Published in final edited form as:

J Am Acad Audiol. 2016 March ; 27(3): 204–218. doi:10.3766/jaaa.15050.

Children with ANSD fitted with hearing aids applying the AAA Pediatric Amplification Guideline: Current Practice and Outcomes

Elizabeth A. Walker, Ph.D.^a, Ryan W. McCreery, Ph.D.^b, Meredith Spratford, Au.D.^b, and Patricia A. Roush, Au.D.^c

^aDepartments of Communication Sciences and Disorders, University of Iowa

^bBoys Town National Research Hospital, Omaha, NE

^cDepartment of Otolaryngology, University of North Carolina, Chapel Hill, NC

Abstract

Background—Up to 15% of children with permanent hearing loss have auditory neuropathy spectrum disorder (ANSO), which involves normal outer hair cell function and disordered afferent neural activity in the auditory nerve or brainstem. Given the varying presentations of ANSD in children, there is a need for more evidence-based research on appropriate clinical interventions for this population.

Purpose—This study compared the speech production, speech perception, and language outcomes of children with auditory neuropathy spectrum disorder (ANSO) who are hard of hearing and children with similar degrees of mild to moderately-severe sensorineural hearing loss (SNHL), all of whom were fitted with bilateral hearing aids based on the American Academy of Audiology (AAA) pediatric amplification guidelines.

Research design—Speech perception and communication outcomes data were gathered in a prospective accelerated longitudinal design, with entry into the study between six months and seven years of age. Three sites were involved in participant recruitment: Boys Town National Research Hospital, the University of North Carolina at Chapel Hill, and the University of Iowa. Study sample: The sample consisted of 12 children with ANSD and 22 children with SNHL. The groups were matched based on better-ear pure-tone average, better-ear aided speech intelligibility index, gender, maternal education level, and newborn hearing screening result (i.e., pass or refer).

Data collection and analysis—Children and their families participated in an initial baseline visit, followed by visits twice a year for children under age 2 years and once a year for children older than 2 years. Paired-sample t-tests were used to compare children with ANSD to children with SNHL.

Results—Paired t-tests indicated no significant differences between the ANSD and SNHL groups on language and articulation measures. Children with ANSD displayed functional speech

Corresponding author: Elizabeth A. Walker, 250 Hawkins Drive, University of Iowa, Iowa City, IA 52252, 319-335-6851, elizabeth-walker@uiowa.edu.

The authors have no financial relationships relevant to this article to disclose.

perception skills in quiet. Although the number of participants was too small to conduct statistical analyses for speech perception testing, there appeared to be a trend in which the ANSD group performed more poorly in background noise with hearing aids, compared to the SNHL group.

Conclusions—The AAA Pediatric Amplification Guidelines recommend that children with ANSD receive a hearing aid trial if their behavioral thresholds are sufficiently high enough to impede speech perception at conversational levels. For children with ANSD in the mild to severe hearing loss range, the current results support that recommendation, as children with ANSD can achieve functional outcomes similar to peers with SNHL.

Keywords

Auditory neuropathy spectrum disorder; children; hearing aids

Introduction

Permanent childhood hearing loss (HL) occurs in one to three per thousand live births (Finitzo et al., 1998; Van Naarden et al., 1999). Among children with a permanent hearing loss, between 2 and 15% will have auditory neuropathy spectrum disorder (ANSD; Sininger, 2002; Rance, 2005; Vlastarakos et al., 2008). Individuals with ANSD present with an abnormal or absent neural response from the auditory nerve or auditory brainstem, in combination with preneural cochlear responses including otoacoustic emissions and/or cochlear microphonic (Starr et al., 1996). Sites of lesion may occur at several different locations along the auditory pathway, including the inner hair cells, the synapse between the inner hair cells and the auditory nerve, or the auditory nerve (Buchman et al., 2006). ANSD may occur as the result of certain risk factors such as prematurity, hyperbilirubinemia, or ototoxic medications, including furosemide, aminoglycosides, and vancomycin (Coenraad, Goedegebure, van Goudoever, & Hoeve, 2011; Lobarinas, Salvi, & Ding, 2013); it may also be due to syndromic or nonsyndromic genetic conditions (Roush et al., 2011). It is possible to identify ANSD at birth, but audiological management is complicated by the fact that objective measures such as auditory brainstem response (ABR) are absent or abnormal and not correlated with behavioral audiometric thresholds.

As would be expected based on the numerous potential sites of lesion and contributing factors, ANSD presents as heterogeneous with respect to a number of factors. Degree of hearing loss based on the audiogram may range from normal to profound (Rance & Starr, 2011). In some cases, the hearing thresholds have been reported to fluctuate or progress (Madden et al., 2002), although Rance (2014) reports that fluctuations in hearing level are rare. Speech perception outcomes are highly variable. There are several characteristic features of speech perception in ANSD: 1) speech perception abilities may be better or poorer than would be expected based on audibility (Rance, 2005) and 2) speech perception in noise is particularly difficult (Kraus et al., 2000; Rance & Barker, 2008). The difficulties with speech perception in noise may be due to a potential reduction in individual ability to integrate binaural cues, resulting in an impairment in localization skills that support listening in noise (Zeng et al., 2005). Some researchers have attempted to explain variability in speech perception abilities. Speech perception abilities appear to be related to the degree to which there are temporal processing deficits, as children with ANSD who have better

temporal resolution tend to have better speech recognition in quiet, unaided conditions (Rance, McKay, & Grayden, 2004). Given the varying presentations of ANSD, researchers and clinicians have noted that there is a need for more evidence-based research on appropriate clinical interventions for this population (Roush et al., 2011).

There are three current forms of hearing technology that are recommended as intervention for children with ANSD: cochlear implants, hearing aids, and FM technology. For the purposes of the current manuscript, we will focus on literature related to the first two options. A number of studies examined outcomes following cochlear implantation in children with ANSD (Shallop et al., 2001; Buss et al., 2002; Raveh et al., 2007; Rance & Barker, 2008; Teagle et al., 2010; Breneman et al., 2012). The majority of these studies focused on children with severe-to-profound hearing loss (Miyamoto et al., 1999; Trautwein et al., 2000; Buss et al., 2002; Mason et al., 2003; Vermeire et al., 2003; Jeong et al., 2007). Some researchers have indicated positive changes in pure-tone thresholds and speech perception outcomes following implantation (Madden et al., 2002; Mason et al., 2003). Several explanations have been posited for why cochlear implants may benefit some patients with ANSD: 1) the electrical stimulation from the array increases the number of firing neural elements, effectively bypassing the damaged inner hair cells and directly stimulating the spiral ganglion or 2) the electrical signal improves the synchronization of neural firing along the auditory nerve (Rance, 2014). Thus, for some individuals with ANSD who have greater degrees of temporal disruption, cochlear implants may lead to improvements in temporal resolution. Successful outcomes with cochlear implants may be mediated by the site of lesion, however, resulting in some reported cases of poorer progress with cochlear implants (Miyamoto et al., 1999; Neary & Lightfoot, 2012). Degree of hearing loss may also have an influence on the success of cochlear implants versus hearing aids for children with ANSD, as there is evidence of children who have less severe degrees of hearing loss and use hearing aids who show better aided speech recognition scores than children with cochlear implants (Rance et al., 2002). Furthermore, a significant number of children with ANSD may have other conditions or co-morbidities that negatively impact outcomes irrespective of the treatment approach (Teagle et al., 2010). Thus, even though cochlear implantation is often cited in the literature as a successful remediation approach for ANSD, clinicians and families should be cautious when deciding on the appropriate management technique (Rance, 2005).

