Children with cochlear implants and developmental disabilities: A language skills study with developmentally matched hearing peers

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ABSTRACT

The number of children receiving cochlear implants (CIs) with significant disabilities in addition to their deafness has increased substantially. Unfortunately, children with additional disabilities receiving CIs have largely been excluded from studies on cochlear implant outcomes. Thus limited data exists on outcomes in this population to guide pre-implant counseling for anticipated benefits. The study objectives were: (1) evaluate differences in post-cochlear implant language skills between children with cochlear implants and developmental disabilities and age/cognitively matched controls; (2) quantify possible discrepancies between language level and cognitive level. Fifteen children with a developmental disability who received a CI were matched 1:1 on nonverbal cognitive ability and age to hearing controls. Language was evaluated using Preschool Language Scale-IV and reported as language quotients. Multivariable mixed models for matched pairs analyzed differences in language levels between groups. No significant differences were seen between CI and control groups regarding insurance, maternal education, or family income level. Results of the multivariable models indicated that compared to matched controls, the CI group had significantly lower mean receptive (24.6 points, \( p = 0.002 \)) and mean expressive (21.9 points, \( p = 0.001 \)) language quotients after controlling for confounders such as number of therapies and weekly hours in therapy. Significant discrepancies between language level and cognitive level were seen among CI participants only. Compared to age- and cognitively matched controls, children with CIs had significantly lower language levels with delays disproportionate to their cognitive potential. Mechanisms behind this performance-functional gap need to be understood to deliver appropriate intervention strategies for this special population.

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1. Introduction

Hearing loss is one of the most common pediatric health conditions in the United States. Moderate to profound bilateral hearing loss is identified in 2–3 infants per 1000 births, increasing to approximately 6 per 1000 children by school age (American Speech-Language-Hearing Association, 2010; Centers for Disease Control and Prevention, 2010; National Institute on Deafness and other Communication Disorders). Approximately 30–40% of children with sensorineural hearing loss demonstrate additional or multiple disabilities that can have profound effects on communication and related cognitive,
A cochlear implant (CI) is a widely embraced technological device used for the deaf child’s auditory system to gain access to a quality of sound experience not available with hearing aids alone. Studies addressing language development of children with implants at early ages (≤36 months old) have found that the rate of language development after a CI exceeded that expected from deaf children without an implant, with the most rapid language growth occurring among children who received the CI at the youngest ages (Anderson et al., 2004; Fryauf-Bertschy, Tyler, Kelsay, Gantz, & Woodworth, 1997; Geers, 2004; Kirk et al., 2002; McConkey Robbins, Koch, Osberger, Zimmerman-Phillips, & Kishon-Rabin, 2004; Miyamoto, Kirk, Svirsky, & Sehgal, 1999; Osberger, 1997; Svirsky, Teoh, & Neuburger, 2004; Tomblin, Barker, Spencer, Zhang, & Gantz, 2005; Waltzman & Cohen, 1998). In addition, upon receiving an implant, language growth rates of children are close to rates of children with normal hearing (Bollard, Chute, Popp, & Parisier, 1999; Kirk et al., 2002; McConkey Robbins et al., 2004; Svirsky & Meyer, 1999; Svirsky, Robbins, Kirk, Pisoni, & Miyamoto, 2000; Svirsky et al., 2004), with the biggest leap in language development happening during the first year post-implant (Cheng, Grant, & Niparko, 1999; Tomblin, Spencer, Flock, Tyler, & Gantz, 1999). While children with CIs approach the language development of their hearing matched peers, language delays may continue to exist in some children post-implant (Bollard et al., 1999; Geers, 2004; Manrique, Cervera-Paz, Huarte, & Molina, 2004; Miyamoto, Svirsky, & Robbins, 1997; Stacey, Fortnum, Barton, & Summerfield, 2006). In the early years of pediatric cochlear implantation, it was typical for children with known disabilities to be considered unsuitable for the procedure. Although the number of children with additional disabilities who are receiving cochlear implants has been increasing over the years (Edwards, 2007), appropriate outcomes in this population are still relatively unknown.

