are common in other syndromes as well as CHARGE, such as cardiovascular malformations, genital hypoplasia, cleft lip/palate, distinctive CHARGE facies (sloping forehead, flattened tip of the nose), growth deficiency, Tracheo-esophageal fistulas, and developmental delay (Blake and Prasad, 2006; Lalani, et al, 2006). An Ocular Coloboma is a slot found in the iris, retina or optic nerve which causes decreased vision. Choanal Atresia is blocking of the nasal passages often requiring immediate surgery and a tracheostomy tube. DiGeorge Sequence, omphalocele, hemivertebra and scoliosis are also commonly associated with the syndrome (Hefner & Davenport, 2006).

In order to obtain a diagnosis of CHARGE syndrome, the child must display all four major categories or three major and three minor categories. Children who display one or two major diagnostic characteristics and several minor characteristics are said to possibly have CHARGE syndrome (Lalani, et al, 2006). Another major diagnostic criterion for CHARGE syndrome is the involvement of cranial nerves. If the Olfactory nerve (CN I) is involved, the child is likely to have anosmia. If CN VII or the facial nerve
is involved, the child is likely to have facial palsy. If the Glossopharyngeal (CN IX), Vagus (CN X), or Spinal Accessory (CN XI) nerves are involved, the child will have difficulty with swallowing, gastroesophageal reflux, and velopharyngeal aspiration. If CN VIII, the auditory nerve, is involved it will cause sensorineural hearing loss (Blake and Prasad, 2006). Common ear anomalies associated with the syndrome are Mondini Malformation or semicircular canal dysfunction/hypoplasia. Eustachian tube dysfunction may also be present. These anomalies lead to hearing loss as well as vestibular deficits (Lalani, et al, 2006).

All of the malformations associated with CHARGE syndrome occur early in the first trimester of pregnancy (Blake & Prasad, 2006). Associated complications with CHARGE syndrome include renal, spinal, hand and neck/shoulder anomalies. The prognosis for life becomes much lower if the child has cyanotic cardiac lesions, bilateral posterior choanal atresia, or tracheoesophageal fistula. There is no cure for CHARGE syndrome. Reconstructive surgeries for structural abnormalities, steroids for reproductive organs, and speech, vision and hearing therapies are common treatment options. The primary concern that begins when the child is born and continues until early years of childhood is survival; communication and other developmental milestones are placed secondary to hospital visitations for various surgeries required for survival (Thelin and Fussner, 2005). Pharyngeal and laryngeal incoordination puts the child at the most risk for hospitalization and death because of aspiration and penetration of boluses. Many children with CHARGE syndrome have compromised airways and have tracheostomy tubes (Blake & Prasad, 2006).
CHARGE syndrome has been found to be a result of a defect in the DNA binding protein gene CHD7. This gene is involved in development in many early embryologic stages and cell cycle development (Lalani, et al, 2006). This is why CHARGE syndrome is a highly involved syndrome having a multitude of characteristics. The majority of children with CHARGE syndrome are sporadic cases (Edwards, Kileny, & Van Riper, 2002). The incidence of CHARGE syndrome is thought to be 1/11,900 births and seen more commonly in females. CHARGE syndrome typically occurs randomly but can have a 1-2% recurrence rate. A high percentage of monozygotic twins are born with CHARGE syndrome, thus supporting the idea of a genetic component to the syndrome (Vissners, et al, 2004). The risk of having a child with CHARGE syndrome if either parent/caregiver has the diagnosis is much larger (Edwards, Van Riper & Kileny, 1995).

Children with CHARGE syndrome often have low adaptive behavior skills which may affect their ability to modify behaviors in particular situations. They also commonly have motor impairments which may require accommodations in the classroom and in various other settings. These motor impairments are likely due to vestibular dysfunction and increased hospitalization during crucial periods of development (Hefner & Davenport, 2006). Children with CHARGE syndrome commonly show characteristics like that of a child with autism spectrum disorder. Children with CHARGE syndrome have difficulty with arousal and self-regulation. As a result of these deficits, behavioral difficulties are typically seen. The extent to which these difficulties develop depends on the individual child. Thus, the wide varieties of characteristic combinations seen in CHARGE syndrome make it difficult to classify just one behavioral phenotype for the syndrome (Blake & Prasad, 2006). Factors that contribute to an “easier” adolescence
have been found to be: degree of visual impairment, degree of cognitive functioning and age of walking. The average age children with CHARGE syndrome begin walking is between three-four years, but this is dependant on the severity of their characteristics (Hefner & Davenport, 2006).

Cochlear Implantation

Ongoing debates have surrounded the necessity of CI’s for as long as they have been in existence. More recently, cochlear implants have become more accepted as a form of treatment for children with hearing loss particularly with hearing parents/caregivers. In 1995, it was determined that over 12,000 individuals with CI had shown improvements in auditory perception. According to Mosheim (2006), the degree of success in symbolic language development with the aid of a cochlear implant depends on the cognitive and developmental levels of the child, but having exposure to communication training and therapy at an early age will give them the amount of exposure that they need.