The American Academy of Audiology (AAA) Pediatric Amplification guidelines state that children with ANSD should have a hearing aid trial if auditory thresholds are insufficient to support speech perception at conversational levels. This trial would consist of a designated time period of experience with appropriately fit hearing aids, although the guidelines are not specific on how long a trial should last. Even though the AAA guidelines recommend a hearing aid trial, there is a paucity of studies examining speech perception, language, or literacy outcomes for children with ANSD who use hearing aids. Of those studies that have been conducted, some have observed poor outcomes with hearing aids, leading some researchers to argue that hearing aids may provide limited benefit because they are merely amplifying an already distorted signal (Berlin, 1999; Berlin et al., 2003; Raveh et al., 2007). These findings have led to general uncertainty as to whether or not children with ANSD

should utilize hearing aids, but there are some limitations to applying this line of research for children who are hard of hearing.

One limitation is that these earlier studies have relied primarily on children with thresholds in the severe-to-profound range, who, due to gain constraints of acoustic amplification, likely would receive only minimal benefit from hearing aids. In contrast, children with thresholds in the mild to severe range have the potential to greatly benefit from the audibility provided by hearing aids. Second, these studies included minimal information regarding characteristics of amplification, such as level of aided audibility and amount of daily hearing aid use. Both level of aided audibility and amount of hearing aid use have been shown to influence outcomes for children with SNHL (Tomblin et al., 2014; Marnane & Ching, 2015; McCreery et al., accepted). Specifically in the ANSD population, some audiologists may follow recommendations that these children be fit with wide dynamic range hearing aids that provide less gain than prescribed based on hearing thresholds (Hood, 1998), resulting in possible under-amplification and limited access to the speech spectrum (Rance, 2005). Thus, it is important to document the level of aided audibility when describing outcomes in this population, because individuals may be intentionally underfit, relative to children with SNHL. It is also important to document the amount of daily HA use in children with ANSD. Children with SNHL who are older and have more severe degrees of hearing loss wear hearing aids more often than younger children and children with milder degrees of hearing loss (Walker et al., 2013). Thus far, there have been no investigations examining amount of hearing aid use in children with ANSD. Given the reported variations in speech perception abilities in children with ANSD, it is possible that families may see less benefit with amplification, leading to more inconsistent HA usage. It is also possible that families receive mixed messages from clinicians regarding expected benefit from amplification.

Finally, there is little information in the literature on timing of service delivery for children with ANSD (i.e., age at confirmation, early intervention, or HA fitting). Researchers have noted that audiological intervention may be delayed for children with ANSD until behavioral thresholds can be obtained (Ching et al., 2013). Deferment in hearing aid fitting may also occur because of concerns about causing acoustic trauma in young children (Rance, 2005). The AAA recommendations acknowledge that an ABR cannot provide valid estimates of behavioral thresholds for children with ANSD, in contrast to children with SNHL. Therefore, the guidelines suggest that hearing aids may initially be fit based on careful behavioral observations to sound, by the clinician and/or the parent, until a reliable behavioral audiogram can be obtained. However, we currently lack evidence-based protocols for using behavioral observations to fit hearing aids in a clinically valid manner. In summary, we know little about children with ANSD who are hard of hearing, have been early identified and have been fit using best practice recommendations, in comparison to children with similar degrees of SNHL.

In one of the few studies that did include information on hearing aid fitting procedures, Rance et al. (2007) examined speech and language outcomes in a group of 12 children with ANSD (mean age = 8.5 years), the majority of whom had hearing losses in the mild to severe range. All of the subjects were fit bilaterally with hearing aids using National Acoustics Laboratories (NAL) prescriptive targets for gain. The authors did not report aided

audibility levels, but it may be surmised that the children had optimal aided audibility based on use of best-practice hearing aid verification protocols by the Australian Hearing service (King, 2010). On average, participants demonstrated below-average performance compared to the test norms on standardized measures of receptive vocabulary and articulation, but no significant differences compared to a group of children with SNHL who were matched by age and degree of hearing loss.

More recently, Ching and her colleagues have investigated the outcomes of children with ANSD who participated in a population-based longitudinal study in Australia. Ching et al. (2013) examined a diverse group of 3-year old children with ANSD ($n = 47$). Twenty-seven used hearing aids, which were all fit based on NAL or Desired Sensation Level (DSL) prescriptive targets, using real ear to coupler difference (RECD) measures for verification. Nineteen children used cochlear implants. Of the 47 total children, 80% used an oral communication mode and 30% had at least one additional disability. Twenty-two children with hearing aids contributed outcomes data. The authors compared the ANSD group to a group of children with SNHL. Results indicated no significant differences between groups on measures of speech production, receptive and expressive language, and functional auditory skills. However, children with hearing aids and cochlear implants were combined into one group when examining scores in these separate domains. Thus, it is difficult to determine if the children with ANSD who used hearing aids experienced more difficulties within certain domains compared to those who used cochlear implants.

In summary, these two studies (Rance et al., 2008; Ching et al., 2013) suggest that there is additional need for research that explores the impact of hearing aid performance on children with ANSD in the mild to severe hearing loss range. As other researchers have noted (Rance, 2005; Roush et al., 2011), there is ambiguity regarding hearing aid management approaches for children with ANSD. This ambiguity in the literature makes it difficult to interpret results with this population. There is also minimal evidence regarding speech production and language outcomes for children with ANSD. The primary purpose of the current paper is to investigate speech perception, speech production, and language outcomes of children with ANSD who were participants in the Outcomes of Children with Hearing Loss (OCHL) study. Our goal is to provide valuable evidence to support clinical decisions about audiological management and intervention for this population. The participants in the current study, including those with ANSD, had hearing loss in the mild to moderately severe range, utilized bilateral hearing aids, and were fit based on best practice recommendations. This paper specifically addresses the following research questions:

1. How do children with ANSD compare to children with SNHL in terms of characteristics of amplification, including ages at service delivery, aided audibility, and amount of daily HA use?
2. How do children with ANSD compare to children with SNHL on measures of language, speech production, and speech perception?

Method

Participants

Participants included children with hearing loss recruited as part of the OCHL study. This multi-center study was designed to explore the developmental outcomes of children who are hard of hearing. Speech perception and communication outcomes data were gathered in an accelerated longitudinal design, with entry into the study between six months and seven years of age. Each child was seen for up to five annual visits and the test protocol was designed to be appropriate for each age level (for more information regarding the accelerated longitudinal design, see Holte et al., 2012). Three sites were involved: Boys Town National Research Hospital, The University of North Carolina at Chapel Hill (UNC), and the University of Iowa. Each site recruited from surrounding areas and states. Children recruited for the OCHL study had a permanent, bilateral HL (sensorineural, mixed, and permanent conductive) with a better-ear three- or four-frequency PTA no better than 25 dB HL and no poorer than 75 dB HL. Children with significant cognitive, visual, or motor impairments were excluded from participation. For all participants, at least one primary caregiver spoke English in the home. Children who used manually-coded English or American Sign Language as their primary mode of communication were excluded from the study.

The children described in the current manuscript received pediatric hearing aid fitting services that were uniform and consistent with the AAA Pediatric Amplification best-practice guidelines (AAA, 2013). In other words, each child's managing audiologist fit the hearing aids according to DSL v5.0 gain targets for speech and MPO targets for pure tones using either simulated real-ear measurements or in-situ real-ear probe microphone measurements (Marlene Bagatto et al., 2005; Scollie et al., 2005). Eleven of the children's fitting audiologists reported that these best practice recommendations were followed by completing an online survey that asked about fitting procedures and verification techniques for that client. The twelfth participant's fitting audiologist did not complete a survey; therefore, it is unknown if that child's hearing aids were fit and verified according to best practice protocols. Due to the small number of participants with ANSD in this study, we decided to keep this participant in the dataset.

Participants in the current study included 12 children (6 females) with ANSD. Eleven had bilateral ANSD and one child had ANSD in one ear and typical SNHL in the opposite ear. At the initial testing visit, the children with ANSD were between the ages of 9 months and 7 years, 0 months ($M = 38.42$ months; $SD = 24.83$ months). Ten children referred on the newborn hearing screen (NHS), two were identified later (one passed the NHS, which was conducted with otoacoustic emissions and the other child's NHS status was unknown; both were later identified due to speech delays).