The impact of hearing loss on children with developmental disabilities or delays has never truly been quantified, yet it has been assumed to be profound. Until recently, most research on deafness and additional disabilities have been qualitative (e.g., surveys, case studies, observations). The few quantitative studies on children with cochlear implants and disabilities have reported on a variety of outcomes, often regarding speech perception or intelligibility (Daneshi & Hassanazadeh, 2007; Dettm et al., 2004; Edwards, Frost, & Witham, 2006; Hamzavi et al., 2000; Nikolopoulos, Archbold, Wever, & Lloyd, 2008; Pyman, Blamey, Lacy, Clark, & Dowell, 2000; Vlahovic & Sindija, 2004; Waltzman, Scalchunes, & Cohen, 2000). Control populations, when available, consist of typically developing children (Holt & Kirk, 2005; Nikolopoulos et al., 2008; Pyman et al., 2000), which are not necessarily appropriate comparisons for this particular group of children. Children with developmental disabilities who received cochlear implants do not meet their typically developing peers in auditory skill development, speech perception, or language skills. Unfortunately, control populations of typically developing children with implants will never help us understand the skills set we would expect to see in context of the developmental concerns of the child.

In light of the lack of developmentally appropriate control groups for children with cochlear implants and additional disabilities, the current study utilized a design that allowed for a control population of hearing children with disabilities. Our cochlear implant center has routinely implemented an evaluation by a developmental pediatrician since 2001 which has been discussed in detail previously (Wiley, Meinzen-Derr, & Choo, 2008). Anticipated expectations for child outcomes are discussed candidly with families during this pre-implantation evaluation. The addition of a developmental pediatrician has allowed the otologists, speech-language therapists, audiologists, and aural rehabilitation therapists an increased comfort level in serving children with additional disabilities. It has also allowed for continuity of care and guidance in altering strategies for interventions as children continue to follow up with the team’s pediatrician. Being extremely aware of the needs for outcomes research in this population of children, the objectives of this study were (1) to evaluate the differences in post-implant language skills among children with cochlear implants and developmental disabilities as compared to hearing children who were matched on age and cognitive abilities; and (2) quantify the gap between language abilities and cognitive abilities in this population.

2. Materials and methods

2.1. Participants

Children identified with a developmental disability who were ≤6 years of age were eligible for the study. Children with a cochlear implant were identified through a clinical cochlear implant registry. Hearing children (controls) with similar age and developmental abilities were identified through a review of clinical charts within the Division of Developmental and Behavioral Pediatrics. Parents of eligible study participants were contacted by letter and follow-up phone call. Parents could also actively contact study personnel through information listed on advertisements posted throughout the medical center. All enrolled participants had completed developmental evaluations by 3 years of age. Children with hearing were matched (1:1) to children with cochlear implants within 12 months of age and within 5 quotient points (per nonverbal cognitive assessment). The nonverbal cognitive abilities, over chronologic age, were considered the priority regarding matching criteria. This study was approved by the institution’s Institutional Review Board. Consent was obtained from all parents prior to study participation.

2.2. Developmental evaluation

All children had been evaluated by a developmental pediatrician prior to the study using the Revised Gesell Developmental Schedules (Ball, 1977). This tool is routinely administered to children under the age of 3 years who are seen

for a multi-factorial developmental evaluation, which included the evaluation required prior to receiving a CI. The scale consists of five sub-categories of skills: gross motor, fine motor, nonverbal cognitive performance, personal-social, and language. Developmental quotients are derived by dividing the age-equivalent level of each domain on the Revised Gesell by the child’s chronologic age at the time of the developmental assessment. A nonverbal cognitive quotient (NVCQ) was derived from the cognitive performance domain. A non-verbal or performance-based IQ was used for one control participant.

Children’s developmental delays were classified according to all developmental domains. Children with either a gross motor or fine motor delay were described as having a motor delay. The diagnosis of cerebral palsy was based on specific neurologic exam (patterns of persistent primitive reflexes, abnormalities in tone and reflexes, presence of abnormalities on MRI of the brain) by the developmental pediatrician paired with MRI findings. The presence of vision impairment was also indicated (based on definition of visually impaired or legally blind). Children with low vision were defined as having significant vision loss that is unable to be corrected by glasses. No children in the study were considered legally blind, which was defined as having visual acuity in the better eye that with correction was not more than 20/200 or a defect in the visual field of less than 20° field in the widest diameter.