A statement set forth by the NIH Consensus (Cochlear Implants in Adults and Children, 1995) determined that CI’s improve the hearing abilities of individuals with severe-profound hearing loss given that the individual who receives the CI fits a certain criteria. Several components are crucial to determining a candidate’s eligibility for cochlear implantation. Children must be at least 12 months old in order to be implanted. It is recommended that children with severe to profound hearing loss be implanted by 24 months old (Holt & Kirk, 2005). The etiology of the hearing loss is significant because it may help to establish several other reasons that have lead to the candidate’s present hearing capabilities. For parents who have chosen to use an oral communication approach
(versus sign language), the age of onset of deafness plays a role in determining the child’s language abilities as they stand without the aid of a CI. Duration of hearing loss is also a factor as it can directly affect the child’s exposure to language and language development. Environment and motivation also contributes to language development and will influence an individual’s candidacy for implantation (Niparko & Blankenhorn, 2003). Any residual hearing present may exclude a person with a hearing deficit from being eligible for cochlear implantation.

**Developmental Disabilities and Cochlear Implantation**

Research is beginning to develop regarding the benefits of cochlear implantation in populations with multiple disabilities. Waltzman, Scalchunes, & Cohen (2000) found that children with multiple disabilities did benefit from having a cochlear implant and that children should not be excluded from cochlear implant candidacy. A study conducted by Wiley, Meinzen-Derr and Choo (2004), determined through parent/caregiver questionnaire that children with multiple disabilities and a cochlear implant received some benefit to their communication abilities as a result of the implantation. The study states that several children who were given cochlear implants early in life were later diagnosed with a developmental disability. Thus, it is important for parents/caregivers to be aware of their options and make informed decisions regarding their child’s care. It is necessary to give a child with a developmental disability that could benefit from a cochlear implant the chance to do so, as gains in communication are possible. Wiley, et al, (2005), found that 96% of the children with developmental disabilities who participated in the cochlear implant study had increased awareness to
environmental sounds, 88% developed speaking skills, 69% had improved interaction with peers, and 88% were more likely to communicate their wants and needs.

**CHARGE syndrome and Cochlear Implantation**

In CHARGE syndrome, documented hearing loss is prevalent. Of the documented cases of CHARGE in one particular study, 13% of children have been found to have normal hearing, 7% have mild loss, 38% have a moderate hearing loss and 42% have severe-profound hearing loss (Mosheim, 2006). Because the syndrome is multi-sensory, early intervention services for hearing are crucial for a child with CHARGE syndrome’s ability to relate to the world around him (Edwards, Kileny, & Van Riper, 2002). Characteristics vary so widely among cases; therefore, it is necessary to examine all possible deficits and to test for hearing loss, even if it seems the child has normal hearing. Otitis Media with effusion can cause temporary hearing loss, so monitoring for this is crucial. In this study by Edwards, et al. (2002) it was determined that an audiologic evaluation should be conducted at one of the child’s early hospitalizations to determine a further course of action. Because these children are so medically fragile, it is crucial to develop a management plan through a multidisciplinary approach to limit time spent in the hospital as well as provide the best overall care for the child (Edwards, Van Riper, & Kileny, 1995). Children with CHARGE who have facial palsy are 2 to 3 times more likely to have hearing loss on (at least) the side of the face that has the palsy (Edwards, et al, 2002; Mosheim, 2006). Surgery during the early months of life can also be dangerous. This is due to the need for sedation during surgery because of airway obstruction, congenital heart defects, swallowing disorders and apnea (Edwards, et al, 1995). Hearing loss in children with CHARGE can be mixed or sensorineural, and can be associated with
cochlear defects, otitis media, and vestibular problems. Cochlear implantation is a successful method of intervention for children with malformations of the inner ear (Mosheim, 2006). A study by Edwards, et al (2002) revealed that as many as 81% of children with CHARGE syndrome have significant levels of hearing loss. Nearly 100% of children with CHARGE syndrome have had otitis media with effusion that affects hearing abilities (Edwards, et al, 2002). Cochlear implantation is not recommended for children with CHARGE who have severe congenital temporal bone abnormalities or lack of a cochlear nerve (Mosheim, 2006).

Many children with CHARGE syndrome have bony or absent semicircular canals, however, the cochlea is normally developed; this fact makes these children excellent candidates for CI (Toriello, 1999). Although temporal bone growth continues throughout childhood, the facial recess is fully developed by the time the child is born, making the cochlear implant surgery possible from a structural standpoint. Occasionally, when looking at candidacy of children with CHARGE, it is necessary to look at facial deformation to determine whether implantation is appropriate. Structural abnormalities can lead to facial nerve rerouting, which may impede surgical procedures. The cochlear implant surgery is very invasive and requires a large amount of therapy post-operatively. Because structural abnormalities of the outer ears are common, hearing aids are often difficult to fit and have very little benefit to the child. Because of the abnormal growth of the ear canal, it is necessary for a child with CHARGE syndrome to have several hearing aid fittings within the first few years of life, making hearing aids difficult to afford and maintain.
Lanson, et al (2007) explored the viability of cochlear implantation on children with CHARGE syndrome. Their results found that there was an increase in auditory benefit but no research to provide evidence to suggest that this auditory benefit also increased development of oral language skills. Audiologic rehabilitation is said to have positive affects on children with CHARGE syndrome and their ability to develop symbolic language. It is known that children with CHARGE syndrome and CI are more aware of their surrounding environment after the CI is placed. The intellectual capabilities of children with a developmental disability are always underestimated (Mosheim, 2006).