Ten of the children were recruited for OCHL by the UNC site and two of the children were recruited by the Boys Town site. Due to the longitudinal nature of the study, most of the children in the ANSD group participated in multiple test visits. In the current analysis, audiological and hearing aid data, including better-ear Speech Intelligibility Index (SII) and daily hours of use, are examined at the first visit in which a full dataset was available for

ANSD subjects to avoid analysis of multiple data points for the same children. Speech perception and language outcomes are examined at the visit in which those data were collected; thus, some participants within the ANSD group may contribute data at several visits, but none are counted for the same speech or language test more than one time. Some children contributed data for specific standardized tests at multiple visits (e.g., GFTA-2 at ages 5 and 7). In those cases, only scores at one visit were included, and the data that were used were selected at random.

The mean better-ear pure-tone average (BEPTA) for the ANSD group at the baseline visit was 57.63 dB HL ($SD = 11.60$). One participant had a mild hearing loss (between 20 to 39 dB HL), 6 had a moderate hearing loss (between 40 to 59 dB HL) and 5 had a moderately-severe hearing loss (between 60 to 75 dB HL). One child with ANSD presented with a progressive hearing loss during the course of the study, as determined by more than a 10 dB HL increase in BEPTA (i.e., decline in thresholds) between the first and last visit. The other 11 children demonstrated stable thresholds throughout the course of the study.

A cross-sectional sample of 22 children (13 females) with bilateral SNHL served as a comparison group (Boys Town, $n = 5$; Iowa, $n = 5$; UNC, $n = 12$). The ANSD and SNHL pairs were matched as closely as possible on chronological age, BEPTA and better-ear aided SII, gender, and maternal education level. We also controlled for NHS status by only matching children who referred on the NHS together, and only matching children who were identified after the NHS together. All of the children in the SNHL group were also fitted with hearing aids based on best-practice protocols.

Data collection

As part of the OCHL study, children and their families participated in an initial baseline visit, followed by visits twice a year for children under age 2 years and once a year for children older than 2 years. At the initial visit, parents completed an intake interview with an examiner. Intake questions documented demographic information related to the child and family, including predictor variables incorporated in the present study (i.e., maternal education level, gender, and age at confirmation of hearing loss, enrollment in early intervention, and HA fitting). It should be noted that age at confirmation of hearing loss only referred to when the child's hearing loss was confirmed through diagnostic testing, and did not specify when they were diagnosed with ANSD or when reliable behavioral thresholds were obtained. The intake interview also documented birth history, including length of pregnancy in weeks, birth weight, and medical treatments and complications related to the pregnancy and birth.

Audiologic assessment—An experienced pediatric audiologist completed all behavioral hearing assessments, with an assistant participating in assessments as needed. The audiologist attempted to obtain air-conduction and bone-conduction thresholds at 500, 1000, 2000, and 4000 Hz at a minimum, using visual reinforcement audiometry, conditioned play audiometry, or conventional audiometry depending on the age of the child. All attempts were made to obtain ear-specific thresholds using insert earphones, supra-aural headphones, or the child's own earmolds paired with insert earphones. The audiologist obtained

soundfield thresholds if the child would not tolerate the testing with earphones or headphones. If a full audiogram could not be completed, the audiologist obtained a copy of the child's most recent unaided audiogram from their clinical audiologist (in the majority of cases this audiogram was obtained within three months of the OCHL test visit). The 4-frequency BEPTA was calculated for subsequent analyses (3-frequency BEPTA when 4-frequency thresholds were not available).

Hearing aid use—As part of each visit, the caregiver completed an interview that asked about daily hearing aid use (see Walker et al. 2013, for an example of the hearing aid use questionnaire). Parents estimated the average amount of time the child used hearing aids per day during the week and on the weekends. To calculate daily use, we devised a weighted hearing aid use measure in which parents' estimates of weekday use was multiplied by 0.71 (5/7 days of the week) and weekend use was multiplied by 0.29 (2/7 days of the week). The two values were then added together. This weighted measure was used to represent hearing aid use time for all participants.

Hearing aid electroacoustic measures—The pediatric audiologist completed electroacoustic hearing aid measurements, including total harmonic distortion, frequency range, and output sound pressure level at 90 dB (OSPL90) following ANSI S3.22 (2003). The audiologist also conducted probe microphone measures to estimate the current speech audibility for the participant at the time of the visit, measuring the RECD when possible. When the RECD could not be measured due to limited cooperation or subject noise, an age-related average RECD was used to estimate the acoustic characteristics of the child's occluded ear canal (Bagatto et al, 2002). Simulated speechmapping was then completed in the 2 cc coupler. Audioscan Verifit™ software calculated the aided SII for all of the participants using the standard male speech signal (carrot passage) presented at 65dB SPL (average speech), following ANSI S3.2–2009.

Speech and language assessment—Examiners administered standardized tests of speech production, language, and phonological processing in a quiet testing room or a mobile testing van. The *Goldman Frisbie Test of Articulation-2* (GFTA-2; Goldman & Frisbie, 2000) is a standardized measure of articulation, in which examinees label single word pictures. The *Peabody Picture Vocabulary Test-4* (PPVT-4; Dunn & Dunn, 2007) is a standardized measure of receptive vocabulary, in which the examiner says a word that describes one of the pictures on a page, and the participant identifies the correct picture. The *Vineland Adaptive Behavior Scales-II* (Vineland; Sparrow, Cicchetti, & Balla, 2005) is a parent-report questionnaire. It examines adaptive behavior, including receptive and expressive language, writing, interpersonal and coping skills, and fine and gross motor skills. For the current analysis, we used the adaptive behavior composite standard score, which is a composite of all of the subtests scored on the Vineland. The *Comprehensive Assessment of Spoken Language* (CASL; Carrow-Woolfolk, 1999) is a standardized measure of global language development. Subtests are designed to evaluate receptive and expressive language in areas of lexical/semantic, syntactic, supralinguistic, and pragmatic development. For the purposes of the current analysis, we included scores on the Pragmatic Judgment and Syntax Constructions subtests to measure pragmatics and expressive morphosyntax scores,

respectively. For all standardized measures, standard scores were derived based on the children's uncorrected chronological age, in order to be consistent across all participants at each age. In other words, we did not adjust for prematurity in the current analysis.

Auditory Development and Speech Perception—The LittEARS Auditory Questionnaire was administered as a measure of early auditory development (Coninx et al., 2009). The measure has been validated for use with children who are hard of hearing who wear hearing aids (Bagatto et al. 2010). The LittEARS is a parent-report questionnaire consisting of 35 questions. Parents indicate whether (1) or not (0) their child exhibits the behavior described in each question. The LittEARS was completed by parents at their child's 12 months, 18 months and 2 year visits. Raw scores were compared to the normative range in the LittEARS manual, based on the uncorrected chronological age of the children.

The *Phonetically-Balanced Kindergarten List* (PBK; Haskins, 1949) was used to assess speech perception in quiet at 5 years of age and older. The PBK test is an open set speech perception measure. The child listened to a recorded list of words presented at 65 dBA at 0° azimuth from a loudspeaker, and repeated back the words to the audiologist. Participants wore hearing aids during test administration. The test is scored as percent correct out of 50 words.

Seven, 8-, and 9-year-olds were tested on their ability to perceive single words in background noise using the Computer-Assisted Speech Perception Assessment (CASPA; Mackersie et al., 2001). The CASPA was presented with noise at 55 dBA and speech at 65 dBA (+10 dB SNR) in the aided condition.

Data analysis

The primary goal of the present study was to compare speech perception, speech production, and language outcomes of children with ANSD to children with SNHL, all of whom were fitted with amplification using the AAA Pediatric Amplification best practice guidelines. Data regarding HA use, PTA, and aided audibility were explored descriptively, in addition to paired t-tests to compare the ANSD and SNHL groups. We compared outcomes between children with ANSD and SNHL using both inferential statistics (paired t-tests) and non-parametric statistics (Wilcoxon Signed Rank test).

