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Impact of the presence of auditory neuropathy spectrum disorder (ANSD) on outcomes of children at three years of age

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Abstract

Objective—To determine the influence of the presence of auditory neuropathy spectrum disorder (ANSD) on speech, language, and psycho-social development of children at three years of age.

Design—A population-based, longitudinal study was performed on outcomes of children with hearing impairment (LOCHI) in Australia. The demographic characteristics of the children were described, and their developmental outcomes were evaluated at three years of age. Performance of children with ANSD was compared with that of children without ANSD in the LOCHI study.

Study sample—There were 47 children with ANSD in the study sample.

Results—Sixty-four percent of children with ANSD have hearing sensitivity loss ranging from mild to severe degree, and the remaining have profound hearing loss. At three years, 27 children used hearing aids, 19 used cochlear implants, and one child did not use any hearing device. Thirty percent of children have disabilities in addition to hearing loss. On average, there were no significant differences in performance level between children with and without ANSD. Also, the variability of scores was not significantly different between the two groups.

Conclusions—There was no significant difference in performance levels or variability between children with and without ANSD, both for children who use hearing aids, and children who use cochlear implants.

Keywords

Auditory neuropathy spectrum disorder; children; language outcomes; psychosocial development; functional performance; hearing aids; cochlear implants; PEACH; PLS-4; PPVT; CDI; DEAP

About 1 in 10 children identified with permanent childhood hearing loss has auditory neuropathy spectrum disorder (ANSD) (Rance et al, 1999; Sininger & Oba, 2001; Tang et al, 2004; Rance, 2005; Kirkim et al, 2008; Vlastarakos et al, 2008; Sanyelbhaa Talaat et al, 2009). Knowledge about the speech and language development of children with ANSD is limited, and evidence on how best to manage children with this disorder is lacking.

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The presence of ANSD is characterized by abnormal or absent neural responses (auditory brainstem response: ABR) together with the presence of pre-neural responses (otoacoustic emissions or cochlear microphonics) (Starr et al, 1996). Through universal newborn hearing screening (UNHS) programs, infants with ANSD, like those with sensorineural hearing loss (SNHL), are detected soon after birth. Unlike the latter group, the hearing sensitivity loss of infants with ANSD cannot be estimated from ABRs. Their behavioural audiograms, when available, may range from normal to profound degrees (Sininger & Oba, 2001; Berlin et al, 2010), fluctuate (Starr et al, 1998; Marlin et al, 2010), or may be transient in nature (Madden et al, 2002; Psarommatis et al, 2006; Attias & Raveh, 2007). Audiological management of infants with ANSD is often deferred until behavioural hearing thresholds can be determined using standard audiometry, which may occur when a child is 8 – 12 months developmental age (Moore et al, 1992; Norton et al, 2000). This means that infants with significant degrees of pure-tone loss — estimated to be about 68 to 90% of ANSD cases (Rance et al, 1999; Sininger & Oba, 2001) — will experience auditory deprivation due to delays in provision of acoustic amplification or cochlear implantation.

Despite general recommendations that children diagnosed with ANSD should be treated with (1) Appropriate hearing aid (HA) amplification when reliable behavioural thresholds demonstrate a significant hearing loss (Hayes & Sininger, 2008), and (2) Cochlear implantation (CI) when poor progress in speech and language development is found despite appropriate amplification (Raveh et al, 2007; Berlin et al, 2010), there is continual controversy about how best to manage individual children with ANSD. A systematic review of the literature conducted by ASHA's National Centre for Evidence-Based Practice in Communication Disorders (NCEBPCD) (Roush et al, 2011) indicated that published data on outcomes of children with ANSD and the relative efficacy of different types of audiological treatment are scant and inconsistent. Most reports were case series reports or single case studies on children's sound detection and speech perception, with very limited data on functional ability. No data on speech production, language, and psychosocial development of three-year-old children with ANSD using hearing aids or cochlear implants had been published. Information about these early outcomes would contribute to decisions about management strategies for individual children.

In the sixteen studies on ANSD reviewed by Roush et al (2011), 91 out of the 116 subjects (78%) with audiological results had pure-tone thresholds in the severe-profound range. Not surprisingly all were considered for cochlear implantation, which is what we would expect for any child with a severe-profound hearing loss, regardless of whether it was due to ANSD. In addition, all subjects showed a considerable improvement in hearing sensitivity following cochlear implantation, which is also not surprising given the degree of their hearing loss.

It could be said that the most challenging group of children with ANSD to manage are those with pure-tone thresholds in the mild-moderate range. Subjects in this group are expected to show an improvement in their sound-detection thresholds with the provision of hearing aids in the same way as subjects with a SNHL. However, studies have shown that not all of these children improved in speech perception ability when aided (Rance et al, 1999, 2002; Berlin et al, 2010). Psychoacoustic studies have shown that children with ANSD have problems processing temporal information (Zeng et al, 1999, 2005; Rance et al, 2004). It has been argued that amplification may cause speech to sound louder but more distorted, whereas cochlear implantation may facilitate more synchronous neural firing. Several case studies on patients with ANSD who had pure-tone thresholds in the moderate range showed an improvement in speech discrimination after cochlear implantation (Berlin et al, 2008). However, other studies reported limited benefit from cochlear implantation (Zdanski et al, 2006; Bradley et al, 2008; Teagle et al, 2010; Neary & Lightfoot, 2012).