**CHARGE syndrome, Receptive and Expressive Language Development and Cochlear Implantation**

Because communication disorders are associated with CHARGE syndrome, many children struggle to live a normal life. In order to communicate effectively, one must develop symbolic language. Approximately two-thirds of children with CHARGE syndrome develop some sort of symbolic language (Mosheim, 2006). Because children with CHARGE syndrome vary so greatly in their degree of disability, it is difficult to state one reason for the variance in language development. There are a significant number of children with CHARGE syndrome who do develop symbolic language that is either spoken or signed (Thelin and Fussner, 2005). Walzman, Scalchunes, and Cohen (2000), determined that while children with additional disabilities had lower scores on speech and language evaluations, they showed improved connectedness with their environment after receiving the implant. According to Lanson, et al, (2007), careful planning for and treatment of children with a cochlear implant and CHARGE syndrome can lead to limited
benefits in auditory perception and advancement in understanding and connecting with
the world around them. This study showed that no oral output in language was attributed
to the cochlear implant.

Thelin and Stephens (1996) determined that 88% of parent/caregivers of children
with CHARGE syndrome interviewed for their study said that their child had a structural
ear abnormality or hearing loss. In this study, parent/caregivers were then asked to rank
the abnormalities in regard to what was most significantly impairing their child’s
communication; the majority (51%) said hearing loss. It was reported that many of the
children in the study used perlocutionary communication including crying, laughing and
babbling (Thelin and Fussner, 2005). Many parent/caregivers who participated in the
study believed that CHARGE syndrome significantly adversely effected their child’s
overall development, communication, and educational development.

Edwards, et al, (2002) determined that the younger a child (regardless of multiple
disabilities) is implanted, the more likely the child is to make gains in the area of
communication, especially when speech and language therapy is implemented. Edwards,
et al, (2002) states that it is necessary to have realistic expectations for the implant
depending on the child’s preexisting cognitive functioning and developmental level.

In CHARGE syndrome, the number and the intensity of anomalies or
impairments is so large that it is difficult to define a characteristic/typical group of
individuals with CHARGE syndrome. Therefore, consequences of certain patterns of
impairments in the association are not yet understood.

The need to investigate the benefits of cochlear implants further for children with
CHARGE syndrome has become necessary so that parents/caregivers can make informed
decisions when it comes to planning the treatment and care of their child. Without research to support that cochlear implants are beneficial to language acquisition for children with developmental disabilities, parents/caregivers have difficulty determining an advantageous course of treatment for the development of their child.
CHAPTER II.

METHODS

This research study is part of a larger study investigating the outcome of deaf children with developmental disabilities being conducted at a large urban hospital. Receptive and expressive language outcomes for the subgroup of children with CHARGE syndrome and CI are the focus for this study. Four children with co-occurring CHARGE syndrome and a CI were evaluated to determine the language benefits that have been acquired as a result of the implantation. Results of a language evaluation were compared among cases in the areas of receptive and expressive language abilities.

The specific aim of this study was to assess the receptive and expressive language skills of children with CHARGE syndrome and CI in comparison to each other and personal language growth since implantation. A qualitative comparison of language scores was completed to determine benefits in communication that each child had acquired post implantation.

Participants

Children were eligible for this study if they 1) were between 2 years, 4 months and 6 years of age at the time of the study and 2) presented with CHARGE syndrome and a cochlear implant. The children in the study must have received their implant before 36 months of age to qualify for the study, and must have received the CI prior to enrollment. Eligible participants for the study were recruited through the hospital’s division of Developmental and Behavioral Pediatrics via letters mailed to their homes, as well as word of mouth recruitment from a developmental pediatrician. Parents/caregivers were
notified the purpose of the study and the extent of their involvement in the study. Parents/caregivers signed informed consent to participate in the study.

Materials/Procedures

All children recruited were administered the Preschool Language Scale – 4th edition (Zimmerman, Steiner, Pond, 2002). The PLS-4 assessed the child’s receptive and expressive language abilities. The Auditory Comprehension subscale (receptive language) included assessment of the child’s basic receptive vocabulary, and understanding of concepts, grammatical markers, complex sentences, comparisons and inferences. The Expressive Language subscale included the assessment of the child’s ability to name objects, use concepts of description, express quantity, use grammatical markers, and produce multi-word utterances. A benefit of this assessment was that it allowed for parent/caregiver input on a variety of questions, so parent/caregiver involvement was a necessary asset during the evaluation.

A hospital interpreter was present during the evaluation if the child used or benefited from sign language. If an interpreter was used during the evaluation, two language scores were reported, one for verbal stimulus only, and another for verbal and sign (total communication). Although sign language was used to facilitate communication, the total communication score is not recognized as a valid PLS-4 score as the test is not normalized using sign language. Instructions were given to the interpreter to sign exactly what the clinician presented verbally. Stimulus was presented verbally first, and repeated in sign, if needed. Some questions were unable to be translated into sign language, such as labeling body parts, as the signs provide the answer.
During the time of the evaluation, the parent/caregivers were asked to complete a questionnaire to collect information on outstanding issues that may have had an effect on the results of the testing. Information collected in this questionnaire (Appendix B) include basic subject characteristics (See Table 3 for a comparison of CHARGE syndrome characteristics across participants), educational programs the child has been involved in, intensity and attendance of therapies, and educational level and other related information of the parent/caregivers.

The information gained during the evaluation was then compared to information obtained through chart review and the results of the Revised Gesell Developmental Schedules assessed by the developmental pediatrician. The areas evaluated were adaptive functioning, gross and fine motor functioning, language functioning and personal-social functioning. The Revised Gesell Developmental Schedules were administered previous to the child receiving the cochlear implant, and thus provided a brief, but appropriate indication of the language level at which the child was functioning prior to receiving the cochlear implant.
CHAPTER III
RESULTS

A qualitative case design study of four participants, 50% male and 50% female, determined that three out of four participants showed increase in both receptive and expressive language as judged by the PLS-4 when compared to the RGDS. Three of the four participants scored within the 1st percentile of the PLS-4 in the areas of auditory comprehension and expressive communication using supplemental sign. The fourth participant scored within the 3rd percentile for auditory comprehension when using supplemental sign, and the 1st percentile for expressive communication using supplemental sign. Tables 1 and 2 show the results of the PLS-4 for each participant in Auditory Comprehension and Expressive Communication. The Revised Gesell Developmental Schedules were administered when the children were between the ages of 9 months old and 3 years old. The results/abilities of each individual case are presented below.