Results

Demographic and audiological characteristics of children with ANSD or SNHL

Table 1 displays data regarding birth history and risk factors in the two groups. The majority of children with ANSD (82%; 9/11) had medical risk factors; in particular, a number of participants were premature, had low birth weight, and/or hyperbilirubinemia. At the time of enrollment in the study, these children demonstrated non-verbal cognitive abilities within normal limits, and did not have severe motor or visual impairments that would have excluded them from participation. Most of the children in the SNHL group did not present with risk factors. Specifically, most of the children with SNHL had an uncomplicated birth history and were full term.

Table 2 describes characteristics related to audiological and service provision factors for the two groups. The data in Table 2 are from the baseline visit of the ANSD participants, and the SNHL participants are matched by chronological age, better-ear PTA and SII. There was one exception to this procedure; one child with ANSD did not have audibility data collected at the baseline visit. Data for that child were obtained from his second visit (with the corresponding SNHL control subject matched for age, PTA, and SII at that respective visit). As expected, there were no significant differences in better-ear PTA or better-ear SII in the ANSD and SNHL groups, because the participants were matched on these factors. Figure 1 displays aided audibility for an average speech input level as a function of BEPTA in children with ANSD and children with SNHL. The 95% confidence intervals for the normative SII range by degree of hearing loss (M. P. Bagatto, Brown, Moodie, & Scollie, 2011) are displayed as dashed lines. All of the participants demonstrated SII levels within the average range.

Results indicated that there were no significant differences in amount of daily hearing aid use based on parent report. When children with ANSD were paired with children with SNHL on age, PTA, and aided audibility, they did not wear hearing aids more often or less often compared to the SNHL group. In terms of service delivery, there were no significant differences between groups for age at first evaluation, confirmation of hearing loss, and entry into early intervention. There was a significant difference in age at HA fitting, $t(10) = 3.63$, $p = .005$. Children with ANSD were fit with hearing aids at significantly older ages ($X = 13.73$ months) compared to children with SNHL ($X = 8.18$ months), with an average difference of 5.55 months between groups.

Standardized test comparisons between children with ANSD and children with SNHL

Expressive morphosyntax—Children in the ANSD group ($n = 9$) demonstrated a mean standard score of 86.56 on the CASL Syntax Construction subtest ($SD = 23.91$) and children in the matched SNHL group ($n = 9$) demonstrated a mean standard score of 94.67 ($SD = 21.45$). The results of the paired samples t-test were not significant, $t(8) = -1.11$, $p = .30$. Due to the small sample size, a Wilcoxon Signed Rank test was also conducted with consistent results. Three children in the ANSD group and five children in the SNHL group showed scores that were within the average range or better compared to standardized test norms. Figure 2 and Table 3 display the individual CASL Syntax Construction subtest standard scores for both groups.

Pragmatics—Children in the ANSD group ($n = 9$) demonstrated a mean standard score of 87.78 on the CASL Pragmatic Judgment subtest ($SD = 18.05$) and children in the matched SNHL group ($n = 9$) demonstrated a mean standard score of 94.44 ($SD = 17.76$). The results of the paired samples t-test were not significant, $t(8) = -.93$, $p = .38$. Due to the small sample size, a Wilcoxon Signed Rank test was also conducted with consistent results. Four children in the ANSD group and seven children in the SNHL group showed scores that were within the average range or better compared to standardized test norms. Figure 2 and Table 3 display the individual CASL Pragmatic Judgment subtest standard scores for both groups.

Vocabulary—Children in the ANSD group ($n = 5$) demonstrated a mean standard score of 87.40 on the PPVT-4 receptive vocabulary test ($SD = 20.40$) and children in the SNHL group ($n = 5$) demonstrated a mean standard score of 104.60 ($SD = 13.09$). We did not conduct inferential or nonparametric statistics due to the very small sample size. Three children in the ANSD group and all five children in the SNHL group showed scores that were within the average range or better compared to standardized test norms. Figure 2 and Table 4 displays the individual PPVT-4 standard scores for both groups.

Articulation—Children in the ANSD group ($n = 9$) demonstrated a mean standard score of 86.33 on the GFTA-2 ($SD = 14.45$) and children in the SNHL group ($n = 9$) demonstrated a mean standard score of 90.22 ($SD = 18.89$). The results of the paired samples t -test were not significant, $t(8) = -.60$, $p = .57$. Due to the small sample size, a Wilcoxon Signed Rank test was also conducted with consistent results. Four children in the ANSD group and seven children in the SNHL group showed scores that were within the average range or better compared to standardized test norms. Figure 2 and Table 5 displays the distribution of the GFTA-2 standard scores for both groups.

Adaptive behavior—Children in the ANSD group ($n = 8$) demonstrated a mean standard score of 91.25 on the Vineland Adaptive behavior composite measure ($SD = 13.01$) and children in the SNHL group ($n = 8$) demonstrated a mean standard score of 98.00 ($SD = 9.40$). The results of the paired samples t -test were not significant, $t(7) = -1.21$, $p = .27$. Due to the small sample size, a Wilcoxon Signed Rank test was also conducted with consistent results. Six children in the ANSD group and all eight children in the SNHL group showed scores that were within the average range or better compared to standardized test norms. Figure 2 and Table 6 displays the distribution of the Vineland adaptive behavior composite standard scores for both groups.

Auditory Development—Figure 3 shows individual data for the parent-report auditory questionnaire, the LittEARS. The established ceiling criterion for this test is a raw score of 28 (Tsiakpini et al., 2004). The average raw score for the ANSD group was 27.75 ($n = 4$; $SD = 3.20$), while the average raw score for the SNHL group was 29 ($n = 4$; $SD = 6.06$). Three out of the four children with ANSD had scores within the normal range on this measure, based on the LittEARS normative data. One child with ANSD (DBQ at the 18 month visit) fell below the normative range, suggesting significant delays in early auditory behavior. All four of the children with SNHL scored within the normal range for their ages.

Speech perception—A limited number of children contributed unique data for speech perception measures because of the accelerated longitudinal design of the study (i.e., there were fewer children tested after age 5 years). Children in the ANSD group ($n = 4$) demonstrated an average score of 77% correct on the PBK ($SD = 10.39$) and children in the SNHL group ($n = 3$) demonstrated an average score of 83% ($SD = 1.15$). Figure 1 shows open-set PBK scores for the ANSD and SNHL groups, plotted as a function of 4-frequency better-ear PTA. All four children with ANSD displayed functional speech perception skills in quiet.

Only three children in the ANSD group contributed unique data on the CASPA. Their individual scores in the +10 SNR aided condition (phonemes correct) were 60%, 77%, and 80% correct. The SNHL matched pair scores were 97%, 93%, and 97%, respectively. Although the number of participants was too small to conduct statistical analyses for the speech perception testing, there does appear to be a trend in which the ANSD group performed more poorly in background noise with hearing aids, compared to the SNHL group. These results should be interpreted cautiously, however, given the small number of subjects.

Discussion

The primary objective of this study was to describe language, speech production, and speech perception outcomes in children with ANSD who were fitted with hearing aids using best practice recommendations. We also examined timing of service provision and amount of daily hearing aid use in children with ANSD, compared to children with SNHL.

There has been a great deal of debate regarding clinical management of children with ANSD. Some researchers have recommended minimal use of acoustic amplification (Berlin, 1999; Berlin et al., 2002), while others have suggested using a conservative approach of providing low-gain wide-dynamic range compression hearing aids, compared to what would be recommended by prescriptive targets based on hearing thresholds and RECD (Hood, 1998). Berlin et al. (1998) reported on two case studies of children who did poorly with hearing aids, but both of those participants had minimal experience with amplification and were fit with hearing aids providing gain lower than would be recommended by prescriptive targets. The difficulty with interpreting the results of studies on ANSD and acoustic amplification is there is little reported evidence on how children with ANSD with hearing thresholds in the mild to severe range perform with appropriately-fit hearing aids set to DSL or NAL prescriptive targets.

