UNIVERSITY OF CINCINNATI

Date: 5/22/09

I, Mark Ostrowski,

hereby submit this original work as part of the requirements for the degree of:
Master of Arts

in Communication Sciences and Disorders

It is entitled:
Cochlear Implants and Language Outcomes in Children with Symptomatic CMV.

________________________________________________________________________

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I have reviewed the Thesis/Dissertation in its final electronic format and certify that it is an accurate copy of the document reviewed and approved by the committee.

Committee Chair signature: Sandra M. Grether
Cochlear Implants and Language Outcomes in Children with Symptomatic CMV

A Thesis Submitted to the Graduate School of the University of Cincinnati in partial fulfillment of the requirements for the degree of Master of Arts in the Department of Communication Sciences and Disorders College of Allied Health Sciences

by

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May 2009

Committee Chair: Sandra M. Grether, Ph.D.
ABSTRACT

Before and after the decision to implant a child, caretakers and professionals need the guidance of data that document the potential effects cytomegalovirus (CMV) may have upon their child’s use of a cochlear implant (CI). This study attempted to contribute to such data through the qualitative analysis of post-implant language abilities of six young children with symptomatic CMV and CI. Receptive and expressive language abilities represented by age-equivalents and language quotients, were obtained from evaluations using the Preschool Language Scale – 4th Edition (PLS-4). Results demonstrated wide variability in receptive and expressive language abilities; however, all scores were significantly below the average range. Three of the four cases with pre-implant scores demonstrated growth in expressive language, but limited gains in receptive language. Analysis of case histories revealed that a diagnosis of cognitive delay may have a significant impact on the language gains after implantation. Findings have implications on the pre-implant decision making process, and indicate the need for future research into cognitive processes that limit outcomes and increase variability.
ACKNOWLEDGEMENTS

I owe tremendous thanks to my thesis committee and others for helping me prepare this thesis. Many thanks to Dr. Meinzen-Derr, whose impressive intellect and statistical insight made this study possible. Thanks to Dr. Wiley, who contributed immensely to my understanding of childhood development, and the fine art of arena assessment. Many thanks to Dr. Creaghead, whose perspective helped redirect and clarify my approach to this study. My sincerest thanks to Dr. Sotto, who has helped me succeed from the very beginning of my graduate years. And of course, I am forever grateful for the guidance of Dr. Grether, who has profoundly influenced my professional development with guidance that will last for years. Special thanks to Mel, my loving wife, and to her patience, which continues to astound me.
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CHAPTER I

Introduction

As candidacy for cochlear implantation expands to include younger children and those with more residual hearing, the same technology has become more accessible to children with additional disabilities (Hamzavi et al., 2000; Nicholas & Geers, 2007; Vlahovic & Sindija, 2004). Benefits are clear in documented cases of typically developing children (Edwards, Frost & Witham, 2006; Wilson, & Dorman, 2008), but the positive impact this technology could have on children with developmental disabilities has yet to be consistently documented (Fukuda et al., 2003; Rajput, Brown, & Bamiou, 2003; Wiley, Jahnke, Meinzen-Derr, & Choo, 2005). The need for clinically meaningful data that contributes to our understanding of this potential benefit is necessary, particularly if one considers the link between congenital hearing loss and developmental delay. In a study by the Metropolitan Atlanta Developmental Disabilities Surveillance Program (MADDSP), 30% of children with sensorineural hearing loss (SNHL) also had neurodevelopmental conditions (Van Naarden, Decoufle, & Caldwell, 1999). Another study concluded that for children with SNHL from a non-genetic origin, an estimated one third will have additional disabilities (Herer, Knightly, & Steinberg, 2007). One congenital etiology that is associated with both SNHL and additional disabilities is the often unpredictable human form of cytomegalovirus.

Cytomegalovirus (CMV)

Human cytomegalovirus (CMV) is the largest member of the Herpesviridae family, and is morphologically indistinguishable from herpes viruses found in humans. In general, the virus is relatively common and clinically silent for the majority of individuals (Schildroth, 1994). However, the virus has been known for many years as a deadly infectious agent to an unborn
infant. CMV transmission is not well understood but is believed to commonly travel from mother to infant through the placenta following either a primary or recurrent infection. Primary infection at an earlier gestational age, particularly an infection within 20 weeks of gestation, has generally been associated with worse outcomes for the child. Prenatal diagnosis of CMV can be obtained through fetal blood tests and amniocentesis, although these methods have not been prognostically reliable. Clinically observable signs and symptoms of CMV infection in the mother are usually similar to mononucleosis, if they are observable at all. Only 35-40% of pregnant women with primary infections transmit the virus to their unborn child, and most are asymptomatic at birth (Revello & Gerna, 2002).

The symptomatic form of CMV, however, results in a complex range of damage to the central nervous system (CNS), and is one of the leading causes of congenital deafness (Noyola et al., 2000; Revello, & Gerno, 2002; Schildroth, 1994). According to the Centers for Disease Control (2008), about 40,000 children in the United States will be born with CMV each year, and approximately 10-15% will be symptomatic at birth. Although up to 90% of infected children will be asymptomatic at birth, they may develop permanent hearing loss along with other mental/physical impairments months and sometimes years later (Dahle et al., 2000; Noyola et al., 2001). Whether symptomatic at birth, or shortly after, the majority of infants who survive an active CMV infection potentially develop not only hearing loss, but mental retardation, psychomotor delays, learning disabilities and expressive language delays (Lee, Lustig, Sampson, Chinnici, & Niparko, 2005). In fact, almost every organ system in the human body is subject to the pathological effects of CMV (Schildroth, 1994).

Although predictions of developmental outcomes are notoriously difficult to make after a child is diagnosed with symptomatic CMV, the literature suggests some potentially reliable
prognostic indicators. For example, the severity and onset of hearing loss appears to be greater and arrive earlier if the child is symptomatic at birth (Fowler, Dahle, Boppana, & Pass, 1999). In addition, a study by Pass, Stagno, Myers and Alford (1980) found that microcephaly along with hearing loss, is associated with consistently poor outcomes (e.g. cognitive impairments and motor delays). Conboy et al. (1987), however, found that more than half of the participating subjects with congenital CMV had a normal IQ. In a more recent study, Pymen, Blamey, Lacy, Clark and Dowell (2000), found that CMV was the only cause of deafness in their subjects that reliably predicted motor and/or cognitive delays.