2.3. Language evaluation

Per the study protocol, receptive (auditory comprehension) and expressive language was assessed using the Preschool Language Scales – 4th edition (PLS-4) (Zimmerman, Steiner, & Pond, 2002). This language assessment tool is designed to be used with children from birth through 6 years 11 months of age and provides norm-referenced test scores as well as age-equivalents. The PLS-4 auditory comprehension subscale targets skills that are important precursors for language development (e.g., attention to speakers, appropriate object play), comprehension of basic vocabulary, concepts, grammatical markers, and the ability to understand complex sentences and make comparisons and inferences. The expressive communication subscale addresses vocal development and social communication, naming common objects, the use of concepts that describe objects, express quantity, prepositions, grammatical markers, sentence structures, and examines pre-literary skills (i.e. phonological awareness tasks, ability to tell a short story in sequence). Because the PLS-4 was used as a language measurement and not merely as a measure of spoken language, both auditory-oral and signed responses were scored and reported as standard scores as well as age-equivalents. Because a standard score of 50 is the floor on the PLS-4, and many children had a score of 50, language age-equivalents were used for this study. Each age-equivalent score was normalized for chronological age by dividing the PLS-4 receptive language age and expressive language age with the child's chronologic age at time of testing and multiplying by 100. Receptive and expressive language quotients (LQs) close to 100 indicate that a child’s language level is age-appropriate. For the purpose of this study, we reported receptive and expressive language quotients and did not report a total language quotient.

Language testing was completed by the same speech-language pathologist who specializes in children with complex developmental issues. If children used any sign language for communication, a certified sign language interpreter was present for the evaluation. Questions were asked auditorially first and then subsequently the same question was asked via the sign language interpreter. As American Sign Language can be iconic and has a different structure from English, Conceptually Accurate Signed English was used first, followed by American Sign Language if needed. Appropriate accommodations were made for children with hearing impairment, per PLS-4 instructions (Zimmerman et al., 2002). Language scores obtained with the sign language interpreter were used for the purpose of analysis if they existed.

2.4. Other data collected

Upon enrollment, parents were asked to fill out a questionnaire that described the following: gender, race, their child’s communication strategy (speech, sign, behavior, other or a combination), educational placement (full or partial mainstream, preschool disabilities, self-contained classroom, home education, oral school, school for the deaf), what percentage of a school day was spent in the educational placement, if interpreter services are used at school, types of therapies (speech, occupational, physical, behavioral, vision, aural rehabilitation), where and how often each week or month therapy was received. Questions about the educational level of the parent, number of siblings at home, household income, and main source of health insurance were also asked.

2.5. Statistical analysis

Medians and ranges were reported for continuous variables (i.e., age of identification, age at implantation). Frequencies with percentages were reported for categorical variables. Differences between CI participants and their matched controls regarding categorical variables were tested using McNemar’s Chi-square. Differences in continuous variables between groups were tested using the Wilcoxon Sign Rank test. Correlations between language quotients and nonverbal cognitive quotients were conducted using the Spearman correlation coefficient. To understand the discrepancy between language level and cognitive abilities, the difference between language quotients (receptive and expressive) and nonverbal cognitive quotients was tested using Wilcoxon Sign Rank test. General linear mixed models were constructed to analyze independent factors related to receptive and expressive language skills separately while accounting for the matched pair design and potential confounders. Possible confounders included those factors which may influence language abilities, such as gender, maternal education, hours spent in

therapy. Potential confounders were included in the models if they remained significant at the $p < 0.20$ level. Results from the regression analyses were reported as beta coefficients ($\beta$) with 95% confidence intervals as estimates of the adjusted differences in language between CI recipients and their hearing matched controls. Adjusted mean receptive and expressive language quotients with 95% confidence intervals for both groups were also reported.

3. Results

3.1. Study participants

Twenty-two participants with cochlear implants were enrolled, though two were excluded: one was a sibling twin and one had incomplete demographic information. Fifteen control participants were matched to CI participants on both chronicologic age and cognitive abilities (Table 1). None of the CI participants had bilateral cochlear implants at the time of this study. Age- and cognitively matched controls were not enrolled for five CI participants (Table 2). One participant had a nonverbal cognitive quotient of 115 (equivalent to one standard deviation above the norm) and it was decided to not locate a match. The other four participants had either matched controls that did not show up for the study or an appropriate match was not located. Although the study was powered for a sample size of 20 controls, our preliminary analyses indicated that we did not require additional controls, due to the larger than expected difference between the two groups. Thus, the study was closed to further recruitment.

The median (range) age of identification of hearing loss for the CI participants was 2.7 months (0–25) and the median age at CI was 21 months (13.5–54). The duration of implant use ranged from 10 to 68 months. All but one CI participant had their implant for more than a year; one participant had his implant for only 10 months at the time of the study. Four participants had CHARGE syndrome and four had congenital cytomegalovirus infection as the etiology of deafness. The remaining etiologies of deafness included prematurity $(n = 2)$, meningitis $(n = 2)$, genetic $(n = 1)$, Infantile Refsums $(n = 1)$, and one auditory neuropathy without known risk factors.