All of the children in the current study were fit with hearing aids based on the AAA recommendations for pediatric amplification (with the possible exception of one child with ANSD, whose audiologist did not complete the service provider survey). The current results provide support for conducting a hearing aid trial with appropriately-fit amplification for children with ANSD who have behavioral thresholds in the mild to severe range. The results also support current hearing aid verification recommendations: audiologists should be verifying gain and comparing to prescriptive targets through the use of probe microphone measurements of the hearing aid in the child's ear canal or simulated measurements of the hearing aid output in a coupler with a real-ear-to-coupler-difference measure (Bagatto et al. 2010; King, 2010; AAA, 2013). These results do not support the need to provide differential intervention or amplification management (i.e., fitting to prescriptive targets and verification techniques) for children who have ANSD with mild-severe behavioral thresholds, and do not support the provision of low-gain hearing aids for this population.

How do children with ANSD compare to children with SNHL in terms of characteristics of amplification, including ages at service delivery, aided audibility, and amount of daily HA use?

In order to understand the outcomes of children with ANSD, we must also understand what their cumulative auditory experiences represent: when are they receiving audiologic intervention, how much access to the speech spectrum do they get from their hearing aids, and how often do they wear hearing aids on a daily basis? Results from the OCHL cohort demonstrate that each of these factors has an effect on outcomes for children who are hard of hearing (Tomblin et al., accepted; McCreery et al., accepted). Unfortunately, we do not have enough participants to determine if these factors influence outcomes for children with ANSD, but we can compare the group to children with SNHL to see if they differ in any of these variables. For most of the factors, including age at confirmation of hearing loss, age at early intervention, aided audibility, and amount of daily hearing aid use, there were no significant differences between groups. It is not surprising that the ANSD and SNHL groups were similar in aided audibility, as that was one of the variables on which the groups were matched. It is encouraging to note, however, that all of the children with ANSD, who had hearing aids fit and verified using AAA guidelines, showed aided audibility levels that were within the average range compared to normative data that were appropriate for their PTAs (Bagatto et al., 2011). Furthermore, they wore hearing aids 11.5 hours per day on average, which can be considered full-time use. It might be presumed that children with ANSD would wear amplification less often compared to children with SNHL, if parents are not seeing benefit or children are receiving a distorted, amplified signal. Based on the current, albeit small, group of children with ANSD, this assumption does not appear to be the case.

On another positive note, children with ANSD did not show differences from their SNHL matches in age at confirmation or enrollment in early intervention. The median age at enrollment in early intervention (6 months) was actually consistent with recommendations by the Joint Committee on Infant Hearing (JCIH, 2007) for initiation of early intervention services. It is also encouraging to note that on average, children with ANSD were not delayed in receipt of early intervention, compared to children with SNHL. Age at hearing aid fitting was the one service delivery variable that was significantly different between the ANSD and SNHL groups, with the ANSD group being fit at significantly later ages. This result is consistent with observations by other researchers that audiological intervention may be delayed for children with ANSD until reliable behavioral thresholds can be obtained (Ching et al., 2013). Behavioral responses through visual reinforcement audiometry (VRA) cannot be consistently obtained until infants are around eight months of age (Widen et al., 2000) and likely at even later ages for infants who are premature. Although the AAA guidelines recommend the use of behavioral observations to fit hearing aids prior to obtaining reliable thresholds, we currently lack strong evidence to support the use of behavioral observation audiometry for appropriate hearing aid fittings in children. Further research is needed to explore the validity of clinical protocols (before 6–8 months of age) for fitting hearing aids in children with ANSD.

How do children with ANSD compare to children with SNHL on measures of language, speech production, and speech perception?

The results of the current study showed no significant differences between children with ANSD and SNHL in terms of outcomes in a variety of different domains. The two groups were well-matched on a number of important factors, including chronological age, degree of hearing loss, amount of aided audibility, maternal education level, gender, and NHS status. The groups were not matched on birth history – the ANSD group presented with a more complicated birth history than the SNHL group, yet this did not appear to have an effect on group-level comparisons, even without adjusting age-based standard scores for prematurity.

These findings are consistent with two other reports on speech and language outcomes of children with ANSD who were fit with hearing aids based on best practice recommendations (Ching et al., 2013; Rance et al., 2007). Rance and colleagues described receptive vocabulary and articulation skills in 12 children with ANSD and 12 children with SNHL between the ages of 4 and 13 years, matched on age and PTA. They found no significant differences between groups on either measure. Ching et al. compared children with ANSD to children with SNHL, all of whom were 3 years of age. They found no significant differences between groups on measures of global language or speech production. Approximately half of the children with ANSD scored within the average range for expressive and receptive language and speech production. The current results expand upon those previous studies, by including measures of expressive morphosyntax, pragmatics, and adaptive behavior (which can be thought of as an overall measure of daily personal and social skills). Taken together, these studies all consistently demonstrate that some children who have ANSD in the mild to severe range can acquire functional language skills in the areas of form, content, and use. These skills can be acquired at levels comparable to children with SNHL, and even within the average range for children with normal hearing.

Although limited in number, the speech perception performance of the current group of children is consistent with the previous literature. On a quiet, open-set monosyllabic word recognition task, in the aided condition, children with ANSD demonstrated scores that were comparable to children with SNHL, suggesting functional speech perception skills in quiet. Furthermore, PBK scores were all within the range that would be expected for adults with SNHL, based on norms provided by Yellin et al. (1989). Rance (2005, 2013) has reported similar findings for some children with ANSD, although he has also shown that a significantly large number of children with ANSD (56% out of 46 children) performed below the normative range. Rance's data in the 2005 paper consists of a compilation of scores from 10 different studies, and little is known about the aided audibility levels of the children in those studies.

Speech perception scores in noise presented a different picture. Even in a favorable SNR condition (+10 dB), the children with ANSD demonstrated significant difficulty compared to the children with SNHL. This finding is also consistent with previous research on individuals with ANSD, such that children and adults have displayed reliably poor performance in background noise (Rance, 2005; Starr et al., 1998). Therefore, an important recommendation for this population is the use of FM systems to improve the ability to listen in noise (Kraus et al., 2000; Rance, 2005). This is particularly true for children with ANSD

and thresholds in the mild to severe range, who are expected to learn academic material while listening in noisy, reverberant acoustics conditions in mainstream classroom settings (Crandell & Smaldino, 2000).

Limitations and Future Directions

Although this paper provides important outcomes evidence regarding clinical management for children with ANSD, we acknowledge that there are several limitations that could be addressed through further research. The current study did not include measures of psychosocial or academic outcomes, including reading or classroom behavior. To date, there have been no studies examining outcomes in these areas. Future directions should include investigations of literacy, academics, and socioemotional aspects of development, as these areas may provide more insight into long-term outcomes of this population.

Another limitation is that we were limited to investigating a very small sample of participants with ANSD. The primary goal of the OCHL study was to examine outcomes of children who were hard of hearing, and children with ANSD were not specifically targeted during recruitment efforts. Furthermore, not every participant had data for every test measure due to the accelerated longitudinal design of the study. This design allowed us to collect data on a wide range of developmental levels and a broad class of test domains, but it limited our ability to analyze specific groups of children with hearing loss on particular measures, as we have attempted to do in the present manuscript.

A third point is that there needs to be caution in generalizing the current findings to all children with ANSD with behavioral thresholds in the mild to moderately-severe range. Recruitment for the OCHL study focused on children with bilateral hearing loss who used hearing aids and did not have major secondary disabilities. Children who had received cochlear implants at the time of enrollment were excluded from the OCHL study, to allow investigators to focus on the influence of hearing aids on outcomes. Children who previously tried hearing aids, but showed little benefit, may have received cochlear implants and were no longer candidates for enrollment into the OCHL study. Furthermore, children with additional conditions (i.e., autism spectrum disorder, developmental delays, visual impairments) were excluded from the study. Therefore, the outcomes reported in this paper could over-represent the success that children who are hard of hearing with ANSD can have with hearing aids.