While research continues to search for prognostic indicators, the fact still remains; symptomatic CMV results in random injury and has a combination of features that cannot be individually confined to the infection (Schildroth, 1994). In addition, congenital CMV infection may be more prevalent than was previously reported, with greater than 40% of children with SNHL of an unknown origin later testing positive for CMV (Barbi et al., 2003). Therefore, children with severe to profound SNHL secondary to symptomatic CMV constitute a significant and heterogeneous population that often present with uncertainties prior to receiving a CI. If parents decide to implant early (e.g. under two years of age) this uncertainty may be important to consider during the pre-implant decision making process (Nicholas & Geers, 2007). For these parents of children with suspected developmental disabilities, data that delineate the variable range of language outcomes offer a critical contribution to help alleviate some of this uncertainty. With an etiology that is as common and unpredictable as CMV, decisions before and after implantation need to be guided by the same degree of data that are available to parents/professionals of typically developing children. More specifically, what are needed are
data demonstrating the receptive and expressive language abilities a child with CMV related hearing loss may acquire with a CI.

Purpose of Present Study & Research Questions

The purpose of this study is to qualitatively describe and interpret language outcomes of children with CI and symptomatic CMV. The following research questions were created to assist in this purpose:

1. What are the receptive and expressive language abilities of children with symptomatic CMV after receiving a cochlear implant?

2. What are the specific impairments in addition to hearing loss that may have an impact on these language outcomes?
CHAPTER II

Review of the Literature

*Cochlear Implants in Children without Additional Disabilities*

As the worldwide cumulative number of individuals with CI exceeds 120,000, the consensus is that cochlear implants provide many benefits to individuals with severe to profound sensorineural hearing loss (Wilson & Dorman, 2008). According to the National Institute on Deafness and other Communication Disorders (2007), cochlear implants coupled with intensive therapy can help young children acquire speech, language and communication skills. Supporting this are research studies confirming individual successes which have shown that, in most cases, CIs are a worthwhile option and more appropriate than alternative measures (e.g. hearing aids). For example, in a study that included 29 prelingually deaf children with three or more years of CI experience, results showed that CI users performed better on expressive language tests than did 29 deaf children who used hearing aids (Tomblin, Spencer, Flock, Tyler, & Gantz, 1999).

Expectations after implantation, however, are not always the same, and the expanding literature suggests that success in the perception and production of speech is variable. In addition to this, Pisoni et al. (2008) listed the following key findings that have been observed across most CI centers.

1. There are large individual differences in language outcomes (e.g. spoken language).
2. Age of implantation is critical for sensitive periods (e.g. the earlier the better).
3. Early language experiences impact language development (e.g. auditory-oral versus total communication).
4. There are no reliable pre-implant predictors of individual outcomes (e.g. receptive and expressive language development).
5. Learning abilities emerge after implantation, and performance with a CI improves in time.

6. Performance with a CI depends upon neural reorganization and cross modal plasticity (e.g. adapting to the degraded acoustic representations).

Whenever possible, early implantation is particularly important. At implant centers around the world, cochlear implantation between one to three years of age is considered preferable to facilitate early language learning (Dettman, Pinder, Briggs, Dowell, & Leigh, 2007; Richter, EbBele, Laszig, & Lohle, 2002). Because the neurological systems behind auditory development depend upon input during sensitive time periods, early implantation is necessary to maximize functional capacity (Tomblin et al., 1999). Researchers in Germany, for example, studied the receptive and expressive language skills of 106 children with a minimum of two years experience wearing a CI. Results showed that age at implantation was the most important prognostic indicator of more typical language learning, as those implanted earlier demonstrated the smallest discrepancy between speech development age and chronological age (Richter et al, 2002).

Also included in the preceding list by Pisoni et al. (2008), is the high degree of variability found across implant centers in the recipients’ acquisition of spoken language after implantation. Where one child could develop exceptionally clear verbal language, another child with the same device may develop only the awareness of sound with little to no speech. It seems that individual differences in cognition, brain plasticity, phonological processing and encoding, may determine the rate and extent to which a child will acquire verbal language (Pisoni, 2000). It could be that, even with the contribution of intensive intervention, a child’s ability to utilize the device’s degraded acoustic signal will ultimately determine his/her success.
Cochlear Implants in Children with Additional Disabilities

As previously mentioned, the expanding criteria for eligibility has contributed to the growing number of children with additional disabilities receiving a cochlear implant. However, the resulting percentage of children with CIs and additional disabilities may be larger than expected, since many developmental disabilities may be undetectable at very young ages. In a retrospective study of 69 children with cochlear implants, Wiley, Meinzen-Derr, and Choo (2004), found that 46% had at least one additional disability, and 16% had two or more additional disabilities. In each case, those with additional disabilities were not diagnosed prior to surgery. Results of a study conducted in Iran found that 15% of 398 prelingually deafened children with cochlear implants were diagnosed as having additional disabilities (Daneshi & Hassanzadeh, 2007).

Because of this large and increasing number of children with CI and additional disabilities, there is a need for reliable prognostic indicators that guide the preimplant decision making and parent counseling process. Wiley, Meinzen-Derr and Choo (2008), argued that an index of development (e.g. developmental quotient) can be used to predict potential difficulties in children with CI. Edwards, Frost, and Witham (2006), studied the practicality of developmental assessment to predict language outcomes up to two years post-implantation. Results of 32 children indicated that in every case, the degree of developmental disability was the best predictor of speech intelligibility and receptive language skills. However, the previously mentioned study by Daneshi and Hassanzadeh (2007), made predictions based not upon developmental status, but developmental checklists that were grouped according to disability type. Although they argued that CI is not contraindicated in prelingually deafened children with
additional disabilities, they noted that severe mental retardation was contraindicated in their program.

Both developmental status and type of additional disabilities, therefore, may provide critical input for caretakers and professionals to consider before and after implantation. However, additional disabilities can vary significantly in their prevalence and in their characteristics. Wiley et al. (2004) found that of the subjects with one or more additional disability, those with two or more comprised a larger proportion than every other individual disability group except for those with motor delay. Furthermore, Rajput et al. (2003) found that in their study of 106 cases, children with diagnosed syndromes had lower speech and language scores 4-5 years after implantation, when compared to those with non-syndromic disabilities. They also found that vision and vestibular impairments may be further negative prognostic indicators for speech and language development.