The cochlear nerve was present in all 15 CI participants, as the absence of nerve would be a contraindication for a cochlear implant. A normal cochlea was observed in 10 participants. Four participants had bilateral cochlear hypoplasia and one participant had bilateral cochlear dysplasia. All but one participant (Child #8 in Table 1) had a full electrode insertion. All four children with CHARGE syndrome had a complete insertion with all electrodes activated except for one child (Child #11) who had 18 of 22 activated electrodes. Post-implant thresholds for the four-frequencies (0.5, 1, 2, 4 kHz) were averaged for each of the 15 children. These average thresholds ranged from 20 dB to 78.3 dB (median 34.4, mean 34.8). Up-to-date post-CI thresholds for the four-frequencies (0.5, 1, 2, 4 kHz) were averaged for all but one participant (Child #8 in Table 1) had a full electrode insertion. All but one participant (Child #8 in Table 1) had a full electrode insertion. All but one participant (Child #8 in Table 1) had a full electrode insertion. All four children with CHARGE syndrome had a complete insertion with all electrodes activated except for one child (Child #11) who had 18 of 22 activated electrodes. Post-implant thresholds for the four-frequencies (0.5, 1, 2, 4 kHz) were averaged for each of the 15 children. These average thresholds ranged from 20 dB to 78.3 dB (median 34.4, mean 34.8). Up-to-date post-CI thresholds were not available for Child #8.

All developmental disabilities were diagnosed clinically prior to study enrollment. The majority of the study participants in both groups had some form of cognitive delay and many had co-existing motor delays (Fig. 1). The diagnosis of cerebral palsy was made in five of the CI participants and in two controls. Four children with CHARGE syndrome in the CI group had developmental issues related to CHARGE, including cognitive, oral-motor, gross motor and vision impairment, though all had functional vision. One CI participant with autism spectrum disorder was matched to a control with an autism spectrum disorder.

Table 1
Cochlear implant study participants and their age- and cognitively matched controls.

<table>
<thead>
<tr>
<th>Cochlear implant participants</th>
<th>Control participants</th>
</tr>
</thead>
<tbody>
<tr>
<td>Match-pair Gender Age Developmental disability NVCQ</td>
<td>Match-pair Gender Age Developmental disability NVCQ</td>
</tr>
<tr>
<td>1 M 28.2 Cognitive + Motor + Oral-Motor 50</td>
<td>6 F 54.6 Cognitive + Motor + Vision 90</td>
</tr>
<tr>
<td>2 F 35.1 Cognitive + Oral-Motor + Vision 83</td>
<td>7 M 49.6 Cognitive + Motor + Vision 87</td>
</tr>
<tr>
<td>3 M 38.5 CP + Oral-Motor* 50</td>
<td>4 M 44.5 Mild Cognitive 50</td>
</tr>
<tr>
<td>4 M 44.5 Mild Cognitive 50</td>
<td>5 M 46.8 Motor + Apraxia 70</td>
</tr>
<tr>
<td>5 M 46.8 Motor + Apraxia 92</td>
<td>6 M 46.6 Mild Spastic Diplegia CP + Mild Cognitive 90</td>
</tr>
<tr>
<td>6 M 46.6 Mild Spastic Diplegia CP + Mild Cognitive 90</td>
<td>7 F 52.4 CP + Cognitive* 80</td>
</tr>
<tr>
<td>7 F 52.4 CP + Cognitive* 50</td>
<td>8 F 55.9 Cognitive 70</td>
</tr>
<tr>
<td>8 F 55.9 Cognitive 70</td>
<td>9 F 57.9 CP + Cognitive* 70</td>
</tr>
<tr>
<td>9 F 57.9 CP + Cognitive* 50</td>
<td>10 F 69.1 Cognitive + Oral-Motor + Vision 75</td>
</tr>
<tr>
<td>10 F 69.1 Cognitive + Oral-Motor + Vision 75</td>
<td>11 M 71.5 Cognitive + Motor + Vision 50</td>
</tr>
<tr>
<td>11 M 71.5 Cognitive + Motor + Vision 50</td>
<td>12 F 71.5 Cognitive + Motor + Vision 80</td>
</tr>
<tr>
<td>12 F 71.5 Cognitive + Motor + Vision 50</td>
<td>13 M 71.5 Cognitive + Motor + Vision 80</td>
</tr>
<tr>
<td>13 M 71.5 Cognitive + Motor + Vision 80</td>
<td>14 F 71.5 Cognitive + Motor + Vision 50</td>
</tr>
<tr>
<td>14 F 71.5 Cognitive + Motor + Vision 50</td>
<td>15 M 81.6 CP + Cognitive + Vision 70</td>
</tr>
<tr>
<td>15 M 81.6 CP + Cognitive + Vision 70</td>
<td>16 F 81.6 CP + Cognitive + Vision 70</td>
</tr>
</tbody>
</table>
| Abbreviations: NVCQ = nonverbal cognitive abilities, CP = cerebral palsy. Cochlear implant participant numbers 1, 2, 11, 13 had CHARGE syndrome. Cochlear implant participant numbers 3, 7, 8, 9 had congenital cytomegalovirus infection. *Deviation from matching protocol criteria for nonverbal cognitive quotient \pm 5 points.

