I. Introduction

A. Nature of the research problem

Over 40% of children with clinically significant permanent sensorineural hearing loss (HL) have another neurodevelopmental disability,\(^1-6\) most frequently cognitive disability.\(^5,6\) The co-existence of HL and low nonverbal cognitive functioning interacts such that the combined result is worse than either condition in isolation.\(^7-9\) Because language skills are highly correlated with academic, occupational, and social success, this dual diagnosis places children at immense risk for life-long disability. Indeed, the lifetime cost for hearing children with cognitive disabilities is 25 times higher, amounting to $50 billion more, than for children with HL alone.\(^10\) Research linking the importance of auditory and cognitive processes in optimizing outcomes in HL is recognized in adults\(^11-16\) and pediatric cochlear implant populations with normal cognitive abilities.\(^17-23\) Similar evidence in children who are deaf/hard of hearing (DHH) who have cognitive disabilities has been severely lacking, stifling progress towards effective interventions. Identifying mechanisms of language deficits among children who are DHH and cognitive disabilities is the basis for novel assessments, and this study creates the foundation for designing effective evidence-based interventions. We expect our studies to improve language outcome prediction and provide critical information about language-based intervention strategies for children with co-existing HL and cognitive disabilities. Providing all children with opportunities to enhance early language and communication will build their capacity to transition into adulthood, decrease life-time functional disability, and thus maximize their potential.

B. Purpose, scope, and methods of the investigation

The overarching goal of this work is to characterize how the co-occurrence of sensorineural HL and cognitive disabilities impacts functional language and communication among children who are DHH with cognitive disabilities. We enrolled a unique cohort of children (DHH with cognitive disabilities, DHH only, cognitive disabilities only), which also allowed us to address potential neurocognitive and audiologic predictors for language in children who are DHH across all levels of ability. The specific aims of this study were to: (1) Determine the extent to which nonverbal cognitive IQ (NVIQ) explains the divergence between language level and cognitive level in children with sensorineural hearing loss; (2) Determine specific audiologic features that impact the divergence between language and cognitive level among children with cognitive disabilities; (3) Characterize the language-based interventions for children with hearing loss and cognitive disabilities.
II. Review of the Literature
Despite our early identification and intervention efforts and emphasis on language goals, relatively large pockets of children who are DHH struggle with significantly lower language levels disproportionate to their scores on nonverbal cognitive assessments\textsuperscript{17,24-29}. These language “deficits,” even when only in the low average or average range, have negative cascading effects on academic success\textsuperscript{30-32}, independent functioning,\textsuperscript{33-35} and employment opportunities in young adulthood.\textsuperscript{36} Relatively large numbers of children who are DHH struggle with significantly lower language levels disproportionate to their scores on nonverbal cognitive assessments\textsuperscript{25,26,28}. These lags in early language often worsen rather than improve through the school years, irrespective of hearing loss severity.\textsuperscript{17,29,37} Because early language difficulties are considered precursors of academic, social, behavioral, and psychiatric problems,\textsuperscript{31,38,39} any additional negative impact places a child with hearing loss at immense risk for persistent underachievement that interferes with optimal health, well-being, and community integration. Cognitive disabilities are the most common comorbidity among children who are DHH.\textsuperscript{5,6} This specific group of children are at highest risk for life-long inadequate communication skills, placing them in danger of secondary disability. Though delayed compared to typically developing children, language of children who are DHH and cognitive disabilities should not be different relative to cognitive-matched peers with normal hearing.\textsuperscript{40,41} Yet, the impact of the dual diagnosis of HL and cognitive disabilities leads to language levels far below what is cognitively commensurate. Research has investigated this important hearing and cognitive interplay in the aging population\textsuperscript{11-16} and cochlear implant pediatric populations.\textsuperscript{17-23} Studies, including our own, have shown that measures of nonverbal cognitive ability are highly predictive of language in deaf children with disabilities.\textsuperscript{25,42} Clinically, HL is often tested and treated in isolation from cognitive abilities. Thus, the physical sound source is separated from the meaning of sound. This is a problem because in challenging listening environments, people will use learned rules and previous knowledge to “fill in the blanks”.\textsuperscript{11,13,14,43} When speech signals are degraded, cognitive skills are used to better resolve acoustic input. In other words, cognitive performance is maximized when listening is effortless. As listening effort increases, fewer cognitive resources are available for listening and reasoning, resulting in reduced processing and comprehension.\textsuperscript{44} The extent to which various levels of nonverbal cognition affect language among the larger pediatric population with HL (specifically those with joint hearing and cognitive deficits) is not fully explained.