<table>
<thead>
<tr>
<th></th>
<th>Age Equivalent</th>
<th>Standard Score</th>
<th>Percentile Rank</th>
<th></th>
<th></th>
</tr>
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<tbody>
<tr>
<td>Bobby</td>
<td>10 mo</td>
<td>57</td>
<td>1</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td>Margaret</td>
<td>6 mo</td>
<td>50</td>
<td>1</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td>Jeff</td>
<td>3 yrs, 6 mo</td>
<td>64</td>
<td>1</td>
<td>3</td>
<td></td>
</tr>
<tr>
<td>Karen</td>
<td>5 mo</td>
<td>50</td>
<td>1</td>
<td>1</td>
<td></td>
</tr>
</tbody>
</table>

*SS – Supplemental Sign
Case History and Development – Bobby

Bobby is two years, 4 months old. He was born at 32 weeks gestational age and hospitalized for the first 4 months of life. He has a significant profound bilateral sensorineural hearing loss that was diagnosed when he was 4 weeks old. He has had choanal atresia repair twice since birth. Bobby has also had patent ductus arteriosus with ligation surgery to repair heart defects. He has Grade IV ureteral vesicular reflux. He is currently using a Tracheostomy tube and is fed via G-Tube and Nissen. Bobby has a history of aspiration of saliva. A high palatal arch makes his ears drain fluid, which required the insertion of PE Tubes twice. He wore hearing aids in the first year of life, but they were difficult to keep on due to outer ear malformations. He received a cochlear implant when he was 15 months old.

According to parent/caregiver and pediatrician report, Bobby has functional vision. An MRI from when he was an infant showed he had an extraaxial hemorrhage in the posterior fossa, blood layering in the occipital horns and blood in the left lateral ventricle near the choroid plexus. He has poor definition of facial nerve canals. He is
recently recovered from a femur fracture that occurred during physical therapy, which delayed his motor growth.

**Therapy Services Received:** Until recently, Bobby received occupational, physical and speech therapy services through a home based program biweekly because of the intensity of his medical needs. At this time he was also receiving Newborn Infant Hearing Screenings on a monthly basis to monitor any fluctuation due to the fluid drainage. He is currently receiving outpatient services for occupational, physical and speech therapies. He also receives early intervention services through his local school district and previously through Help Me Grow. In addition, He receives services through MR/DD and Job and Family Services/Social Services.

**Current Language Functioning according to PLS-4 Scores**

**Receptive Language** – Bobby was able to demonstrate anticipation for what would happen next, this was seen with games like “peek-a-boo”, “playing ball” and “stacking blocks”. He was able to actively search for the source when sounds were made. He would occasionally turn when his name was called, but this was not done consistently. Bobby was able to properly respond to the command ‘no’. He was able to use more than one object in play and showed appropriate use of these objects. Parent/caregivers report that he has difficulty following routines or directions, understanding specific words in phrases, and identifying familiar objects or photographs.
**Expressive Language** – Bobby’s main strategy for expressing himself is through eye gaze and facial expressions. When he is angry, his most common way of expressing himself is kicking his feet. Mother reports grunting/squealing is occasionally made. Bobby was able to sign the word “more” consistently and in meaningful contexts. He was able to reach for different toys that he wanted, and would make eye contact to look for reactions. He would clap for himself and enjoyed praise from others. Mother reports that he imitates what to do with common objects, for example, with his eye drops; he will put the bottle up to his eye.

The Revised Gesell Developmental Schedule (RGDS) was given when Bobby’s chronological age was 11 months. At this time in the area of communication, he demonstrated an alert face and direct regard, giving him a RGDS score of 8 weeks old in the area of communication/language.

**Case History and Development – Margaret**

Margaret is a 2 year, 11 month old female. Margaret was born a full term infant with multiple congenital anomalies including visual and hearing impairments. She has bilateral coloboma. She was in the NICU and the RCNIC for 2 months after birth. At birth she exhibited hemivertebra and scoliosis, hydrocephalus, and laryngomalacia. Margaret has an absent corpus colosum and septum pallucidum. She currently uses a tracheostomy tube due to choanal stenosis. She is fed through a G-Tube and has Gastroesophageal Reflux as well as difficulty with respiration. She had a heart murmur
on this first day of life, leading to coarctation of her aorta. Margaret is on a BiPAP at night to aide with breathing.

Margaret has absent semicircular canals and has had two sets of PE Tubes. The configuration of her right cochlea is dysmorphic. She wore hearing aids at 3 months. She has abnormalities in both temporal bones and she has left facial nerve paralysis. Margaret received her cochlear implant at 18 months of age.

**Therapy Services Received:** Margaret received early intervention services through Help Me Grow. She currently is receiving aural rehabilitation, speech therapy, physical therapy and occupational therapy. She also is followed by the Regional Infant Hearing Program and the city’s Association for the Blind.