Just as individual differences in typically developing children affect outcomes after implantation, children with additional disabilities are individually unique and resistant to comparisons. Such differences as cognition, and brain plasticity, are not exclusive to typical children. Marschark and Hauser (2008), poignantly write “Regardless of the source of their differences, such individuals [those requiring special education] are likely to evidence greater diversity than the majority who reside near the center of myriad normal distributions” (p.16). It has been suggested that this greater diversity limits the reliability of prognostic indicators, such as age at time of implantation, degree of pre-implant language exposure, and the presence and type of additional disabilities (Edwards, Frost, & Witham, 2006). Therefore, while developmental status and disability type may alter expectations of language outcomes, their
universal applicability may fall short if individual differences perpetually contradict professional predictions.

This is not to say, however, that specific disabilities have no prognostic value to pre and post implant decisions and expectations. Children diagnosed with autism spectrum disorder (ASD), for example, have common traits with similar implications on parent and professional expectations. Donaldson, Heavner and Zwolan (2004), studied six children with ASD, and results indicated small gains made in comparison to the general population. Whereas spoken language is the goal of most implantations, they found that only one subject in their study used spoken language to communicate. However, they offer an important consideration; when compared to themselves before the implant, these children with ASD made notable improvements in behaviors and interaction as well as overall quality of life. Furthermore, they state that for the parents and professionals working with children with ASD in their study, expectations were not the same as those made for typically developing children. Parents of those with a diagnosis before implantation discussed the possibility that oral communication was not a realistic goal, and subsequent perceptions of benefit where therefore adjusted. However, for those parents who were unaware of a later diagnosed ASD, higher expectations led to perceptions of worse outcomes. Ultimately, with or without a diagnosis before implantation, evaluation of success in children with ASD demands a more subjective approach.

In fact, a different measure of benefit from CI that reflects more positive life changes may be appropriate for the majority of children with additional disabilities (Vlahovic & Sindija, 2004). Hamzavi et al. (2000), argued that measuring success in children with multiple disabilities is often a subjective process, and consideration of family surveys, for example, should compliment objective data for greater insight. Further evidence of this need was provided
by Wiley et al. (2005), who surveyed 19 families with children with developmental disabilities and found that all children made communication progress post implant based on the perceptions of the family. Furthermore, each family said that if presented with the chance, they would choose to implant their child all over again. Donaldson, Heavner, & Zwolan (2004) found that the families they interviewed said they would recommend a CI to others in a similar situation, and gave examples of how their child with ASD responds to the environment, to music and sign language with greater frequency.

**Cochlear Implants in Children with Symptomatic Cytomegalovirus**

It would not be an exaggeration to say that articles on language outcomes in children with cochlear implants and CMV are practically non-existent. In fact, this author was able to find only one source with CMV and cochlear implants as its central focus. In Lee et al. (2005), a retrospective study was conducted of 13 patients with a wide range in ages, duration of deafness, and speech development. Each of the seven males and six females were diagnosed with hearing loss from CMV before they were two years old, three of which had progressive hearing loss leading up to their implantation. The youngest subject to be implanted was 1.2 years, while the oldest was 12.1 years, and the mean age equaled 5.6 +/- 3.6 years. For those who were old enough, a variety of audiometric assessments were administered initially then converted into categories of speech perception. Vocabulary development was assessed using the Peabody Picture Vocabulary Test-Revised, and the One Word Picture Vocabulary Test, while language development was evaluated using Reynell Language Developmental Scales, Clinical Evaluation of Language Fundamentals-Preschool, and the Clinical Evaluation of Language Fundamentals-3, based upon age appropriateness. The majority of subjects had pre-CI audiometric data.
available; however only five of the 13 had follow up data that came roughly one to two years after implantation.

Results of the study by Lee et al. (2005) indicated that some subjects experienced minimal improvement of speech perception, and this was seen as a likely result of their older age of implantation. The authors suggested, however, that they would see improvement steadily as they continue with therapy, and would additionally advance because they were without any other complications related to CMV other than their deafness. Another subject also displayed modest gains in speech perception, although this was linked to his cerebral palsy. Contrary to this, however, another subject with encephalitis and cognitive delay achieved relatively significant growth in receptive language scores.

Out of all 13 subjects in the study by Lee et al. (2005), 73% achieved closed set word recognition, while 64% had open set recognition. Five patients with language scores demonstrated a range of improvements, with the most improvement seen by the three subjects who did not have any other complications related to CMV other than deafness. The first had a moderate receptive vocabulary delay and a mild expressive vocabulary delay post-CI. The second achieved a similar pattern of vocabulary development, and had mild receptive delays and expressive skills within normal limits. The third subject was similar to the second, and also demonstrated receptive and expressive language skills that developed to within normal limits two years after implantation.

It is worth noting the range of additional disabilities that were documented in Lee et al. (2005). These included cardiac anomalies, ophthalmologic manifestations, and motor and cognitive delays. However, cognitive delay and other neurological sequelae did not prevent these subjects from demonstrating significant improvement in speech perception and language
skills. The authors linked all subjects’ successes with the weekly therapies each received, and they argued that early aural habilitation was critical for this population.

In another study, Pyman, Blamey, Lacy, Clark, and Dowell (2000), conducted a retrospective analysis of 75 Australian children who had received cochlear implants before the age of five. The study grouped the children by known cause of hearing impairment and classified them according to evidence of motor and/or cognitive delays, which included CMV. Though no single assessment procedure was used consistently across the sample, results were obtained from detailed analysis of history and various measurements of speech perception. They found that motor and/or cognitive delays had a dramatic impact on the rate of speech development, and children with such delays appeared to begin at lower levels than other children who were not developmentally delayed. According to their analysis, Pyman and colleagues found that CMV was the only cause of deafness that reliably predicted motor and/or cognitive delay. Their research seemed to indicate that the predictive ability of all other etiologies they studied was insignificant and could possibly explain the variability in speech perception.