III. Study Design and Methods
A. Study design
This was a prospective cross-sectional study of children with permanent bilateral hearing loss. Subjects were recruited from the Division of Developmental and Behavioral Pediatrics, the Ear and Hearing Center, and through advertisement flyers. Participants were enrolled into one of three study groups: 1) HL with normal NVIQ, 2) HL with low NVIQ (dual diagnosis), and 3) normal hearing with low NVIQ. These three groups served to help us understand the relationship of the combination of hearing deficits and cognitive deficits with achieved language. All subjects receive standardized neurocognitive, language, and functional assessments which occurred during a one-time study visit.

B. Population studied
We enrolled a total of 123 participants between the ages of 3-6 years; 93 children had bilateral hearing loss. The hearing controls were
frequency matched to children with lower cognitive abilities, regarding nonverbal IQ and age. Figure 1 highlights the enrollment strategy. Children with other known disabilities or syndromes that potentially impact communication (e.g., significant motor disabilities, autism, Williams syndrome), children with vision impairment, neurocognitive testing standard scores <40 or unable to complete testing were excluded. The age range of 3-6 years was chosen because: a) many disabilities are diagnosed by three; b) HL management is established for most children; c) represents an important transition period after early intervention prior to academics.

1. Children with bilateral permanent hearing loss and normal NVIQ. To help delineate the joint impact of HL and cognitive deficits on language levels, this group of children with HL only was critical and allowed us to analyze a continuum of NVIQ. We enrolled subjects who had mild to profound sensorineural HL and had NVIQ ≥80. Children with suspect or concern for developmental delay were excluded from this group. Frequency matching to dual diagnosis group occurred according to severity of HL.

2. Children with bilateral permanent hearing loss and low NVIQ. We enrolled subjects who had a cognitive disability (study definition using NVIQ). Our strict inclusion/exclusion criteria helped define a homogenous group of children.

3. Children with normal hearing and low NVIQ. Children with normal hearing and cognitive disabilities served as a reference regarding language levels commensurate with cognitive abilities. This group was necessary to delineate effects of HL on outcomes above and beyond the effects of cognitive deficits. Children with no documentation of normal hearing test were excluded. Frequency matching to the dual diagnosis group occurred regarding nonverbal cognitive abilities and age. We enrolled a total of 123 participants into the study. The mean age of our overall study population was 57.7 months (SD 13.7) and 62% (n=76). Among children who were DHH (n=93), the degree of hearing loss ranged from mild to profound, with 42 (45%) using a cochlear implant for deafness.

C. Sample selection
Participants were recruited from the Division of Developmental and Behavioral Pediatrics (DDBP), the Ear and Hearing Center (EHC), and through advertisement fliers. All children who are DHH are referred for a clinical developmental assessment and followed clinically by Dr. Wiley (co-PI) through the EHC either upon the initial EHC visit or upon suspicion of delay. For study inclusion, all diagnoses of HL and cognitive disability had been documented prior to enrollment. Both children with HL and children with hearing were recruited and enrolled via the same or similar methods.

D. Instruments used
The standardized assessment battery was carefully chosen by the research team and was based on what is most appropriate for the study population. Test administration occurred by trained professionals (i.e., speech-language pathologists, neuropsychologists) at the study visit. A brief summary of assessments are as follows:

- **Language Measures (outcome):** Preschool Language Scale-4th edition, Peabody Picture Vocabulary Test-IV.
A short interview with parents occurred to collect information on maternal education, socioeconomic status, insurance status, number of siblings in household, and intervention-specific information. Intervention-specific information was also abstracted from data sources described under Aim 3.