**Current Language Functioning according to PLS-4 Scores**

**Receptive Language** – Margaret was able to locate the source of environmental sounds, such as a door slamming, and was able to turn her head in the direction of the sound. Margaret looks intently at objects within her focus. She is able to shake and bang two objects together in play, she did well using more than one object in play. Margaret was observed anticipating what would happen next during a block game that was being played with the clinician. Margaret’s mother reports that Margaret understands “no-no” when given verbally, but only in direction to leave her tracheotomy tube alone. Mother reports that “no-no” has not generalized to other areas. Margaret and the clinician read the story *The Very Hungry Caterpillar* by Eric Carl and Margaret was very focused on the pictures. She sat on her mother’s lap during the evaluation and the book was held 2-3
inches from her face. She demonstrated pre-literacy skills by turning the pages of the book. Her mother reported that she enjoys watching TV/videos and her favorite are shows that incorporate sign language.

Expressive Language – Margaret was able to vary her pitch when laughing. She was able to show pleasure/displeasure with her vocal tone/grunting and facial expression. She was able to play simple games with the clinician such as the “block game” and the “tickle game”. During these games, Margaret was able to communicate her wants by non-verbally pushing the blocks toward the clinician to have her begin the game again. Her mother reported that she consistently pushed things away when she is done. She will also move her mother’s hands around her when she wants a hug. She kisses by putting her mouth to your forehead. Margaret was able to respond to the speaker through smiling. Margaret has not yet vocalized any babbling or sounds/syllables. She makes choices through reaching, but is sometimes slow to make a choice; however, her mother reported that she is improving.

The RGDS was administered when Margaret was 9 months old. At this time her language functioning was at 16-20 weeks. She was able to laugh aloud.

Case History and Development – Karen

Karen is a five year old female. She was born at full term. She has a history of cleft lip and palate, and had a maxillary appliance placed at 3 months of age. Her cleft lip and nasal repair was done at 4 months and her palate was repaired at 9 months of age.
Karen’s medical history is also significant for patent ductus arteriosus and left superior vena cava to the coronary sinus. In addition she has left ventricular hypertrophy. Her vision is impaired by bilateral coloboma. Karen is near sighted and sees best with high contrast. She also has behavioral difficulties, such as obsessive compulsive disorder, tic like motions, difficulties with sleeping, and cyclical vomiting. Recently she was noted to have episodic facial pain. She received bilateral PE tubes at 2 months of age and a cochlear implant at 18 months of age.

**Therapy Services Received** – Karen receives speech therapy and physical therapy once a week, aural rehabilitation every other week, and occupational therapy through her county Early Intervention program. Services had been provided with the Regional Infant Hearing Program prior to the age of 3 years. She is followed by the area’s Association for the Blind.

**Current Language Functioning according to PLS-4 Scores**

**Receptive Language** – Karen was able to glance momentarily at the person who talked to her. Her parent/caregivers reported that she reacts to songs on the television. She looked intently at the speaker and mouthed several objects before shaking them to examine them. She was more engaged when an object was within 16 inches of her face. Her parent/caregivers also reported that she is able to anticipate what will happen next by using her picture schedule at school and at home. She was able to use more than one object in play, but did not have the concept of pretend play; she examined the toys as a form of interacting with them. Karen was able to respond to “no-no” when it was paired
with the sign. She also is able to understand words and phrases when given to her in sign. Mother reports that she has a receptive sign vocabulary of 60-70 words. She was able to point to colors that her mother signed to her, such as orange, red, and green. Mom reports that she knows several other colors. Karen did not actively search to find the person who was speaking or a sound when the source was not available. She was not able to identify familiar objects from a group of objects. Her mother reports that when she is at home, they work on identifying flashcards and she is able to point to pictures such as “cookie”, “ball”, and “shoe”, but this was not seen during the evaluation. She can identify some letters of the alphabet, and turns when her name is called. Karen knows signs such as “mom, dad, dog, car, baby, hat, book”. She can match photographs to objects, consistently make a selection when presented with at least four pictures, and distinguish between a conversation partner and bystander. She uses a picture schedule throughout the day.

**Expressive Language** – Karen was able to vocalize soft, throaty sounds and was able to vary her pitch length for the /uh/ sound she produced. She was able to vocalize pleasure and displeasure sounds and protest. She vocalized /uh/ to show disapproval during the evaluation. Karen primarily communicated nonverbally through gestures and pushing/pulling behaviors. For example, mother reports that when she wants something, she will pull her to the object and then point. Her parent/caregivers report that she is able to sign a few words, such as “milk”, “more”, and “up” and that she signs mostly with her mother. Her parent/caregivers say that she is able to communicate her wants using gestures and actions. If she wants to go outside, she will go and get her mother’s shoes.
The RGDS was administered when Karen was 11 months old. At this time, her communication/language level was 9-10 months. This was scored with regard to supplemental sign and with regard to visual impairment. Her verbal only abilities were not reported on this scoring the RDGS. She was able to vocalize a single consonant sound, cry, respond to "no-no", and use gestures to communicate. She used sign approximations for “da-da” and “ma-ma” with meaning according to parental report. This language score is higher than the language score found by the PLS-4, but it also does not provide as detailed a description of the child’s receptive and expressive language abilities as the PLS-4 does.