Finally, we did not collect data involving psychophysical measures such as temporal resolution, which have been shown to be predictive of the speech perception difficulties in children with ANSD (Rance et al., 2004). Such measures were not documented in the OCHL study, but further research in this area may aid in the development of specific processing strategies that help with the perception of temporal cues (Rance, 2005) and explain variability in developmental speech and language outcomes.

Conclusion

The AAA Pediatric Amplification Guidelines recommend that children with ANSD receive a hearing aid trial if their behavioral thresholds are reliable and sufficiently high enough to

impede speech perception at conversational levels. For children with ANSD in the mild to severe hearing loss range, the current results appear to support those guidelines.

Acknowledgments

Funded by NIH/NIDCD R01DC013591, R01DC009560, P50DC000242

Abbreviations

AAA	American Academy of Audiology
ABR	Auditory brainstem response
ANSD	Auditory neuropathy spectrum disorder
CASL	Comprehensive Assessment of Spoken Language
CASPA	Computer-aided Speech Perception Assessment
DSL	Desired Sensation Level
GFTA	Goldman-Fristoe Test of Articulation
HL	Hearing loss
SNHL	sensorineural hearing loss
NAL	National Acoustics Laboratory
OCHL	Outcomes of Children with Hearing Loss
PPVT	Peabody Picture Vocabulary Test
PBK	Phonetically-balanced Kindergarten lists
PTA	Pure-tone average
RECD	Real ear to coupler difference
SII	Speech intelligibility index

References

- American Academy of Audiology. Clinical practice guidelines: Pediatric amplification. Reston, VA: 2013.
- Bagatto MP, Brown CL, Moodie ST, Scollie SD. External validation of the LittleEARS® Auditory Questionnaire with English-speaking families of Canadian children with normal hearing. *International Journal of Pediatric Otorhinolaryngology*. 2011; 75(6):815–817. [PubMed: 21492945]
- Bagatto M, Moodie S, Scollie S, Seewald R, Moodie S, Pumford J, Liu KR. Clinical protocols for hearing instrument fitting in the Desired Sensation Level method. *Trends in Amplification*. 2005; 9(4):199–226. [PubMed: 16424946]
- Bagatto M, Scollie SD, Hyde M, Seewald R. Protocol for the provision of amplification within the Ontario infant hearing program. *International Journal of Audiology*. 2010; 49(sup1):S70–S79. [PubMed: 20109090]
- Bagatto M, Scollie SD, Seewald RC, Moodie KS, Hoover BM. Real-ear-to-coupler difference predictions as a function of age for two coupling procedures. *Journal of the American Academy of Audiology*. 2002; 13(8):407–415. [PubMed: 12371658]

- Berlin CI. Auditory neuropathy: using OAEs and ABRs from screening to management. Paper presented at the Seminars in Hearing. 1999
- Berlin CI, Hood L, Morlet T, Rose K, Brashears S. Auditory neuropathy/dys-synchrony: Diagnosis and management. *Mental Retardation and Developmental Disabilities Research Reviews*. 2003; 9(4):225–231. [PubMed: 14648814]
- Breneman AI, Gifford RH, DeJong MD. Cochlear implantation in children with auditory neuropathy spectrum disorder: long-term outcomes. *Journal of the American Academy of Audiology*. 2012; 23(1):5–17. [PubMed: 22284837]
- Buchman CA, Roush PA, Teagle HF, Brown CJ, Zdanski CJ, Grose JH. Auditory neuropathy characteristics in children with cochlear nerve deficiency. *Ear and Hearing*. 2006; 27(4):399–408. [PubMed: 16825889]
- Buss E, Labadie RF, Brown CJ, Gross AJ, Grose JH, Pillsbury HC. Outcome of cochlear implantation in pediatric auditory neuropathy. *Otology & Neurotology*. 2002; 23(3):328–332. [PubMed: 11981390]
- Carrow-Woolfolk, E. CASL: Comprehensive assessment of spoken language. American Guidance Services; 1999.
- Ching TY, Day J, Dillon H, Gardner-Berry K, Hou S, Seeto M, Zhang V. Impact of the presence of auditory neuropathy spectrum disorder (ANSD) on outcomes of children at three years of age. *International Journal of Audiology*. 2013; 52(S2):S55–S64. [PubMed: 24350696]
- Coenraad S, Goedegebure A, van Goudoever JB, Hoeve L. Risk factors for auditory neuropathy spectrum disorder in NICU infants compared to normal-hearing NICU controls. *The Laryngoscope*. 2011; 121(4):852–855. [PubMed: 21305553]
- Coninx F, Weichbold V, Tsiakpini L, Autrique E, Bescond G, Tamas L, Le Maner-Idrissi G. Validation of the LittlEARS Auditory Questionnaire in children with normal hearing. *International Journal of Pediatric Otorhinolaryngology*. 2009; 73(12):1761–1768. [PubMed: 19836842]
- Crandell CC, Smaldino JJ. Classroom acoustics for children with normal hearing and with hearing impairment. *Language, Speech, and Hearing Services in Schools*. 2000; 31(4):362–370.
- Dunn, D.; Dunn, L. Peabody Picture Vocabulary Test 4. Minneapolis, MN: NCS Pearson: Inc; 2007.
- Finitzo T, Albright K, O'Neal J. The newborn with hearing loss: detection in the nursery. *Pediatrics*. 1998; 102(6):1452–1460. [PubMed: 9832584]
- Gilbertson M, Kamhi AG. Novel word learning in children with hearing impairment. *Journal of Speech, Language, and Hearing Research*. 1995; 38(3):630–642.
- Goldman, R.; Fristoe, M. Goldman-Fristoe Test of Articulation 2. Circle Pines, MN: American Guidance Service; 2000.
- Haskins, J. Kindergarten phonetically balanced word lists (PBK). St. Louis: Auditec; 1949.
- Joint Committee on Infant Hearing. Year 2007 position statement: principles and guidelines for early hearing detection and intervention programs. *Pediatrics*. 2007; 120(4):898–921. [PubMed: 17908777]
- Holte L, Walker E, Oleson J, Spratford M, Moeller MP, Roush P, Tomblin JB. Factors influencing follow-up to newborn hearing screening for infants who are hard of hearing. *American Journal of Audiology*. 2012; 21(2):163–174. [PubMed: 22585937]
- Hood LJ. Auditory neuropathy: what is it and what can we do about it? *The Hearing Journal*. 1998; 51(8):10–12.
- Jeong S-W, Kim L-S, Kim B-Y, Bae W-Y, Kim J-R. Cochlear implantation in children with auditory neuropathy: Outcomes and rationale. *Acta Oto-Laryngologica*. 2007; 127(S558):36–43.
- King AM. The national protocol for paediatric amplification in Australia. *International Journal of Audiology*. 2010; 49(sup1):S64–S69. [PubMed: 19919326]
- Kraus N, Bradlow A, Cheatham M, Cunningham J, King C, Koch D, Wright B. Consequences of neural asynchrony: a case of auditory neuropathy. *Journal of the Association for Research in Otolaryngology*. 2000; 1(1):33–45. [PubMed: 11548236]
- Lobarinas E, Salvi R, Ding D. Insensitivity of the audiogram to carboplatin induced inner hair cell loss in chinchillas. *Hearing Research*. 2013; 302:113–120. [PubMed: 23566980]