D. Statistical techniques employed

All statistical analyses were performed using SAS v9.3 software. Descriptive statistics such as means, medians, ranges, standard deviations, frequencies and proportions as appropriate were computed and continuous variables were examined for normality. Bivariate analysis occurred between each outcome variable (absolute language scores and the ratio of language level to NVIQ). The ratio was calculated to define the divergence between language and cognitive level (language score divided by cognitive score multiplied by 100).

The relationship between continuous values of NVIQ and other factors was investigated using Pearson correlation with continuous variables and analysis of variance (ANOVA) or t-test for categorical variables. We constructed multivariable general linear models to assess the amount of variability (using the coefficient of determination or \(R^2\)) in outcomes explained by NVIQ. Because predictability of NVIQ may differ by study group, we stratified our analysis and evaluated possible differences across study groups regarding \(R^2\) and slope (\(\beta\) coefficient). We also explored the relationship between communication and social functioning (subscales of the Vineland and the PEDI) as outcomes with NVIQ and language to determine if similarities exist with the NVIQ relationship. Model development was then extended to examine the link between audiologic characteristics and language. Potential confounders and covariates evaluated in all analyses (and models) included the following (though not an inclusive list): Child information such as age of identification and amplification, communication mode, device use (hearing aid, FM system), and types of intervention programs and the background of the providers (i.e., deaf education, speech language pathologist). An SES score that was created using maternal education, family income, and The Hollingshead Four-Factor Index of Social Status will be used to quantify SES. Other information will include date of birth, gender, and race. We will add adaptive functioning scores to the models. Detailed audiometric information will be tested with Aim 2.

IV. Detailed Findings

We have found a series of interesting and important findings regarding our study, reported below by study aim. The mean (SD) nonverbal IQ for 93 children who were DHH was 94.8 (19). The mean (SD) receptive language standard score was 81.3 (18.6) with 47% of children scoring <80 on the standardized assessment. Children’s language performance was significantly lower (average 15 points, \(p<.0001\)) than their nonverbal IQ, indicating a significant divergence in language to cognitive abilities. Detailed summary of findings by aim are listed below. Among the three groups of children, children who were DHH had significantly higher mean receptive language scores than children who were DHH + low cognition or hearing +low cognition (89.1 (16), 66.3 (13.1), and 66.8 (13.1) respectively, \(p<.0001\)). However, the ratio of language to cognitive abilities was not significantly difference across all three groups. (84.9 (12.7) vs. 84.5 (27), vs. 91.6 (15.7), \(p=0.39\)). The language gaps identified in children who were DHH occurred across all levels of cognitive ability (a ratio of 100).

Aim 1. Cognition and Language: Among children who were DHH, nonverbal IQ measures were significantly correlated with language (Pearson rho=0.67, \(p<.0001\) for nonverbal IQ composite score and receptive language), supporting Aim 1. Importantly, specific components or subscales of nonverbal IQ appear more predictive of language than others. These include the Form Scale...
on the Leiter-R (ρ=0.60, p<.0001) vs. the Figure-ground scale (ρ=0.44). Executive function subscales (from the BRIEF) were also found to be correlated with language, such as working memory (ρ=−0.29, p=0.005). Among children with hearing (controls), working memory was not associated with language (ρ=−0.02, p=0.93). Results of the general linear models indicated that NVIQ explained 18% of the variance in receptive language among children with NVIQ≥85 (average to above average IQ). However, two subscales of the Leiter (Form completion and Repeated patterns) explained 30% of the variance in receptive language. Amongst participants with NVIQ<85, NVIQ explained only 11.5% of the variance, though Form completion alone significantly accounted for ~25% of the variance.