Case History and Development – Jeff

Jeff is a 5 year, 9 month old male. Jeff has CHARGE syndrome with co-occurring Klinefelter’s syndrome. Jeff has profound sensorineural hearing loss in both ears, and received a cochlear implant when he was 4 years, 3 months old. In addition to the sensorineural hearing loss, Jeff has vestibular and cochlear dysplasia. Hearing loss was identified two weeks after birth and he was amplified at 2 years old. He has nearly absent semicircular canals. He has developmental delays and blindness in one eye due to bilateral coloboma. He has hypoplastic left prechiasmatic optic nerve and chiasm. Jeff has an abnormal auditory nerve in the right ear. He has a history of an aortic arch problem. He also has orchiopexy. He was previously on a G-tube, but eats orally now. Oral motor skills are lacking in imitating mouth movements and difficulty feeding.
Therapy Services Received: Jeff receives services through his local school district’s early intervention/preschool programs where he receives occupational therapy, physical therapy and speech therapy. He uses sign language in school. He receives private aural rehabilitation, speech, physical and occupational therapies.

Current Language Functioning according to PLS-4 Scores

Receptive Language – Jeff was able to correctly perform more tasks when supplemental sign language was used. He demonstrated the ability to identify basic colors and common objects from a variety of pictures. He was able to do these tasks with auditory directions only. Using auditory directions only, he was also able to understand basic picture analogies through pointing to the correct picture. In addition, Jeff also demonstrated understanding of basic shapes. With the use of supplemental sign, Jeff was able to understand expanded sentences and passive voice sentences. Jeff was able to identify general time concepts such as night and day, but was unable to identify time-sequence concepts such as first/last. Although he was able to identify number words, Jeff had difficulty understanding more complex quantitative concepts.

Expressive Language – Jeff uses gestures, signs, and facial expressions during spontaneous conversation. When he does produce speech, Jeff primarily uses 1 word utterances consisting of nouns and a few modifiers (numbers and colors). Jeff was able to name common objects. He also demonstrated the ability to express how common items are used (i.e. Tell me what you do with a spoon/towel). However, Jeff demonstrated difficulty answering logic questions. He was unable to express possessives or complete simple analogies using opposite concepts.
The RGDS was administered at 3 years, 11 months. At that time, Jeff’s language abilities were that of a 15-18 month old child when scored with consideration for expressive sign language rather than verbal language. He was able to understand turn taking, and used several spontaneous signs, he was not able to combine more than one sign into a phrase.

### Chart 3

The following chart shows a summary of the common diagnostic characteristics common to a diagnosis of CHARGE syndrome exhibited by each subject.

<table>
<thead>
<tr>
<th>Characteristic</th>
<th>Bobby</th>
<th>Margaret</th>
<th>Jeff</th>
<th>Karen</th>
</tr>
</thead>
<tbody>
<tr>
<td>Coloboma</td>
<td>-</td>
<td>+</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>Choanal Atresia</td>
<td>+</td>
<td>+</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>Cranial Nerve Anomalies</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>Hearing Loss</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>Inner Ear Abnormalities</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>External Ear Malformation</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>Cardiovascular Malformation</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>Genital Abnormality</td>
<td>+</td>
<td>-</td>
<td>+</td>
<td>-</td>
</tr>
<tr>
<td>Cleft Lip/Palate</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>+</td>
</tr>
<tr>
<td>Tracheo-Esophageal Fistula</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>Growth Retardation</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>Developmental Delay</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
</tr>
</tbody>
</table>

*+ characteristic exhibited, - characteristic not present*
Tables 4 and 5 show the results of the PLS-4 by task, demonstrating which subject was able to complete which task.

**Table 4.**

<table>
<thead>
<tr>
<th>PLS-4 Receptive Language Comparison</th>
<th>Bobby</th>
<th>Karen</th>
<th>Jeff</th>
<th>Margaret</th>
</tr>
</thead>
<tbody>
<tr>
<td>Shakes and Bangs Objects</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>Has name recognition</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>-</td>
</tr>
<tr>
<td>Demonstrates Anticipation</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>Looks at things pointed out</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>Understands Hand Gestures</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>-</td>
</tr>
<tr>
<td>Understands no-no</td>
<td>+</td>
<td>(s)</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>Understands words/phrases</td>
<td>-</td>
<td>(s)</td>
<td>+</td>
<td>-</td>
</tr>
<tr>
<td>Can play with +1 object</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>Understands Routines</td>
<td>-</td>
<td>-</td>
<td>+</td>
<td>-</td>
</tr>
<tr>
<td>Plays appropriately</td>
<td>+</td>
<td>-</td>
<td>+</td>
<td>-</td>
</tr>
<tr>
<td>Identifies Objects</td>
<td>-</td>
<td>-</td>
<td>+</td>
<td>-</td>
</tr>
<tr>
<td>Identifies Photographs</td>
<td>-</td>
<td>-</td>
<td>+</td>
<td>Shows interest</td>
</tr>
<tr>
<td>Understands “my turn”</td>
<td>-</td>
<td>-</td>
<td>+</td>
<td>-</td>
</tr>
<tr>
<td>Can label body parts</td>
<td>-</td>
<td>-</td>
<td>+</td>
<td>-</td>
</tr>
</tbody>
</table>

*+ behavior present, - does not exhibit behavior, (s) present with supplemental sign*