However, as Lee et al. (2005) demonstrated, not all CMV related deafness resulted in additional neurological sequelae. Even with CNS involvement, positive language growth resulted from CI in all the subjects they tested, and they strongly confirmed that a CI is a safe and effective means to improve communication and quality of life. For those individuals with CNS involvement, the kind of variability that Pyman et al. (2000) documented may continue to be linked with poorer outcomes, which was also observed to an extent in Lee et al. Therefore, if symptomatic CMV is a consistent indicator of poor outcomes, then the variability within this population needs additional study to help delineate the range of factors influencing outcomes.
CHAPTER III

METHODS

Subject Identification

Subjects for this study were between the ages of 36 and 84 months at the time of evaluation. Each was diagnosed with symptomatic CMV and had received a cochlear implant before 40 months of age. Subjects were chosen from a larger study investigating the outcome of deaf children with cochlear implants and developmental disabilities at Cincinnati Children’s Hospital Medical Center. Recruitment was through the hospital’s division of Developmental and Behavioral Pediatrics via letters mailed to their homes, and/or word of mouth through the developmental pediatrician. Parents/caregivers were informed on the purpose of the study and the extent of their involvement. Parents/caregivers signed informed consent to participate.

Subject Characteristics

As detailed in Table 1, a total of six subjects with CMV and CI were included in this study, four females and two males. Age of implantation ranged from one year, one month to three years, two months. The total length of time between implantation and evaluation ranged from four months to four years, two months. Figure 1 illustrates these differences in time spans between important dates.

Table 1: Basic Demographic Information

<table>
<thead>
<tr>
<th>Case</th>
<th>Age (months)</th>
<th>Gender</th>
<th>Ethnicity</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>80</td>
<td>Female</td>
<td>African American</td>
</tr>
<tr>
<td>2</td>
<td>55</td>
<td>Female</td>
<td>African American</td>
</tr>
<tr>
<td>3</td>
<td>51</td>
<td>Female</td>
<td>African American</td>
</tr>
<tr>
<td>4</td>
<td>49</td>
<td>Male</td>
<td>Caucasian</td>
</tr>
<tr>
<td>5</td>
<td>38</td>
<td>Male</td>
<td>Caucasian</td>
</tr>
<tr>
<td>6</td>
<td>27</td>
<td>Female</td>
<td>Caucasian</td>
</tr>
</tbody>
</table>
Materials/Procedures

Language evaluations of case subjects were conducted between August 2007 and February 2009. All participants were administered the Preschool Language Scale – 4th edition (Zimmerman, Steiner, & Pond, 2002) by a supervised speech and language pathology graduate student. The PLS-4 assessed the child’s receptive and expressive language abilities. The Auditory Comprehension subscale (receptive language) assessed basic receptive vocabulary, and understanding of concepts, grammatical markers, complex sentences, comparisons and inferences. The Expressive Language subscale assessed the child’s ability to name objects, use concepts of description, express quantity, use grammatical markers, and produce multi-word utterances.

Parent/caregiver involvement was a necessary asset during the evaluation, as the PLS-4 protocol allowed for parent/caregiver input on a variety of questions. Another asset of the PLS-4 was that it provided a large age range (up to 6 years, 11 months) and allowed for a modified administration for special populations including children with hearing impairments. The
accommodations given in the examiner’s manual allow use of the norm-referenced test scores (standard scores, percentile ranks, and age equivalents) when using an interpreter. During the evaluation a hospital interpreter was present if the child used or benefited from sign language. In the event an interpreter was used, two language scores were reported, one for verbal stimulus only, and another for verbal and sign (total communication). Stimulus was presented verbally first, and repeated in sign, if needed. Since American Sign Language can be iconic, Conceptually Accurate Signed English was used for those test items with high iconicity.

Results of each evaluation using the PLS-4 provided norm-referenced test scores as well as age-equivalents. Because the lowest possible standardized score on the PLS-4 is 50, this study focused on the language age-equivalents given by the PLS-4. Age-equivalents were also normalized by dividing the language age-equivalents given by the PLS-4 with the child’s chronologic age at time of testing and multiplying by 100. Language quotients close to 100 indicate that a child’s language level is age-appropriate, while quotients below 80 indicate that a child’s language level is significantly delayed.

Results for each evaluation were then compared to information obtained through chart review. This information included medical histories, pre-implant development and interventions they were receiving. Developmental information included developmental quotients from the Revised Gesell Developmental Schedules (Knobloch, Stevens, & Malone, 1980). The Revised Gesell was administered by the developmental pediatrician previous to the child receiving the cochlear implant, and thus provided a brief, but appropriate indication of the developmental level at which the child was functioning prior to receiving the cochlear implant. Also obtained through chart review were pre-implant language age equivalencies taken from the Rossetti Infant-Toddler Language Scale (Rossetti, 2006). However, these scores were only available for four of the six
cases. Age equivalents were also normalized through language quotients in the same manner as those obtained from PLS-4. This allowed comparisons between pre and post-implant language abilities for these four case subjects.
CHAPTER IV

Results

A qualitative study of six children with cochlear implants (CI) and symptomatic cytomegalovirus (CMV) found severely delayed abilities in both receptive and expressive language. With the exception of case 5, receptive and expressive standard scores on the PLS-4 were all below 50 (<1%). Each case, including case 5, demonstrated age-equivalents that were 20 months or more below their chronological age. In addition, all six cases had language quotients that were below 80, indicating receptive and expressive language abilities significantly below the average range. Individual results are listed in Tables 2 and Table 3, followed by a summary of results for each case, with a review of pertinent developmental information and pre-implant language scores (if available).

Table 2: Receptive scores obtained from the PLS-4 for all cases post-implant.

<table>
<thead>
<tr>
<th>Case</th>
<th>S.S.</th>
<th>S.S. w/Sign</th>
<th>LA</th>
<th>LA w/Sign</th>
<th>LQ</th>
<th>LQ w/Sign</th>
</tr>
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<tbody>
<tr>
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<td>50</td>
<td>13</td>
<td>20</td>
<td>16.3</td>
<td>25</td>
</tr>
<tr>
<td>2</td>
<td>50</td>
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<td>5</td>
<td>n/a</td>
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<td>n/a</td>
</tr>
<tr>
<td>3</td>
<td>50</td>
<td>n/a</td>
<td>10</td>
<td>n/a</td>
<td>19.2</td>
<td>n/a</td>
</tr>
<tr>
<td>4</td>
<td>50</td>
<td>50</td>
<td>8</td>
<td>15</td>
<td>16.3</td>
<td>30.6</td>
</tr>
<tr>
<td>5</td>
<td>50</td>
<td>50</td>
<td>13</td>
<td>17</td>
<td>34.2</td>
<td>44.7</td>
</tr>
<tr>
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<td>2</td>
<td>n/a</td>
<td>7.7</td>
<td>n/a</td>
</tr>
</tbody>
</table>

S.S. = Standard Score  L.A. = Language Age Equivalent  L.Q. = Language Quotient

Table 3: Expressive scores obtained from the PLS-4 for all cases post-implant.