No significant differences were noted between the CI participants and their matched controls regarding the basic demographic characteristics collected at the time of the study (Table 3). Children with cochlear implants attended a significantly higher number of different therapies compared to their matched controls, though no differences between groups were seen regarding the total hours per week spent in therapy.

### 3.2. Language outcomes

Among the 15 CI participants, the median (range) receptive and expressive language quotients were 30.2 (6–116) and 23.8 (11–107.5) respectively. The median (range) receptive and expressive language quotients for the hearing controls were 70.5 (14–127) and 64.0 (11–115) respectively. Duration with the implant (or implant experience) was negatively correlated with both receptive (rho = −0.47, p = 0.08) and expressive (rho = −0.38, p = 0.16) language, though these findings were not quite statistically significant. The age at which the implant was received was not significantly correlated with either receptive or expressive language (p > 0.6 for both LQs). Average four-frequency thresholds were negatively correlated with

### Table 2

Description of cochlear implant participants without matches (not included in analysis).

<table>
<thead>
<tr>
<th>Child</th>
<th>Gender</th>
<th>Age</th>
<th>Developmental disability</th>
<th>NVCQ</th>
<th>Hearing loss etiology</th>
<th>Reason no match found</th>
</tr>
</thead>
<tbody>
<tr>
<td>16</td>
<td>M</td>
<td>80.7</td>
<td>Severe Global CP + Cognitive</td>
<td>33</td>
<td>EVA</td>
<td>Matched control no-show</td>
</tr>
<tr>
<td>17</td>
<td>M</td>
<td>52.2</td>
<td>Motor Delay</td>
<td>83</td>
<td>Prematurity</td>
<td>Unable to find eligible match</td>
</tr>
<tr>
<td>18</td>
<td>F</td>
<td>81.0</td>
<td>Cognitive</td>
<td>64</td>
<td>CMV</td>
<td>Unable to find eligible match</td>
</tr>
<tr>
<td>19</td>
<td>F</td>
<td>69.5</td>
<td>Ataxic CP</td>
<td>115</td>
<td>Viral Encephalitis</td>
<td>Decided to not locate match</td>
</tr>
<tr>
<td>20</td>
<td>M</td>
<td>56.9</td>
<td>Cognitive</td>
<td>46</td>
<td>EVA/genetic</td>
<td>Matched control no-show</td>
</tr>
</tbody>
</table>

CP = cerebral palsy; CMV = cytomegalovirus infection; EVA = enlarged vestibular aqueduct.

![Fig. 1. Areas of developmental delay or disability among cochlear implant participants and age and cognitively matched hearing controls.](image)

No significant differences were noted between the CI participants and their matched controls regarding the basic demographic characteristics collected at the time of the study (Table 3). Children with cochlear implants attended a significantly higher number of different therapies compared to their matched controls, though no differences between groups were seen regarding the total hours per week spent in therapy.

### Table 3

Characteristics of 15 cochlear implant participants and matched hearing controls.

<table>
<thead>
<tr>
<th>Characteristic</th>
<th>CI participants</th>
<th>Controls</th>
<th>p-Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age of child*</td>
<td>52 (28–81)</td>
<td>66 (31–81)</td>
<td>–</td>
</tr>
<tr>
<td>Nonverbal cognitive quotient*</td>
<td>50 (27–92)</td>
<td>54 (22–89)</td>
<td>–</td>
</tr>
<tr>
<td>Number of developmental issues</td>
<td>2 (1–3)</td>
<td>2 (1–2)</td>
<td>0.13</td>
</tr>
<tr>
<td>Gender – male</td>
<td>7 (47%)</td>
<td>11 (73%)</td>
<td>0.16</td>
</tr>
<tr>
<td>Insurance type (private only)</td>
<td>6 (40%)</td>
<td>6 (40%)</td>
<td>1.0</td>
</tr>
<tr>
<td>Maternal education beyond high school</td>
<td>12 (80%)</td>
<td>11 (73%)</td>
<td>0.65</td>
</tr>
<tr>
<td>Income &lt; $40,000</td>
<td>5 (33%)</td>
<td>7 (47%)</td>
<td>0.48</td>
</tr>
<tr>
<td>Receiving speech/language therapy</td>
<td>13 (87%)</td>
<td>11 (73%)</td>
<td>0.41</td>
</tr>
<tr>
<td>Total hours per week spent in all therapies</td>
<td>3 (0–26)*</td>
<td>2.2 (0–7)</td>
<td>0.28</td>
</tr>
<tr>
<td>Total hours per week in speech/language therapy</td>
<td>1 (0–12.5)*</td>
<td>1 (0–2.5)</td>
<td>0.24</td>
</tr>
</tbody>
</table>

Data reported as medians with ranges in parentheses and frequencies with percentages in parentheses.