**Table 5.**

<table>
<thead>
<tr>
<th>PLS-4 Expressive Language Comparison</th>
<th>Bobby</th>
<th>Karen</th>
<th>Jeff</th>
<th>Margaret</th>
</tr>
</thead>
<tbody>
<tr>
<td>Varies pitch/length/volume</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>Vocalizes pleasure/displeasure</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>Vocalizes when talked to</td>
<td>+</td>
<td>-</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>Protests by gesturing/vocalizing</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>Vocalizes two different sounds</td>
<td>-</td>
<td>-</td>
<td>+</td>
<td>-</td>
</tr>
<tr>
<td>Combines sounds to form syllables</td>
<td>-</td>
<td>-</td>
<td>+</td>
<td>-</td>
</tr>
<tr>
<td>Seeks attention from others</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>Plays simple games</td>
<td>+</td>
<td>-</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>Communicates nonverbally</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>Participates in 1-2 minute play routine</td>
<td>+</td>
<td>-</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>Babbles two syllables together</td>
<td>-</td>
<td>-</td>
<td>+</td>
<td>-</td>
</tr>
<tr>
<td>Has vocabulary of at least 1 word</td>
<td>(s)</td>
<td>(s)</td>
<td>+</td>
<td>(s)</td>
</tr>
<tr>
<td>Initiates turn taking</td>
<td>+</td>
<td>-</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>Extends toy/points to object to show others</td>
<td>-</td>
<td>-</td>
<td>+</td>
<td>-</td>
</tr>
<tr>
<td>Uses vocalizations/gestures to request toys/food</td>
<td>-</td>
<td>+</td>
<td>+</td>
<td>-</td>
</tr>
<tr>
<td>Imitates words</td>
<td>-</td>
<td>(s)</td>
<td>+</td>
<td>-</td>
</tr>
<tr>
<td>Has vocabulary of 5-10 words</td>
<td>-</td>
<td>(s)</td>
<td>+</td>
<td>-</td>
</tr>
</tbody>
</table>

*+ behavior present, - does not exhibit behavior, (s) present with supplemental sign*
CHAPTER IV.
DISCUSSION

The purpose of this study was to determine the amount of increase (if any) that a cochlear implant had on receptive and expressive language development. This study found that the three out of the four participants involved showed an increase in receptive and expressive language when compared to their RGDS scores, but it is unclear if this increase is due to the cochlear implant. Parents reported that their children increased in awareness to their environments after the implantation. While only one of the participants developed verbal language, parent/caregivers reported all of the subjects showed an increase in overall demeanor following implantation. Various forms of communication (signs, gestures) were demonstrated by the children, only in one case did verbal language develop post implantation. While that particular child was able to verbalize, he primarily used sign to communicate. Two of the parents/caregivers noted that if their child’s implant was removed, the children would make vocalizations in protest/recognition of the change in hearing.

Results of this evaluation were scattered and it was found that no consistent pattern of language for children with CHARGE syndrome was demonstrated. Appendix A shows the months delayed that each child demonstrated according to the results of the PLS-4 and Tables 4 and 5 depict the four participant’s performance on various language tasks on the PLS-4. Jeff’s baseline language performance exceeded the average range on this evaluation and as a result his true language abilities are not shown in Tables 4 and 5. Parents/caregivers of children with CHARGE syndrome reported that their child showed increased awareness to the surrounding environment. In all but one participant, receptive
language saw increases according to the PLS-4 when compared to the RDGS. Verbal expressive language was developed only in one participant. Each child had developed some use of sign and gestures to communicate expressively. Although one child did demonstrate verbal output, he communicated primarily via sign language. All parents/caregivers had seen positive results that they attributed to the cochlear implant surgery and found that the increase in awareness added to the quality of life for their child. Parent/caregivers reported that the cochlear implants helped their child, but this study did not yield statistical documentation to support this.

The results of this study aid parent/caregivers in determining whether or not it is beneficial to include cochlear implantation on their child’s treatment plan. Because the parent/caregivers of the children included in the study reported the increase in awareness of environment, it is important for parent/caregivers who are considering cochlear implants to understand the importance of this benefit and how it will impact their child.

The finding that children with CHARGE syndrome and a cochlear implant do not necessarily develop a verbal language schema is consistent with the results from Waltzman, Scalchunes & Cohen (2000). They previously found that children with multiple disabilities showed benefit from the cochlear implant, but not necessarily in language ability. Wiley, Meinzen-Derr and Choo (2004), also mentioned previously, found in their study that children with developmental disabilities and a cochlear implant received some benefit to communication ability due to the implant, but not necessarily verbal language. While these results are similar, the criteria through which the children were chosen and the methods of how they were evaluated differed. The children in this study were chosen because their primary diagnosis was CHARGE syndrome, received a
cochlear implant by 36 months of age, and met the original inclusion criteria for the study. Because of the time variable involved, the study compared the children to each other and also looked at their growth in language since the administration of the RGDS.

An alternative explanation for the communication gains found in this study could be the intensity of therapies received by each child. This was not something that was controlled for, and it likely has a significant impact on language development. Also, overall growth and development of the child and natural progression may account for the language that each child has gained, it may not necessarily be a direct result of the cochlear implant.

Clinical Relevance

This study supplements data currently being obtained to support that language gains are made after cochlear implantation in children with CHARGE syndrome. It provides evidence for cochlear implant candidacy in children with CHARGE syndrome and determines the degree of benefit for children with CHARGE syndrome.

Limitations

There were several limitations included in this study. There were only 4 subjects all with varying ages and multiple diagnoses. The subjects also demonstrated differences in vision, posing a difficulty when viewing visual stimuli. This may have altered the results of the study because of the visual stimuli present in the Preschool Language Scale -4th edition. This was modified during the study by holding the visual stimulus closer to the child’s face and allowing them to investigate each picture for extended time if needed.
Information obtained through this evaluation was compared to the results of the Revised Gesell Developmental Schedules given by the developmental pediatrician. The RGDS provides a general description of receptive and expressive language to compare to PLS-4 which looks at receptive and expressive language more thoroughly. It is not known whether the increase in communicative abilities is a result of interventions given to the child, natural growth of language development or a direct result of the cochlear implant. Varying amounts/intensities of therapies were provided to each of the children, so it is difficult to determine the impact of this therapy on receptive and expressive language development.