**Aim 2.** Hearing loss severity and language: Among participants who were DHH, the degree of hearing loss (measured with PTA) was significantly (p≤.0001) correlated with both receptive (rho=−0.41) and expressive (rho=−0.38) standard scores. The aided thresholds (using either the aided values from SAT or PTA) were also correlated with receptive (rho=−0.39, p=0.0003) and expressive language scores (rho=−0.35, p=0.001). Hearing loss severity was also significantly correlated with the discrepancy between language and cognition (language-cognitive ratio) (rho=−0.36, p=0.0006) as well as with the aided thresholds (rho=−0.29, p=0.01). As the audibility worsens the gap between language and cognitive abilities widens. These findings indicate that degree of hearing loss appears to play a role regarding language acquisition, but that delays in language learning are mitigated by better aidability (the better access to sound, the better language is acquired). Aidability variables (level of aided hearing and/or having a cochlear implant) were included in the original neurocognitive models (language as the outcome). Better aided speech awareness thresholds were significantly associated better language in all children with hearing loss, though children who had received cochlear implants had significantly lower language than those who had hearing aids (p<.0001). The neurocognitive subscales and executive functioning, having a cochlear implant, SES (a composite score of income, maternal education, and insurance), hours of speech therapy per week, and age of child accounted for 67% of the variance in receptive language. In addition, after adjusting for the factors in the regression model, there was no difference in the average standard scores between NVIQ groups (least square or adjusted means 83.6 for NVIQ≥85 and 81.2 for NVIQ<85, p=0.58).

**Children with cognitive disabilities.** Children who were DHH with cognitive disabilities had similar language levels than their age and cognitively-matched peers. No differences between groups in the standard scores were seen (65.8 (13.1) vs. 67.8 (12.8) respectively for receptive language, p=0.6.) There were similar findings seen with expressive scores. Although the ratio of language scores to nonverbal IQ appeared slightly lower (84.5 (27) vs. 91.6 (15.7), p=0.26) this was not a statistically significant finding.

**Aim 3.** Characterization of language-based interventions: We collected detailed information on the different interventions (including therapies) that participants received. Each child’s detailed record was pulled and organized by our research assistant (Smith) and a speech and language pathologist (Hibner) codes each one according to a standardized rubric that has been discussed at length with significant input by Dr. Grether (co-I). Eighty-five percent of participants who were DHH were enrolled in some form of speech therapy; 26% were enrolled in aural rehabilitation therapy for hearing. Among participants who were receiving speech therapy, the median hours per week spent in speech therapy was 1.25 hours (range 0.25-12.5). Parents who reported ≥4 hours of speech therapy each week had children enrolled in an oral deaf school program, which incorporates speech and language therapy as part of the regular curriculum. Only 20% of children had goals specifically targeting social functioning, while 75% had a speech-related goal. The number of syntactic goals was significantly correlated with receptive and expressive
language (Spearman rho=0.70 & 0.65 respectively), the gap in language (rho=0.47), as well as functional communication (rho=0.47) and social functioning (rho=0.55).

**Additional important findings:** Because functional skill assessment is a high priority in early intervention and developmental research,1,2 we assessed functional skills in our participants using the VABS and the PEDI. These functional assessments focus on adaptive skills and identify the kinds of support the child needs in daily activities. Using this framework, we examined children’s abilities and challenges in functional skills in our cohort of children. Our data (Meinzen-Derr et al 2014) showed that on average children who are DHH continued to have standard scores on the functional assessments that were significantly lower than the population mean across the different domains. Specifically interesting was the relationship between the deficits in communication and social functioning with the gap in language (the language-cognitive gap). *This study was the first to document lower functional skills among this population of children.* Our data suggest that children who are DHH continue to have mean communication, social, and self-care function standard scores that are significantly lower than the population mean. In fact, >60% of this population have social functioning scores that are at least 1 SD below the mean; 30% of these children have a NVIQ>100. Social functioning scores were not significantly correlated with degree of HL, maternal education, or SES (p>0.3 for all).