*p-Values for comparisons of continuous variables from Wilcoxon Sign Rank test and for categorical variables from McNemar’s test.

* Not applicable due to matching.

* One child in an oral deaf school program.

receptive (rho = −0.58, p = 0.03) and expressive (rho = −0.47, p = 0.09) language quotients. However, this association disappeared upon controlling for nonverbal cognitive abilities (partial correlation rho = −0.38, p = 0.21 and rho = −0.14, p = 0.64 respectively).

Five children had inner ear anomalies (cochlear hypoplasia and dysplasia) that could affect the results. However, the children with the inner ear anomalies had better median receptive and expressive language skills compared to the 10 children with no inner ear anomalies (data not shown). Thus, in this particular cohort of children with additional disabilities who had received implants, the inner ear findings were not associated with the poorer language skills within the group. Post-implant thresholds were not correlated with language outcomes in this population, after controlling for NVCQ (partial correlation coefficient Spearman rho = −0.37 receptive (p = 0.21) and rho = −0.14 expressive (p = 0.64)).

In order to determine differences in language skills between the CI participants and their matched controls, multiple regression analyses were conducted using general linear mixed models. These models controlled for potential confounders that may influence language outcomes, while simultaneously accounting for the matched design. In constructing the models, one pair (Child #5 in Table 1) was deemed to be a significant influential outlier. CI Child #5 had a nonverbal cognitive quotient (92) considered to be within one standard deviation of the population mean and attended an all day oral deaf education program where she received speech and language services every day throughout the day. Due to results of model fit statistics in combination with the high level of nonverbal cognitive abilities of CI Child #5 compared to the other CI recipients, this pair was not included in the multiple regression analyses.

Results of the models indicated that cochlear implant participants scored 24 points lower (β = 24.6, 95% CI 11.2, 38.1) on receptive language than the age and cognitively matched controls. Cochlear implant participants also scored approximately 22 points lower (β = 21.9, 95% CI 10.2, 33.7) than controls on expressive language testing (Fig. 2). Both models controlled for the number of different therapies and the weekly hours in speech-language therapy (p < 0.1). Additionally, gender was included in the model for receptive language, although it was not quite statistically significant (p = 0.08). No other variables met the model inclusion criteria described in the methods section.

3.3. Language-cognitive difference

Although language was highly correlated with nonverbal cognitive abilities for both the cochlear implant participants (receptive rho = 0.71; expressive rho = 0.77) and their matched controls (receptive rho = 0.84; expressive rho = 0.81), it appeared that children with implants were not reaching language levels that would be commensurate with their cognitive potential. Fig. 3a and b illustrates the nonverbal cognitive abilities for each group as a function of their language levels, reported as receptive and expressive language quotients. The CI group appeared to have a fairly large discrepancy between their language quotients and their cognitive quotients. The controls, however, were more likely to meet their cognitive potential. Children with CIs and additional disabilities had a significant difference in median quotient values between LQ and NVCQ for both receptive language (−25 (range −65 to 24)) and expressive language (−22 (range −56 to 15.5)). Differences between language and nonverbal cognitive quotients were not significant among the control group (Fig. 4a and b).

4. Discussion

Children with cochlear implants who had additional disabilities had significantly lower receptive and expressive language quotients (at least 20 quotient points lower) compared to their hearing peers of the same age and nonverbal
cognitive abilities. In addition to the lower language levels, children with additional disabilities appeared to have significant delays in their language levels that were disproportionate to their nonverbal cognitive abilities, or “cognitive potential.” The receptive and expressive language quotients were significantly lower than the nonverbal cognitive quotients among the cochlear implant group only. Children in the hearing control group did not have this same linguistic-cognitive discrepancy. Although the finding that children with cochlear implants and additional disabilities exhibit language delays is not new, quantifying the discrepancy between language and cognition relative to age and cognitively matched peers is novel.