<table>
<thead>
<tr>
<th>Case</th>
<th>S.S.</th>
<th>S.S. w/Sign</th>
<th>L.A.</th>
<th>L.A. w/Sign</th>
<th>L.Q.</th>
<th>L.Q. w/Sign</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
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<td>50</td>
<td>16</td>
<td>19</td>
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</tr>
<tr>
<td>2</td>
<td>50</td>
<td>n/a</td>
<td>11</td>
<td>n/a</td>
<td>20</td>
<td>n/a</td>
</tr>
<tr>
<td>3</td>
<td>50</td>
<td>n/a</td>
<td>9</td>
<td>n/a</td>
<td>17.2</td>
<td>n/a</td>
</tr>
<tr>
<td>4</td>
<td>50</td>
<td>50</td>
<td>9</td>
<td>12</td>
<td>18.4</td>
<td>24.5</td>
</tr>
<tr>
<td>5</td>
<td>68</td>
<td>74</td>
<td>23</td>
<td>26</td>
<td>60.5</td>
<td>68.4</td>
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<td>50</td>
<td>n/a</td>
<td>8</td>
<td>n/a</td>
<td>28.6</td>
<td>n/a</td>
</tr>
</tbody>
</table>

S.S. = Standard Score  L.A. = Language Age Equivalent  L.Q. = Language Quotient
Case 1: female, 6.6 years.

Hearing History: Case 1 was diagnosed with severe-profound sensorineural hearing loss (SNHL) around 10 months of age, and subsequently received her CI about 19 months later when she was two years, four months of age. At the time of the evaluation, case 1 was six years, six months old, and had been wearing her CI for four years.

Developmental and Intervention History: Case 1 has a diagnosis of cognitive delay and an additional history of left cortical dysplasia, spastic quadriplegia, and seizures. Her developmental quotient was 64 prior to implantation. At the time of the evaluation she was receiving weekly speech and language therapy (ST) at school for half hour sessions. She also was receiving half hour sessions of occupational therapy (OT), and physical therapy (PT), both at school and at home. Case 1 was also receiving visual therapy (time unspecified), but not aural rehabilitation.

Current Receptive Language Abilities: At the time of testing (48 months after implant) case 1 had a chronological age of 80 months with a receptive language age of 13 months without sign, and 20 months with sign on the PLS-4. Her calculated receptive language quotient was 16.3 without sign, and 25 with the use of sign. Case 1 was able to follow simple routines and point to a few familiar objects without the use of sign language. She was able to identify objects more consistently with the use of sign. Also with sign, case 1 was able to understand verbs in contexts (e.g. The bear is thirsty. Give him something to drink).

Current Expressive Language Abilities: At the time of testing, case 1 had an expressive language age of 16 months without sign, and 19 months with sign. Her expressive language quotients from the PLS-4 were 20 without sign, and 23 with signed responses. Case 1 communicated primarily through gestures and behavior, although she was also able to use a few signs. Her
mother reported that she could say “mama” and “bye.” While she was able to produce /m/, and /b/ and imitate signs, she was unable to imitate words verbally. At the time of this evaluation it was reported that she was able to use around 30 different signs. Noted in the report was case 1’s interactive personality and frequent calls to another’s attention with points and vocalizations (e.g. grunts).

Language Growth Since Pre-Implant Evaluation: No pre-implant information available.

Case 2: female, 4.6 years.

Hearing History: Case 2 was three months old when she was diagnosed with profound SNHL. She received her CI about 35 months later when she was three years, two months. At the time of the evaluation she was four years, six months and had been wearing her CI for 16 months.

Developmental and Intervention History: Case 2 has a diagnosis of mild cognitive delay, and has a history of patent ductus arteriosis (e.g. enlarged heart), and seizures. Her developmental quotient was 42 prior to implantation. At the time of the post implant evaluation, Case 2 was attending a preschool for children with disabilities, and was receiving ST, OT, and PT in the school each week for a half hour per therapy. Case 2 was also receiving aural rehabilitation at school and visual therapy at home (time unspecified).

Current Receptive Language Abilities: At the time of testing (16 months after implant) case 2 had a receptive language age of five months with a chronological age of 54 months on the PLS-4, and a calculated receptive language quotient of 9.1. She did not use supplementary sign. Receptive skills were assessed mostly through parent report and observation. Although she typically looked at the speaker during communication, she did not turn to locate the source of sound, or respond when her name was called. Case 2 therefore was limited in receptive abilities,
and her mother reported that she was unsure whether her daughter was able to hear anything from the CI.

**Current Expressive Language Abilities:** At the time of testing, case 2 had an expressive language age of 11 months on the PLS-4, and a calculated expressive language quotient of 20. Case 2 did not vocalize during the evaluation, however her mother reported that she does vocalize when pleased or frustrated, and does so with different variations in pitch and loudness. Her mother also reported that she was able to communicate non-verbally with gestures and other behavior, and will initiate a turn taking game (e.g. peek-a-boo). No evidence of consonants, and only the vowel /a/ was heard or reported.

**Language Growth since Pre-Implant Evaluation:** A period of 19 months elapsed between pre and post-implant testing. Over this time frame a gain of five months was achieved in expressive age (6 months-11 months). Expressive language quotient showed an increase of three points (17-20) over the same period. No pre implant receptive score information was available. Case 2 did not require an interpreter during the evaluation and did not use sign language.

**Case 3: female, 3.2 years.**

**Hearing History:** Case 3 was diagnosed with severe to profound SNHL at one month of age and received a CI at 13 months of age. At the time of the evaluation, case 3 was four years, three months old, and had been wearing her CI for about three years, two months.