**V. Discussion and Interpretation of Findings**

**A. Conclusions to be drawn from findings (with reference to data supporting each).**

Data supporting the following conclusions were detailed in the above section. As detailed in the findings above in Section IV, we found that language levels of our sample of children who were DHH with lower cognitive abilities were not significantly lower than their age-cognitively matched peers. This finding perhaps speaks to our work around improving the outcomes for children with the dual diagnosis of HL and other developmental disabilities. However, we found deficits in language among children who were DHH with average to above average cognitive abilities (NVIQ>85). In fact, the language gaps (as defined by the ratio of language to cognitive scores) were similar in both DHH groups. Over 35% of children who were DHH had a ratio<80, indicating that their language levels were 20% lower than their cognitive levels.28 Children with significant language gaps had social and communication functioning that looked like children with cognitive disabilities who had appropriate language (language commensurate with cognitive abilities). The biggest impact of this gap was seen in children with higher NVIQ (>100) across all degrees of hearing loss.28 The strongest predictors of language were the results from neurocognitive assessments, with some of the subscales acting as stronger predictors than the overall NVIQ score (i.e. Form subscale on the Leiter-R). This finding has allowed us to consider potential intervention strategies that target specific identified neurocognitive factors that are important to social and communication functioning. Because hearing children do not need to rely as heavily on nonverbal cognition to compensate for language deficits, the identification of both the verbal and nonverbal correlates to functional outcomes becomes very important in children who are DHH. Aidability was the best audiologic predictor of outcome, though this was not particularly a robust finding.

During our classification of speech-language therapy goals, we found that social skills and pragmatic concerns were not included within the priorities of out-patient clinical settings, despite considerable under-performance on functional communication measures in our study. Pragmatic goals need to be identified and established sooner and more often. This emphasizes a substantial gap in recognizing and intervening for the social function needs of children who are DHH. Therapy services have been slow, if not reluctant, to trial new language interventions and service
delivery models. This finding has compelled us to design an intervention that is tailored not only to the cognitive level of the child, but to focus on pragmatics.

B. Explanation of study limitations
Although 40% of children who are DHH have an additional developmental disability, the prevalence of children with a dual diagnosis is relatively low. Thus, the identification of children who are DHH with cognitive disabilities was the largest limitation. We made a few adjustments to help overcome some challenges, such as expanding our inclusion criteria from moderate hearing loss to include mild hearing loss. What we have gained from this expansion is generalizability to children with bilateral hearing loss of all degrees, which has been important. We have made significant progress in achieving our study aims, as evidenced by our publications and presentations. The study was designed as cross-sectional, though a longitudinal assessment would provide us with more meaningful information regarding language and outcome trajectories. A major limitation for Aim 3 was in the number of participants for whom we had complete therapy records. Although many children had an evaluation, these evaluations were for one time assessments, and continued therapy either had not taken place at our institution, or there were few goals listed (we found some instances where a child had no goals). This makes findings from this Aim difficult to generalize. However, Aim 3 was designed to be more qualitative and exploratory and was very instrumental in providing supporting evidence for the importance of future intervention work.

C. Comparison with findings of other studies
Sections I and II report details from other published studies, which mirror the findings in our study, reporting that average language scores in for children who are DHH are in the average to low average range. However, these studies do not consider the cognitive abilities of the child in their determinations of appropriate language development. Just as children with cognitive disabilities should have language expectations that reflect their cognitive level, children with above average cognitive abilities should have the same degree of expectations on their language development. By using a ratio of language to cognitive ability, we are able to “normalize” language levels to the capability of the child (and not just to the age of the child, as standardized scores will do). We have found that >35% of children who were DHH had some form of a language gap or deficit. After controlling for potential confounders, we found no differences in the language and in the ratio between children with low NVIQ and higher NVIQ. This concerning finding speaks to the growing language deficit among children with higher NVIQ. We also reported the impact of this language deficit or gap on social and communication functioning. Our study was the first to report functional skill development in a) children with other disabilities (Wiley et al 2012) and b) among the broader group of children who are DHH. (Meinzen-Derr et al, 2014).

D. Possible application of findings to actual MCH health care delivery situations
Our results have implications for the Early Hearing Detection and Intervention (EHDI) system, which has strong support from the Maternal and Child Health Bureau (MCHB). Understanding and monitoring developmental profiles in young children who are DHH who have been identified through newborn hearing screening programs have the potential to improve intervention services designed to prevent the development of a language gap. It is important for Early Intervention Systems that serve children who are DHH to regularly evaluate cognitive and language functioning and adapt services when language progress is not commensurate with cognitive functioning. MCHB is poised to encourage state EHDI programs to collaborate with
intervention services to monitor and maximize outcomes for children identified through Universal Hearing Screening (UNHS)/EHDI Programs. As members of our research team (specifically Dr. Wiley) have served on the EHDI Task Force, and have played instrumental roles in the EHDI system, both locally and nationally, we have used results from this study to encourage EHDI programs to better consider cognitive abilities for children who are DHH.