Parental/caregiver input accounted for a large amount of the information compiled for this study. Input was given through the parent/caregiver questionnaire, as well as on those items of the PLS-4 evaluation that allowed for parent/caregiver response if the child did not demonstrate the skill when prompted on the assessment. This may have skewed the results because parent/caregiver perception that a skill has been mastered may be different than what the speech language pathologist would judge based on actual evidence of the child’s performance.

No consistent interpreter was used to evaluate the children, therefore the amount and complexity of what was interpreted may have varied across evaluations. The actual signs chosen to represent specific vocabulary may have also varied. The children had each been implanted at different ages, and as a result, all had a different “hearing age”. This is a limitation to the study because some children have received more input from their implant because it was turned on earlier.
Further Research

There are several areas in this research study that can be further developed. The impact of the environmental awareness that each child received and the significance it had on quality of life would be an area to be further addressed. Determining an amount of environmental awareness that is consistent or expected for this group of children would be beneficial to parent/caregivers determining a course of treatment and would assist in forming expectations of their child’s abilities.

Re-administering the Preschool Language Scale – 4th Edition at a later date would allow for research to be completed in the area of growth over a certain time period. This could be helpful as a longitudinal study to see at what age level the most gains are made, and if there is any further correlation to the cochlear implant. This would allow for researchers to track each child’s progress and determine whether continued language growth is seen or whether the child has reached a plateau in their development. It may also be interesting to compare the language levels of the children with CHARGE and a CI with a control group of hearing children matched by developmental level only.

A longitudinal study evaluating these four children again in a year would yield information comparing the rate of language growth across subjects. Controlling for therapy intensity may also produce more consistent results. Comparing the children based on hearing age rather than chronological age may also present differing results.

Children with CHARGE syndrome and a cochlear implant can make gains in language and communication, although not necessarily verbally. Children with CHARGE syndrome who have a hearing loss who receive a cochlear implant are better able to connect with the hearing world and as such, have a better quality of life.
BIBLIOGRAPHY


disease, and choanal atresia with multiple anomalies: CHARGE syndrome.


APPENDIX A. The following graphs show the amount of delay (in months) of the 4 cases discussed in the study based on the age equivalents (in months) from the Preschool Language Scale 4th edition. The delays have been computed both with and without the use of supplemental sign language and were compared to each other in the following bar graphs.
APPENDIX B. The questionnaire completed by the parents during the time of the evaluation. It allows the clinician to gain an understanding of possible outlying factors that may have an influence on the child's receptive and expressive language development.

**Background information about your child**

1. What is your child’s gender?
   - _____Male  ______Female

2. Child’s date of birth:
   - ___________ month/day/year

3. What is your child’s race: (please check all that apply):
   - ___African-American/Black
   - ___Caucasian/White
   - ___Hawaiian/Pacific Islander
   - ___American Indian or Alaskan Native
   - ___Hispanic/Latino
   - ___Unknown
   - ___Other (specify)________________

4. Please indicate how your child communicates (check all that apply):
   - ___Speech  ___Signing  ___ Behavior  ______ Other (please specify)________________

Please circle the category that best describes the type of school or day care your child attends (circle all that apply).

Circle the percent of your child’s school day that is spent in each of the schools.

- _____ My child does not attend school

<table>
<thead>
<tr>
<th>School Type</th>
<th>1-25%</th>
<th>25-50%</th>
<th>51-74%</th>
<th>75-100%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Fully mainstream</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Partially mainstream</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Day special school</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Preschool disabilities</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Self-contained classroom</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Ohio Valley Voices</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>St. Rita School for the Deaf</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Home education</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Other (please specify)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
6. If your child has a cochlear implant, does he/she use interpreter services at school?  

___Yes    ____No

7. Please indicate how often (i.e. 1 hour/week, 1 hour/month) does your child receive the following therapies at each of the locations below (check all that apply):  

<table>
<thead>
<tr>
<th>Therapy</th>
<th>School</th>
<th>Private agency</th>
<th>Home based</th>
</tr>
</thead>
<tbody>
<tr>
<td>Speech therapy</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Occupational therapy</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Physical therapy</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Behavioral psychologist</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Vision Specialist</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Aural Rehabilitation</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

**Background information about yourself**

1. What is your gender?  

___Male    _____Female

2. Please indicate which category describes your relationship to your child.  

___Mother/Stepmother    _____Father/Stepfather    ____Grandparent

___Other (Please specify) __________________________________

3. What is the highest grade or level of school you have completed?  

___8th grade or less  
___Some high school  
___High school graduate/high school degree (GED)  
___Some college or technical school  
___Completed college  
___Post graduate training/degree

4. How many siblings does your child have? ______
5 Which category best describes your annual household income before taxes?

___ Under $10,000
___ Between $10,000 and $19,000
___ Between $20,000 and $29,000
___ Between $30,000 and $39,000
___ Between $40,000 and $49,000
___ Between $50,000 and $59,000
___ Between $60,000 and $69,000
___ Between $70,000 and $79,000
___ $80,000 or more

6 Which of the following best describes your child’s main source of health insurance?

___ Private health insurance provided by a commercial carrier (such as an HMO or PPO)
___ Public health insurance (such as Medicaid, Medicaid HMO, or BCMH)
___ A combination of Private and Public
___ No health insurance