**Developmental and Intervention History:** Birth history is notable for mother’s report of decreased fetal movement just prior to term birth. She was diagnosed with cerebral palsy and cognitive delay. Additional history includes seizures, microcephaly, oral-motor delay, spastic quadriplegia, cortical dysplasia. Her pre implant developmental quotient was 50. Case 3 was
receiving one hour treatment sessions of ST, OT and PT once a week at her school. She was not receiving aural rehabilitation.

Current Receptive Language Abilities: At the time of testing (38 months after implant), case 3 had a receptive language age of 10 months with a chronological age of 51 months on the PLS-4, and a calculated receptive language quotient of 19.2. Case 3 demonstrated sensory awareness of sound by interrupting activity when her name was called, and actively searching for the source of a sound when not visible. Case 3 attended to objects called to her attention, and responded appropriately to familiar phrases (e.g. give me a kiss). She also showed emerging skills in her ability to identify photographs of some familiar objects and could follow familiar directions. Case 3 was unable to identify familiar objects from a group of objects, respond appropriately to inhibitory words (e.g. wait), and did not show comprehension of more advanced language such as verbs in context.

Current Expressive Language Abilities: At the time of testing, case 3 had an expressive language age of 9 months, and a calculated expressive language quotient of 17.2. Case 3 was able to vocalize sounds suggesting pleasure and displeasure, and vocalized along with gesture when protesting. Case 3’s mother reported she was able to initiate a turn-taking game, and request objects or actions through gesture and vocalizations. One of Case 3’s primary means of communicating was through the use of eye gaze, however she was unable to label objects or photographs of familiar objects using this method of communication. Case 3 did not have the ability to produce at least two consonants, and did not demonstrate any babble like vocalizations.

Language Growth since Pre-Implant Evaluation: Between pre and post implant evaluations, 45 months had passed. Receptive age grew by seven months (3 months - 10 months), while expressive age grew by six months (3 months - 9 months). In terms of language quotients,
however, receptive skills declined by 23.7 (42.9-19.2) points, and expressive skills dropped by 25.7 points (42.9-17.2). An interpreter was used for all post implant testing.

**Case 4: male, 4.0 years.**

**Hearing History:** At one year, three months of age, case 4 was diagnosed with profound SNHL. He received his CI five months later when he was one year, eight months of age. Case 4 was four years old when he was evaluated with the PLS-4, and had been wearing his CI for two years, four months.

**Developmental and Intervention History:** Case 4 has a diagnosis of cognitive delay and has a history of microcephaly, cortical dysplasia, and mild hypotonia. Case 4’s pre-implant developmental quotient was 70. Case 4 was receiving ST, OT, and PT in the school once a week for an hour, and was receiving private ST three times a week at home for an hour each time. Case 4 also was receiving aural rehabilitation each week at home for an hour.

**Current Receptive Language Abilities:** At the time of testing (28 months after implant), case 4 had a chronological age of 48 months, with a receptive language age of eight months without sign, and 15 months with sign on the PLS-4. His calculated receptive language quotient was 16.3 without sign, and 30.6 with sign. Case 4’s receptive language skills without the use of sign were limited to responding to his name, and following simple verbal directions (e.g. come here). When sign was added, case 4 was able to look at objects that were called to his attention, and identify familiar objects from a group of objects. Case 4 was unable to follow routine or familiar directions with cues. With or without sign, case 4 was not able to identify photographs of familiar objects, understand inhibitory words, identify body parts, or understand verbs in context. His mother reported that he understands more than 100 signs.
**Current Expressive Language Abilities:** At the time of testing, case 4 had an expressive language age of 9 months without sign, and 12 months with sign. His calculated expressive language quotient was 18.4 without sign and 24.5 with the use of sign. Case 4’s vocalizations consist mainly of /mmm/, /ahhh/ and /uuh/, and can use two different consonant sounds, /m/ and /g/. Case 4 was able to combine sounds to form one syllable, /ma/. Even though he is able to produce /ma/, his mother reported that it did not indicate her or any other person at this time. Although case 4 did not yet have any words verbally, he was able to express more than 100 signs, and imitate other signs for such things as “ball”, and “cookie.” Case 4 was able to use sign to name at least one object in a photograph and his mother reported that he used sign to request toys, label actions, and indicate when he wants something to happen again. Case 4, however, did not use sign to ask questions, or combine multiple signs to create a multi-word utterance.

**Language Growth Since Pre-Implant Evaluation:** A total period of 33 months had passed between pre and post implant language evaluations. Over this time, case 4’s language age equivalent gained nine months receptively (6 months -15 months), and nine months expressively (3 months -12 months). Receptive language quotients declined by 6.9 points (37.5-30.6) and expressive language quotient grew by 5.7 points (18.8-24.5). A sign language interpreter was used for all post implant testing.
Case 5: male, 3.2 years.

Hearing History: Case 5 was diagnosed with profound SNHL at one month of age, and received an implant when he was 13 months old. At the time of the post implant evaluation, case 5 was three years, two months of age, and had been wearing his CI for two years.

Developmental and Intervention History: Case 5, had a developmental quotient of 83 prior to implantation. He has a history of gross motor delays related to a diagnosis of ataxic cerebral palsy. He also had apneic episodes related to gastric-esophageal reflux. Case 5 attends a school for children with hearing impairments. He was receiving two hour sessions of ST twice a week in the school setting, and private ST once a week for an hour. He also has received aural rehabilitation once a week for an hour, two half hour sessions of OT per month, and visual therapy twice a year.

Current Receptive Language Abilities: At the time of testing (24 months after implant) case 5 had a chronological age of 38 months, with a receptive language age of 13 months without sign, and 17 months with sign on the PLS-4. His calculated receptive language quotient was 34.2 without sign, and 44.7 with sign. Case 5 demonstrated the ability to detect and orient to sound sources, and could reliably pinpoint the speaker who called his name. Case 5 was able to respond to specific words or phrases, and follow simple routines, (e.g. putting a ball in a box) without sign. When provided with the written word, case 5 could identify body parts (e.g. nose, eyes) and could identify specific clothing (e.g. shirt, shoes). Reportedly, case 5 could read 40 words, and can point to familiar objects if the word is written down for him. With or without sign, Case 5 was unable to identify familiar objects from a group of objects, and understand verbs in context (e.g. The bear is thirsty. Give him something to drink).
Current Expressive Language Abilities: At the time of testing, case 5 had an expressive language age of 23 months without sign, and 26 months with sign. His calculated expressive language quotient was 60.5 without sign, and 68.4 with sign. Case 5 produced a variety of consonant sounds including /t/, /d/, and his mother reported he is working on /r/, /l/, /sh/, and /ch/. Case 5’s vowels appeared to be very accurate. Case 5 spoke primarily with single words (e.g. ball) and was able to imitate words (e.g. duck) and answer ‘what’ and ‘where’ questions such as (e.g. where is he?) Using sign, case 5’s mother reported he could ask questions (e.g. “where is dad”), request items and indicate that he wants more of something (e.g. more cookies). Through sign language, his mother also reported that he was able to use possessive pronouns (e.g. my) and 2-3 word combinations (e.g. school + learn).