- Mackersie CL, Boothroyd A, Minniear D. Evaluation of the Computer-assisted Speech Perception Assessment Test (CASPA). *Journal of the American Academy of Audiology*. 2001; 12(8):390–397. [PubMed: 11599873]
- Madden C, Rutter M, Hilbert L, Greinwald JH Jr, Choo DI. Clinical and audiological features in auditory neuropathy. *Archives of Otolaryngology-Head & Neck Surgery*. 2002; 128(9):1026–1030. [PubMed: 12220206]
- Marnane V, Ching TY. Hearing aid and cochlear implant use in children with hearing loss at three years of age: Predictors of use and predictors of changes in use. *International Journal of Audiology*. 2015; (0):1–8.
- Mason JC, De Michele A, Stevens C, Ruth RA, Hashisaki GT. Cochlear implantation in patients with auditory neuropathy of varied etiologies. *The Laryngoscope*. 2003; 113(1):45–49. [PubMed: 12514381]
- McCreery RW, Walker E, Spratford M, Oleson J, Bentler R, Holte L, Roush P. Longitudinal results from auditory development questionnaires and speech recognition in children who are hard of hearing. *Ear and Hearing*. accepted.
- Miyamoto RT, Kirk KH, Renshaw J, Hussain D. Cochlear implantation in auditory neuropathy. *The Laryngoscope*. 1999; 109(2):181–185. [PubMed: 10890762]
- Neary W, Lightfoot G. Auditory neuropathy spectrum disorder: Examples of poor progress following cochlear implantation. *Audiological Medicine*. 2012; 10(3):143–150.
- Rance G. Auditory neuropathy/dys-synchrony and its perceptual consequences. *Trends in Amplification*. 2005; 9(1):1–43. [PubMed: 15920648]
- Rance, G. A Sound Foundation through Early Amplification: Proceedings of the 2013 International Conference. Chicago, IL: Phonak AG; 2014. An update on auditory neuropathy spectrum disorder in children; p. 137-144.
- Rance G, Barker EJ. Speech perception in children with auditory neuropathy/dyssynchrony managed with either hearing aids or cochlear implants. *Otology & Neurotology*. 2008; 29(2):179–182. [PubMed: 18165792]
- Rance G, Barker EJ, Sarant JZ, Ching TY. Receptive language and speech production in children with auditory neuropathy/dyssynchrony type hearing loss. *Ear and Hearing*. 2007; 28(5):694–702. [PubMed: 17804983]
- Rance G, Cone-Wesson B, Wunderlich J, Dowell R. Speech perception and cortical event related potentials in children with auditory neuropathy. *Ear and hearing*. 2002; 23(3):239–253. [PubMed: 12072616]
- Rance G, McKay C, Grayden D. Perceptual characterization of children with auditory neuropathy. *Ear and Hearing*. 2004; 25(1):34–46. [PubMed: 14770016]
- Rance G, Starr A. Auditory neuropathy/dys-synchrony type hearing loss. *Comprehensive handbook of pediatric audiology*. 2011:225–242.
- Raveh E, Buller N, Badrana O, Attias J. Auditory neuropathy: clinical characteristics and therapeutic approach. *American Journal of Otolaryngology*. 2007; 28(5):302–308. [PubMed: 17826530]
- Roush P, Frymark T, Venediktov R, Wang B. Audiologic management of auditory neuropathy spectrum disorder in children: a systematic review of the literature. *American Journal of Audiology*. 2011; 20(2):159–170. [PubMed: 21940978]
- Scollie S, Seewald R, Cornelisse L, Moodie S, Bagatto M, Lurnagaray D, Pumford J. The desired sensation level multistage input/output algorithm. *Trends in Amplification*. 2005; 9(4):159–197. [PubMed: 16424945]
- Shalloo JK, Peterson A, Facer GW, Fabry LB, Driscoll CL. Cochlear implants in five cases of auditory neuropathy: postoperative findings and progress. *The Laryngoscope*. 2001; 111(4):555–562. [PubMed: 11359119]
- Sininger, YS. Identification of auditory neuropathy in infants and children; Paper presented at the Seminars in Hearing; 2002.
- Sparrow, S.; Cicchetti, D.; Balla, D. survey interview form/caregiver rating form. Livonia, MN: Pearson Assessments; 2005. Vineland adaptive behavior scales: (Vineland II).
- Starr A, Picton TW, Sininger Y, Hood LJ, Berlin CI. Auditory neuropathy. *Brain*. 1996; 119(3):741–753. [PubMed: 8673487]

- Teagle HF, Roush PA, Woodard JS, Hatch DR, Zdanski CJ, Buss E, Buchman CA. Cochlear implantation in children with auditory neuropathy spectrum disorder. *Ear and Hearing*. 2010; 31(3):325–335. [PubMed: 20090530]
- Tomblin JB, Oleson JJ, Ambrose SE, Walker E, Moeller MP. The Influence of Hearing Aids on the Speech and Language Development of Children with Hearing Loss. *JAMA Otolaryngology - Head & Neck Surgery*. 2014; 140:403–409. [PubMed: 24700303]
- Tomblin JB, Harrison M, Ambrose SE, Walker EA, Oleson JJ, Moeller MP. Language outcomes in young children with mild to severe hearing loss. *Ear and Hearing*. accepted.
- Trautwein PG, Sininger YS, Nelson R. Cochlear implantation of auditory neuropathy. *Journal of the American Academy of Audiology*. 2000; 11(6):309–315. [PubMed: 10858002]
- Van Naarden K, Decouflé P, Caldwell K. Prevalence and characteristics of children with serious hearing impairment in metropolitan Atlanta, 1991–1993. *Pediatrics*. 1999; 103(3):570–575. [PubMed: 10049958]
- Vermeire K, Brokx JP, Van de Heyning PH, Cochet E, Carpentier H. Bilateral cochlear implantation in children. *International Journal of Pediatric Otorhinolaryngology*. 2003; 67(1):67–70. [PubMed: 12560152]
- Vlastarakos PV, Nikolopoulos TP, Tavoulari E, Papacharalambous G, Korres S. Auditory neuropathy: endocochlear lesion or temporal processing impairment? Implications for diagnosis and management. *International Journal of Pediatric Otorhinolaryngology*. 2008; 72(8):1135–1150. [PubMed: 18502518]
- Walker EA, Spratford M, Moeller MP, Oleson J, Ou H, Roush P, Jacobs S. Predictors of hearing aid use time in children with mild-to-severe hearing loss. *Language, Speech, and Hearing Services in Schools*. 2013; 44(1):73–88.
- Widen JE, Folsom RC, Cone-Wesson B, Carty L, Dunnell JJ, Koebse K, Trouba S. Identification of neonatal hearing impairment: hearing status at 8 to 12 months corrected age using a visual reinforcement audiometry protocol. *Ear and Hearing*. 2000; 21(5):471–487. [PubMed: 11059705]
- Zeng F-G, Kong Y-Y, Michalewski HJ, Starr A. Perceptual consequences of disrupted auditory nerve activity. *Journal of Neurophysiology*. 2005; 93(6):3050–3063. [PubMed: 15615831]

Figure 1.

Aided SII values for average speech input levels (65 dB SPL), plotted as a function of BEPTA. The filled circles are children with ANSD and the open circles are children with SNHL. The normative SII range is plotted as solid (mean) and dashed (upper and lower 95% confidence intervals) lines (Bagatto, 2011).

Figure 2.

Individual data points for children with ANSD (filled circles) and SNHL (open circles) on standardized speech production (*Goldman-Fristoe Test of Articulation-2*), language (*Comprehensive Assessment of Spoken Language and Peabody Picture Vocabulary Test-4*), and adaptive behavior (*Vineland Adaptive Behavior Scales-2*) measures. The hatched region represents the average range for the normative samples of these measures.

Figure 3.

LittleEARS raw scores, plotted as a function of chronological, uncorrected age. The filled circles are children with ANSD and the open circles are children with SNHL. The normative LittleEARS range is plotted as solid (mean) and dashed (upper and lower 95% confidence intervals) lines (Coninx et al, 2009).

Figure 4. PBK percent correct scores in quiet, plotted as a function of BEPTA. The dashed line represents the minimum expected score for ears with SNHL (Yellin et al, 1989).

Table 1

Data related to birth history and risk factors for children in the ANSD and SNHL groups.