Providing cochlear implants to deaf children with developmental disabilities can result in substantial benefit. Improvements in speech perception, sentence recognition or speech production (Dettman et al., 2004; Edwards et al., 2006; Nikolopoulos et al., 2008; Pyman et al., 2000; Trimble et al., 2008; Waltzman et al., 2000) and auditory skills (Daneshi & Hassanzadeh, 2007; Hamzavi et al., 2000; Wiley et al., 2008) have been reported in this population. Among children with mixed additional disabilities, improvements in speech and/or word recognition occur in anywhere from 10% to 70% of study populations (Berrettini et al., 2008; Hamzavi et al., 2000; Nikolopoulos et al., 2008; Vlahovic & Sindija, 2004; Waltzman et al., 2000; Winter, Johnson, & Vranesic, 2004). A few studies have reported on language skills among children with additional disabilities. Post-implant studies among children with additional disabilities with language as an outcome described either below average language skills or slower rates of language acquisition when compared to typically developing children with implants (Holt & Kirk, 2005; Pyman et al., 2000; Waltzman et al., 2000). Studies that use typically developing comparison groups are unable to address the fundamental questions regarding expected language levels among children with developmental disabilities. Because language levels should at least be on par with a child’s cognitive abilities, hearing control groups of typically developing children are inappropriate comparisons.

Children with developmental disabilities are a heterogeneous group making it very difficult to categorize them. Even among specific diagnoses (e.g. cerebral palsy), heterogeneity in disability severity and a wide range of abilities exist. Thus, outcome expectations established on the disability “label” would be neither accurate nor appropriate. Nonverbal cognitive abilities, which provide the clinician with a picture concerning a child’s potential for performance, could be used as a guide.
for expected outcome development. Although the current study suggested that children with cochlear implants were not meeting their cognitive potential regarding their receptive and expressive language levels, the nonverbal cognitive abilities remained highly correlated with language. In addition, results from the overall study of 20 children with implants reported that nonverbal cognitive abilities continued to be highly predictive of language post-implant (Meinzen-Derr, Wiley, Grether, & Choo, 2010). Using cognitive standardized quotients in the developmental evaluations would have strengthened our study. However, clinically this is not always feasible due to limited access to cognitive testing and funding for this service in our patient population. Our measures used and reported in the two groups were the same, thus imparting the same testing bias in both groups.

The language measure used for the study, the PLS-4, provides standard scores, though only age-equivalents were reported. Optimally, standard scores would have been used to provide us with information on nonverbal cognitive abilities. Most (10/15) of the children in the CI group had a standard score of 50, which was the floor on the PLS-4. Thus, the standard score would possibly have overestimated language levels and not given a clear picture of a child's true language abilities. A major strength of the current study is the prospective language testing of all children and the obtainment of a language score, regardless of disability severity. This is a testament to the high level of experience of the speech-language pathologist with children who are diagnosed with multiple disabilities. In addition, the PLS-4 is not simply a measure of spoken language. The auditory comprehension component is used to evaluate how much a child understands and the expression communication component evaluates how well a child communicates with others (Zimmerman et al., 2002). This tool was determined to be appropriate for our population because the administrator can modify a test task for a child with special needs. Accommodations can also be made regarding the use of an interpreter for children who use sign language. A certified interpreter was always used when children used sign language. Although it can be argued that a child receiving information both auditorily and then subsequently through sign language may have an advantage over children who only received the testing auditorily, we would like to emphasize that children with implants continued to do worse than their hearing cognitively matched controls, regardless of the use of sign language.

Fig. 4. Line plot of the difference in the language quotient (LQ) and the nonverbal cognitive quotient (NVCQ) for receptive and expressive language. Dots above the dotted line (at zero) indicate that the LQ was higher than the NVCQ. Dots below the dotted line indicate that the LQ was lower than the NVCQ. The CI participants (a) had significant lower LQs compared to their NVCQs while no significant difference between LQ and NVCQ was seen among the controls (b).
Although we attempted to strictly match on both age and nonverbal cognitive abilities, we made the decision a priori that nonverbal cognitive ability was the most important matching criterion. Five of the 15 pairs deviated from the age-match, with the largest age discrepancy seen in Child #10 (22 months apart). Because the controls had a higher median age at the time of the study, we investigated in the regression models whether these deviations allowed for confounding by age. Age was neither significant (p > 0.5) in the either of the models, nor did it contribute to the relationship between group and language outcomes. Additionally, the number of disabilities was not a factor in our current analysis, nor was it a factor in our previous study (Meinzen-Derr et al., 2010). We also recognize that some children did have limited “hearing” experience compared to the control group who had hearing since birth (all but one child with a CI had their implant for more than 1 year). Although it is possible that more time is needed for children to gain improvements post-implantation, results from our study of 20 children with additional disabilities showed otherwise (Meinzen-Derr et al., 2010). Children with longer implant duration had worse language post-implant compared to those with shorter implant duration. Cognitive abilities were the strongest predictor of language, regardless of the duration of implant experience.