Language Growth Since Pre-Implant Evaluation: Over approximately 15 months, case 5 gained 11 months in receptive language age (6 months - 17 months), and 20 months in expressive language age (6 months - 26 months). According to pre and post language quotients, a decline of 9.8 points occurred in receptive language; however, a gain of 13.9 points occurred in expressive language quotients. A sign language interpreter was used for the post implant evaluation.

Case 6: female, 2.4 years.

Hearing History: Case 6 was diagnosed with profound SNHL at two months and received her CI when she was two years old. At the time of the evaluation, case 6 was two years, four months of age, and had only been wearing her CI for four months.

Developmental and Intervention History: Case 6 was diagnosed with global developmental delay, with additional diagnoses of microcephaly, mild brachiocephaly, and concerns for eye deviations that could be related to seizures. She had a developmental quotient of 41 prior to
receiving an implant. At the time of the post implant evaluation, case 6 was receiving ST, OT and PT therapy through an early intervention program since she was four months (length of sessions unspecified). Case 6 was evaluated only four months after she received her implant, and arrived for the language evaluation after having her CI re-calibrated by the audiologist.

**Current Receptive Language Abilities:** At the time of testing (four months after implant), case 6 had a receptive language age of 2 months with a chronological age of 28 months on the PLS-4, and a calculated receptive language quotient of 7.7. Case 6 was able to startle from a sound, though she will not attempt to find the source. Although she did not interrupt her activity when her name was called, case 6’s mother reported she showed anticipatory behavior when playing chase with her father. Case 6 did respond to “no”, but was not reported to respond to any other verbal communication.

**Current Expressive Language Abilities:** At the time of testing, Case 6 had an expressive language age of 8 months, and a calculated expressive language quotient of 28.6. Case 6 was able to produce limited speech sounds (/uh/, /oh/, /ah/, /m/) and her mother reported she used vocalizations for communicative purposes (e.g. protest, attention seek). Case 6 communicated nonverbally by using pushing and pulling gestures with the toys. There was no evidence of consonant sounds, or babble-like vocalizations.

**Language Growth Since Pre-Implant Evaluation:** No pre implant information available.
CHAPTER V

Discussion

Summary

This study investigated the language outcomes of children with cochlear implants (CI) and symptomatic cytomegalovirus (CMV). The study itself and its participants are a subset of a larger study investigating children with CI and additional disabilities. Each of the six cases participated in a post implant language evaluation using the PLS-4. All subjects demonstrated significantly delayed receptive and expressive language skills represented by age equivalents and language quotients. Results of four cases with pre implant language data showed receptive and expressive language growth in age equivalents; however language quotients indicated receptive declines and only mild expressive gains. Cases communicated primarily through sign, or behavior. Only two participants demonstrated communicative speech, and both continued to rely on sign or behavior for most of their communications.

Post-Implant Language Abilities

As a group, results were variable across all six cases, which is consistent with findings from the literature describing wide variability in post-implant language outcomes. As Pisoni et al (2008) argued, variability is a fundamental part of outcomes. While there were some similarities between scores (e.g. all language quotients below 80), the range of scores was wide and indicative of a myriad of individual differences. Age of implantation, length of sensory deprivation prior to implant, and the length of time between implantation and evaluation are all factors that likely had enormous implications on individual outcomes. It can also be assumed that each participant’s receptive and expressive language abilities are the result of demographic influences (e.g. intervention type and frequency).
While cautious interpretation is warranted because of numerous confounders, a discussion of variability between subjects may facilitate a deeper understanding of this population. One aspect that varied from case to case was the degree to which receptive and expressive skills were unequal. Half of the cases achieved post-implant scores that were stronger receptively, while the other three cases demonstrated stronger expressive skills. For the three cases that used supplemental sign during the post implant evaluation, the discrepancy between receptive and expressive age-equivalents and language quotients changed when scores included signed answers. However, the way in which these changed varied for each participant. For example, the gap between case 1’s receptive and expressive age-equivalents was smaller when scores included signed responses. This was not the case, however, for case 4, who had a larger gap between receptive and expressive age equivalents after scores included signed responses.

Variable language gains were also observed for four subjects with pre-implant data available. From the perspective of age-equivalents, all subjects grew both receptively and expressively between pre and post implant evaluations. However, language quotients show receptive language declines for three of the four cases, with an average of 14 points being lost between evaluations. Expressive quotients actually show the opposite trend for every one of the four cases, except case 3, who lost 25.7 points between evaluations. The difference in perspective between age-equivalents and language quotients is likely due to the comparative nature of quotients, which indicate how appropriate an individual’s language age is to their chronological age. Because language development involves an exponential accumulation of skills, the degree to which a child is behind may grow even when age equivalencies steadily progress.
There is no easy explanation, however, that would explain why three cases had receptive quotients decline at the same time their expressive quotients rose. Perhaps the adaptations each has made in the form of alternative communication (e.g. gesture, sign) have provided support for more complex communication, while the same has not been true for comprehension skills. Expressive language may also have received a boost from the new sensory awareness of their own voice as a means to an end, and that the CI in conjunction with other methods (e.g. sign, behavior) gave language production the support that language comprehension has yet to utilize because of cognitive, physical and developmental limitations. Their expressive efforts may have received additional support from successful communicative exchanges with individuals who have the ability to infer meaning (e.g. parents, other adults). Their comprehension, however, of similar communicative attempts could be beyond their current abilities. In other words, with a variety of tools at their disposal, the expression of simple linguistic concepts is less cognitively demanding than the comprehension of another’s expression of the same.