E. Policy implications
This study has implications for two different policy sectors: insurance and educational policies. The implementation of the Affordable Care Act (ACA) has the potential to increase the number of children who are DHH who will receive speech and language services. This is especially true with the elimination of pre-existing condition exclusions and the inclusion of habilitative services and devices as essential health benefits (Section 1302, Patient Protection and Affordable Care Act, Pub. L. 111-148). However, each state has some autonomy in deciding what health benefits are deemed “essential” for all plans in the state. This means that insurance companies may limit access to rehabilitative and habilitative speech-language pathology services. In addition, different states cover habilitation to different degrees. For example, though ACA plans will cover 30 visits for speech-language therapy each year, the eligibility for coverage is variable by plan and state. Standardized tests are often used as a measure for eligibility determination, with requirements that standard scores must be at least 1.75-2 standard deviations below the mean for a child to be eligible for coverage. Based on these varying aspects of care delivery, our study results are providing evidence to help support a broader definition of appropriate service delivery for children who are DHH, aligning well with the current Medicaid program definition of habilitation services.

Results from our study also have educational implications. The current educational system through Individuals with Disabilities Education Act (IDEA) supports children who are DHH through a primary disability level. School systems tend to identify the primary disability level as hearing loss. This poses challenges when children have a hearing loss as well as a co-existing developmental disability. Educational teams are left to identify a primary educational label which may guide discussions around interventions to focus on one aspect of a child rather than the child’s full needs. Furthermore, the definition of a specific learning disability by IDEA indicates that the learning problem cannot be due to a hearing or vision impairment. It is understandable that children who are DHH need interventions by providers with knowledge in deaf education. However, by limiting the educational label, co-existing needs may be left to be managed by deaf educators with minimal education and expertise in the expanded learning strategies appropriate for the co-existing conditions. Our data would also support the exploration of the possibility of a child who is DHH having a language based learning disability or specific learning disability.

F. Suggestions for further research
(1) Cross-sectional studies are ill-equipped to understand the impact of language deficits on the development of functional outcomes. Longitudinal studies are necessary to allow us to assess the development of functional outcomes (specifically communication and social functioning) and the relationship with language acquisition to this development. Expanding the current work to focus on children entering primary education is important to understand the effects of these language and social deficits on education and further social functioning. (2) Novel language interventions are necessary for this population of children. Thus, future research should also include the development of novel intervention strategies that would focus on combined neurocognitive and language processes. (3) We should consider defining specific language
impairment (SLI) in children who are DHH. Our study findings indicate that children who are DHH have the potential for SLI and research is necessary to provide evidence to support this diagnosis in this population. Evidence-based research is necessary to begin to shift the clinical paradigm that defines language impairment for children who are DHH.

VI. List of products (peer reviewed articles, books, chapters in books, master and doctoral dissertations, conference presentations, etc.).

Peer reviewed publication (to date)


Manuscripts in progress

- Wiley, Meinzen-Derr, Choo, Grether. Does hearing loss modify the role of nonverbal cognition in language among young children?
- Wiley, Meinzen-Derr. Early developmental assessments and later nonverbal intelligence quotients among children who are deaf or hard of hearing.

Conference presentations/proceedings


References


Maternal Child Health Measurement Research Network. Christina Bethell (2016-2020) Supported by HRSA’s Maternal and Child Health Bureau, the Measurement Research Network (MRN) is a multidisciplinary, collaborative network of experts who represent the MCH lifespan and who are active in the measurement of health and well-being of MCH populations. Additionally, the DRI supports Title V Maternal and Child Health Service Block Grant programs and partners in accessing and effectively utilizing data from each level. Learn more at the National Data Resource Center. Back to Top. Principal responsibilities included project management, data analysis, and report and manuscript preparation. (Completed). @inproceedings{Hepburn2012ComprehensiveFR, title={Comprehensive Final Report Maternal and Child Health Research Telehealth Delivery of a Family-Focused Intervention to Reduce Anxiety in Youth with Autism Spectrum Disorders in Rural Colorado HRSA Award: #R40MC15593-01-00 PI:}, author={Susan Hepburn}, year={2012} ).