	ANSD (n = 12)		SNHL (n = 22)	
Birth history¹	Mean	SD	Mean	SD
Pregnancy length (weeks)	30.54	6.47	37.61	3.50
Weight at birth (pounds)	3.54	2.31	7.12	1.95
	n	%	n	%
Prematurity ²	9	82%	4	19%
NICU stay > 5 days	9	82%	3	14%
Risk Factors: Complications during pregnancy, delivery, or after birth				
	n	%	n	%
Eclampsia	2	18%	2	10%
Fetal distress	3	27%	2	10%
Premature rupture of membranes	0	0	1	5%
Preterm labor	6	55%	3	14%
Perinatal hypoxia	1	9%	1	10%
Meconium aspiration	0	0	0	0
Jaundice	8	73%	8	38%
Risk factors: Interventions during delivery or after birth				
ECMO	1	9%	0	0
Assisted ventilation	7	64%	3	14%
High O ₂ concentrations	5	45%	1	5%
Aminoglycosides	3	27%	2	10%
Loop diuretics	2	18%	0	0
Exchange transfusion	2	18%	0	0

Note.

¹ One child with ANSD did not have birth history information available. One child with SNHL had some birth history information missing due to adoption.

² Prematurity was defined as less than 37 weeks gestation.

Table 2

Descriptive data for ages at service delivery and audiologic characteristics for ANSD/SNHL matched pairs at first visit with complete audiological data.

Test variable	ANSD (n=12)				SNHL (n=12)				p-value
	Mean	Med	SD	range	Mean	Med	SD	range	
Better Ear PTA (dB HL)	56.96	58	11.11	29– 71.25	56.88	58.13	10.74	30–71	.90
Better Ear SII	.74	.73	.12	.55– .95	.72	.69	.08	.60– .88	.58
Amount of daily HA use (parent report in hours)	11.46	12	2.11	6.71– 15	12.33	12.25	1.78	9–15	.31
Age at Service Delivery (months)									
First evaluation ^a	8.42	4	10.64	1–36	6.25	2	8.74	.5–25	.17
Confirmation ^b	8.95	5.25	10.67	1–36	7.3	2	9.80	.5–25	.31
HA fitting ^c	13.73	12	9.48	4–38	8.18	4	9.48	1.5–27	.005*
Entry into early intervention ^d	10.73	6	11.22	3–41	7.05	3.5	7.04	1–24	.25

Note:

^a ANSD group, n = 11; SNHL group = 12;

^b ANSD group, n = 10; SNHL group, n = 12;

^c ANSD group, n = 11; SNHL group, n = 12;

^d ANSD group, n = 11; SNHL group, n = 12

Table 3

Better-ear pure-tone average, better-ear aided speech intelligibility, chronological age, and standard scores for ANSD/SNHL matched pairs on CASL Syntax Construction and Pragmatic Judgment

	Better-ear PTA		Better-ear SII		Age at testing (months)		CASL Syntax Standard Scores		CASL Pragmatics Standard Scores	
	ANSD	SNHL	ANSD	SNHL	ANSD	SNHL	ANSD	SNHL	ANSD	SNHL
Matched Pair										
CID ⁴	29	30	.95	.88	47	48	113	117	102	127
RDU ³	53	51.25	.69	.71	36	36	83	87	73	88
CHI ⁶	55	53	.76	.78	71	71	72	95	98	72
EAR ⁶	60	65	.70	.74	72	71	63	116	72	95
SGU ⁴	63	61.25	No data	.41	48	48	110	81	99	99
OMA ⁴	71.25	76	.65	.63	50	52	68	84	83	89
LNK ³	71.25	71	No data	.60	34	35	71	71	73	88
STL ³	75	73	.54	.59	36	38	71	71	73	73
SLC ⁸	71	73	No data	.64	96	95	128	130	121	102
Mean	60.94	61.50	.72	.66	54.44	54.89	86.56	94.67	87.78	94.44
SD	14.30	14.78	.14	.14	21.00	20.21	23.91	21.45	18.05	17.76

Note. The letter codes for the matched pairs represent an arbitrary assigned code for the participants with ANSD. The superscript numbers represent the age (in years) at which the test visit took place.

Table 4

Better-ear pure-tone average, better-ear aided speech intelligibility, chronological age, and standard scores for ANSD/SNHL matched pairs on PPVT-4.

	Better-ear PTA		Better-ear SII		Age at testing (months)		PPVT-4 Standard Scores	
	ANSD	SNHL	ANSD	SNHL	ANSD	SNHL	ANSD	SNHL
Matched Pair								
CHI ⁵	53	53.75	.76	.79	61	61	88	97
ICT ⁷	55	51	No data	.76	82	83	79	112
EAR ⁵	61	66.25	.65	.69	61	62	61	109
SLC ⁷	70	69	.66	.73	84	83	117	86
CID ⁷	27.5	25	No data	.95	86	85	92	119
Mean	53.3	53	.69	.79	74.8	74.8	87.4	104.6
SD	15.9	17.5	.06	.10	12.7	12.2	20.4	13.1

Note. The letter codes for the matched pairs represent an arbitrary assigned code for the participants with ANSD. The superscript numbers represent the age (in years) at which the test visit took place.

Better-ear pure-tone average, better-ear aided speech intelligibility, chronological age, and standard scores for ANSD/SNHL matched pairs on GFTA-2.

Table 5

	Better-ear PTA		Better-ear SII		Age at testing (months)		GFTA-2 Standard Scores	
	ANSD	SNHL	ANSD	SNHL	ANSD	SNHL	ANSD	SNHL
Matched Pair								
CID ⁵	26	26.25	.94	.94	64	58	108	106
OMA ³	51.25	56	.90	.69	37	37	84	53
RDU ³	53	51.25	.69	.71	36	36	97	89
CHI ⁵	53	53.75	.76	.79	61	61	80	86
EAR ⁷	61	71.25	.70	.67	84	83	75	106
SGU ³	61	61	.56	.64	38	38	100	111
SLC ⁷	70	69	.66	.73	84	83	95	96
LNK ³	71.25	71	No data	.60	34	35	64	96
STL ³	75	73	.54	.59	36	38	74	69
Mean	57.9	59.2	.72	.71	52.7	52.1	86.3	90.2
SD	14.8	14.8	.14	.11	21.0	20.0	14.5	18.9

Note. The letter codes for the matched pairs represent an arbitrary assigned code for the participants with ANSD. The superscript numbers represent the age (in years) at which the test visit took place.

Table 6

Better-ear pure-tone average, better-ear aided speech intelligibility, chronological age, and standard scores for ANSD/SNHL matched pairs on the Vineland.

	Better-ear PTA		Better-ear SII		Age at testing (months)		Vineland Adaptive Behavior Composite Score	
	ANSD	SNHL	ANSD	SNHL	ANSD	SNHL	ANSD	SNHL
Matched Pair								
CID ⁴	29	30	.95	.88	47	48	97	108
OMA ³	51.25	56	.90	.69	37	37	76	67
CHI ⁴	53	53	.79	.80	49	49	102	93
MSP ²	56	59	.58	.66	24	25	86	94
EAR ⁴	58	56.25	.75	.69	54	48	79	87
STL ¹	61	61.25	.76	.69	12	11	117	106
SGU ³	61	61	.56	.64	38	38	100	95
RDU ²	51	50	.71	.68	24	24	90	102
Mean	52.5	53.3	.75	.73	35.6	35.1	91.3	98.0
SD	10.3	10.2	.14	.08	14.5	13.8	13.0	9.4

Note. The letter codes for the matched pairs represent an arbitrary assigned code for the participants with ANSD. The superscript numbers represent the age (in years) at which the test visit took place.

Table 7

Better-ear pure-tone average, better-ear aided speech intelligibility, chronological age, and standard scores for ANSD/SNHL matched pairs on the LittIEARS.

	Better-ear PTA		Better-ear SII		Age at testing (months)		LittIEARS raw score	
	ANSD	SNHL	ANSD	SNHL	ANSD	SNHL	ANSD	SNHL
Matched Pair								
MSP ¹	58	60	.55	.60	11	12	29	33
STL ¹	61	61.25	.76	.69	12	11	29	20
SGU ^{1.5}	63	63	.63	.66	20	18	30	31
DBQ ^{1.5}	64	62.5	.71	.79	21	18	23	32
Mean	61.5	61.69	.66	.69	16	14.75	27.8	29
SD	2.65	1.34	.09	.08	5.23	3.78	3.2	6.1