One further possibility behind stronger expressive scores could be hidden within the test itself. Future task analysis could expose error patterns that resulted from the unequal demands that the test protocol placed upon receptive and expressive domains. Lower receptive scores could have resulted merely from the lack of opportunity to demonstrate a broader range of abilities. While he/she may have no problem spontaneously labeling an item with a sign or vocal approximation, understanding multiword directions, even when signed, may be beyond his/her capabilities. Furthermore, it is not unreasonable to think that some of the expressive scores of these CI subjects could be the result of the evaluator’s generous interpretation of communicative attempts of participants during the evaluation, and that receptive scores are lower simply because the same flexibility is not possible in a young child with a cognitive impairment.
Relationships between Additional Disabilities and Language Outcomes

In their own study of CI in children with CMV, Lee et al. (2005) found that some children with cognitive delays related to CMV made significant gains in speech perception and language skills. While follow-up periods were longer within their research, the present study did not observe the same level of success in the five case subjects with cognitive delays. Case 2 for example, has a similar profile to one particular subject in Lee et al., as both have a cognitive delay and implantation age around 3.5 years. Although the length of time after implantation was shorter, Case 2’s receptive language outcomes were significantly lower than those measured in Lee et al, who achieved open set word recognition. Case 2 had yet to orient to the source of a sound by the time she was evaluated. Interestingly, both subjects received the same type of intervention services at similar frequencies including aural rehabilitation, which Lee et al. argued was critical to successful outcomes.

Another point of interest between Lee et al. and the present study is the type of communication mode used by cases with similar disability types. In Lee et al. (2005), each subject who had cognitive or developmental delays used total communication, and those who were without additional neurological impairments other than deafness, used strictly oral communication. The same was true for some of the cases in the present study; however, subjects such as case 2 had yet to begin to use the total communication approach they were being taught at school.

In this present study, the only case to demonstrate significant progress was also the only case without a cognitive delay. In both language age equivalents and language quotients, case 5 demonstrated the most growth out of the six participants. Like the majority of the participants, case 5 had confirmed brain calcifications, and a history of epileptic seizures. Case 5, however,
was without the additional abnormalities that were noted in other subjects, including vestibular asymmetries, unspecified focal abnormalities, and cortical dysplasia, which likely contributed to cognitive deficits and/or global developmental delays. It is also important to note that prior to implantation, case 5 received a developmental quotient of 83, and is the only case to have a developmental level close to the average range. Case 5, therefore, potentially represents a subset of the population with symptomatic CMV that have greater pre implant potential based upon developmental level and the absence of certain impairments (e.g. microcephaly).

Case 4’s developmental quotient of 70 was not far below the 83 that case 5 had before implantation. At the time of the evaluation, case 4 also had four more months of experience with the CI than did case 5. However, case 4 did not achieve the same level of functional language that was observed in case 5; in fact his expressive language quotient was less than half of case 5’s. In this comparison, case 4’s cognitive impairment may be related to his relatively lower scores. Like cases 3 and 6, case 4 had a history of microcephaly, which was shown by Pass et al. (1980) to consistently impair neurological development. For case 4, as well as cases 3 and 6, the diagnosis of microcephaly, compounded by the sensory deprivation of his deafness, may have imposed significant limitations in their ability to utilize the degraded acoustic representation of sound that the CI provides. In situations where children like case 4 are eligible for implantation, careful consideration of the child’s additional disabilities and the impact they may have on outcomes may assist in establishing realistic expectations.
Limitations of the Present Study

Consistent with the nature of qualitative research designs, this study likely fails to reflect the general population of children with symptomatic CMV and CI, or the full spectrum of CMV sequelae that impact language outcomes. Furthermore, results of this study are a small glimpse into the even greater variability that is likely to be found in such a large but discrete population.

Additional limitations in research design include the use of two test protocols to evaluate pre and post-CI language abilities. In comparing results from two different tests, (the Rossetti Infant Toddler Scale for pre-CI data, and the PLS-4 for post-CI data) the interpretation of language growth may be less convincing. The addition of more naturalistic assessment, including repeated measures over a variety of the subjects’ daily routines could have provided more evidence for or against skills that were observed during the short evaluations.

Perhaps one of the more serious limitations to this study, are the inconsistent intervals between critical points in time, particularly the lengths between pre and post-CI evaluations. Controlling for this with continuing measures of the same protocol may be needed for future studies with the intention of demonstrating benefits of CI. With a population such as the one included here, a developmental delay may necessitate more longitudinal measures to allow sufficient time to fulfill potential growth. Including measurements of hearing acuity, or data concerning individual hearing levels over time, could also provide more insight into the specific gains or lack thereof, in receptive and expressive language.

Future Research

In order to bridge the gap between research and practice, it seems the traditional and ongoing approach is to investigate intervention types and educational settings, demographic and medical variables, and the degree of parental involvement. However, as variability in outcomes
and individual differences will likely continue to limit the capability of research to determine efficacy of CI in children with additional disabilities, focus may need to expand and collaborate with leading researchers in the domains of cognitive and neural sciences. Without understanding the cognitive underpinnings that create or inhibit success with a CI, traditional variables may only contribute to the nebulous ranks of important considerations parents and professionals can turn to when deciding whether or not to implant a child. What may ultimately provide the predictive clues into a child’s response to CI, are the complexities yet to be understood in the automatic and controlled processes linked to language comprehension and production by current research. Understanding how individual neurological impairments limit neuroplasticity and adaptation to the degraded presentation of acoustic signals may help explain the kind of poorer outcomes observed in this study.

Conclusion

Ultimately, across five of the six cases were variable degrees of limited language abilities, while the only case without a cognitive delay achieved the highest level of language functioning. Although three cases demonstrated an increase in expressive language between pre and post-implant evaluations, these gains were not necessarily within spoken language. All cases utilized either sign or communicative behavior and, with the exception of case 5, demonstrated little to no spoken language. While these results suggest that significant cognitive impairments likely limit language outcomes, what are unfortunately missing from these results are the subjective accounts of the benefits that a CI has provided. As research continues, parents and professionals will need to remain patient and strive for greater cooperation to ensure that research obtains clinically meaningful information that can be translated to the caretakers during and after the decision to implant their child. Likewise more subjective measures of post-implant
outcomes that validate parent perspectives could strengthen this parent-professional relationship and increase their willingness to participate in research.
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