A couple of factors came into play when choosing our control population. The use of a hearing control group is a common occurrence in cochlear implant studies of typically developing children. Some studies have, in fact, used the “hearing age” of children in their comparisons to control groups to account for the time of auditory stimulation. Unfortunately, the “hearing age” of a child who has received a cochlear implant does not accurately quantify the actual language gap a child may have relative to his or her age. We also encourage the use of other communication modalities (not only auditory-specific) when a child is identified with their hearing loss. This provides some amount of language or communication foundation for the child prior to implant experience. The other reasoning for our design was around the inappropriate use of typically developing control groups when studying children with developmental disabilities. Our research team felt that the most appropriate measuring stick for a child with a disability and a CI (or any child for that matter) is a control with similar developmental or cognitive abilities. It is a disservice to simply state that children with additional disabilities have poorer language performance than their typically developing hearing or cochlear implant peers. We sought to quantify the true difference between children with additional disabilities and their hearing cognitively matched peers.

Due to the matched design of the current study, it was not possible to include factors such as duration with implant, age of implant, or age of hearing loss identification in the regression models (only the CI group had values). Our hypothesis was that the groups were different regarding language abilities; implant and hearing loss factors are part of the group differences. In the overall cohort of 20 children with cochlear implants, age at implantation did not contribute to the variability in language outcomes (Meinzen-Derr et al., 2010). Longer duration with the implant was associated with poorer language skills, which was surmised to be more related to the complexity of the child versus the implant experience per se. We also investigated the possibility that poorer language performance could be due, in part, to poor post-CI thresholds. Language levels were not correlated with post-CI thresholds upon controlling for NVCQ. It was determined that any potential association between threshold and language was confounded by nonverbal cognitive abilities, which was deemed the most important predictor of language outcomes among this population of children (Meinzen-Derr et al., 2010).

Measures of early development may not always coincide with later outcomes (Aylward, 2004; Bornstein et al., 2006). In the current study, however, early measures of cognitive abilities among children with normal hearing appeared to coincide fairly well with later language abilities. The control group had fewer types of developmental delays per child and required fewer types of interventions compared to the CI group. The overall developmental needs were less from very early on in this group. Therefore, it is possible that the early interventions received by the control group were better targeted and more effective. The cohort of children with cochlear implants was fairly complex, and some children had more developmental issues than the controls which could alter the language trajectory. Although the current study did not find a relationship between language and the number of issues, we did control for the number of different therapies received as a marker of severity of developmental issues. Developmental or cognitive trajectories over time would be helpful in understanding a child’s long-term potential.

It may seem that our results are disconcerting. However, as these language outcomes have never been quantified in this population, this work helps us consider appropriate next steps to diminish this gap in communication skills for this group of children. It is also important to recognize that language is not always the only anticipated outcome for cochlear implantation in this group of children. Our center tends to take a fairly generous approach to implanting children that other centers may not consider appropriate for implantation. However, we believe our philosophy in improving quality of life is an appropriate minimum outcome.

Our study has helped us consider the clinical implications that children with cochlear implants and co-existing disabilities potentially need more frequent monitoring of language and communication progress. Adaptive and/or augmentative communication strategies should be considered when either oral or sign language progress is not occurring. As a direct result of the current study, two children with implants had a change in their therapy management. One child with CHARGE syndrome was receiving speech therapy, but focused solely on feeding issues. Communication strategies using pictures were implemented for this child once the results of the study language evaluation were known. Another study participant with severe cerebral palsy has since completed an augmentative communication evaluation for a speech generating device. It is important to emphasize that the current study used a speech-language pathologist with expertise in children with complex needs. It is feasible to consider that children with hearing loss who have therapy needs above and beyond that for hearing loss alone would require therapists who have a high level of knowledge of severe developmental disabilities as well as good working knowledge of child development. It is also important to consider other outcomes beyond
speech and language for this group of children. Quality of life benefits may be equally important to families of this group of children.

Although children with cochlear implants and additional developmental disabilities make some language progress post-implant, it is clear that they significantly under-perform compared to developmentally matched hearing peers. Understanding the reasons for profoundly lower than expected language levels among children with co-existing hearing loss and developmental disabilities is the first step in determining effective interventions to improve outcomes.

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