Across studies reviewed by Roush and colleagues (Roush et al, 2011), about 50% of children who received either hearing aids or cochlear implants obtained some speech perceptual benefits. Of children who used CI, about 26% did not have open-set word recognition. The wide range of performance reported in these case studies possibly reflect the differential extent of neural distortions associated with ANSD and the heterogeneity of the cases being measured. As the studies used convenience samples and unblinded assessments and did not take into account basic characteristics of participants that might affect performance, the published findings do not resolve the controversies surrounding the relative efficacy of different audiological treatment for children with ANSD. Furthermore, few studies provided a comparison between subjects with ANSD and sensorineural loss of similar degree and age at intervention. This makes it difficult to discern whether poor outcomes were due to the ANSD or were typical of the degree of pure-tone hearing loss.

The evidence-based systematic review on audiological management of ANSD conducted by ASHA (NCEBPCD) (Roush et al, 2011) concluded that published studies had methodological limitations; and that current evidence is insufficient to guide clinicians in the management of children with ANSD or to guide families in the decision-making process. The review called for “prospective, longitudinal studies on larger groups of children with detailed descriptions of participants,” to “address the efficacy of acoustic amplification and cochlear implantation in children with ANSD and the impact of this disorder on developmental outcomes” (Roush et al, 2011).

In order to meet this demand and address the shortcomings of previous studies, we measured speech, language, functional and psychosocial outcomes in a sample of three-year-old children with ANSD, drawn from the population-based cohort of the longitudinal outcomes of children with hearing impairment (LOCHI) study (Ching et al, 2013b). We aimed to (1) Describe the characteristics of a population of children with ANSD; (2) Describe their developmental outcomes; (3) Investigate whether children with ANSD perform differently from those with SNHL, separately for those who use HA, and those who use CI; and (4) Investigate whether the variability in performance for children with ANSD is different from those with SNHL, separately for those who use HA, and those who use CI.

If electrical stimulation is superior to acoustic stimulation in providing access to acoustic signals that support more synchronous neural firing (Berlin et al, 2010) the negative impact of ANSD would be greater in children with HA relative to those with CI. We hypothesized that (1) There is no difference in outcomes between children with ANSD and children with SNHL who use HA; (2) There is no difference in outcomes between children with ANSD and children with SNHL who use CI; (3) There is no difference in the effect of ANSD on outcomes between those who use HA and those who use CI; and (4) There is no difference in variability of outcomes of children with or without ANSD.

Method

Design of the study

This is a population-based cohort study.

Participants

The participants were children enrolled in the LOCH study (Ching et al, 2013b). All families of children with hearing loss born between 2002 and 2007 were invited to participate in the LOCHI study if they (1) were residing in the Australian states of New South Wales, Victoria and Queensland (excluding regional Queensland), and (2) first presented for hearing services at Australian Hearing paediatric centres before three years of age. Australian Hearing (AH) is the government-funded national service-provider for all children with

hearing loss in Australia. Recruitment was completed in 2007. The cohort of the LOCHI study comprised 451 children, of whom 47 (10%) were diagnosed with ANSD. Three of these children had ANSD in one ear, the remaining children had the disorder in both ears.

Hearing-aid fitting of children

All children enrolled in the study were fitted with hearing aids according to the AH national paediatric amplification protocol (King et al, 2005) by audiologists of the national service network. For children without ANSD, hearing thresholds were estimated from ABR measured at diagnostic hospitals, and standard behavioural audiometry was performed at hearing centres where applicable. For children with ANSD, the audiogram for fitting was estimated on the basis of the minimum response levels obtained during behavioral observation audiometry (BOA) assessments, together with measurements of cortical auditory evoked potentials (CAEP) using speech-sound stimuli developed in a previous study (Golding et al, 2009), as well as parental reports (Ching & Hill, 2007). Children who enrolled in the study prior to initial amplification were randomly assigned to being fitted with either the National Acoustic Laboratories prescription for non-linear hearing aids, version 1 (NAL), or the desired sensation level, version 4.1 (DSL) prescription (Ching et al, 2013a). Those who enrolled after initial fitting were fitted with hearing aids using the NAL prescription.

Hearing-aid fitting for all children involved the use of real-ear-to-coupler differences (RECD) to derive custom prescriptive targets, and the measurement of hearing aids in an HA2-2cc coupler to verify that targets were matched to within 5 dB at four of the five octave frequencies between 0.25 and 4 kHz.

Participant-related measures: Predictor variables

To control for baseline characteristics that potentially influence outcomes, we collected information about a range of child-, family- and intervention-related characteristics that have been reported in the literature to be associated with child outcomes. The child-related variables used for predicting outcomes included: age at first-fitting of hearing aids, age at switch-on of cochlear implants, device (hearing aids or cochlear implants), prescription used for fitting (NAL or DSL), severity of hearing loss, gender, birth weight, the presence or absence of additional disabilities, and the presence or absence of ANSD. Other family-related variables included: maternal education, socio-economic status, and communication mode used at home. In addition, intervention-related variables included: communication mode used in early education, and whether a child changed communication mode in early education during the first three years of life.

Data relating to audiological management of children were collected from the databases held at AH and other relevant intervention agencies.

Information about demographic characteristics was solicited from parents or caregivers using custom-designed questionnaires. Family socio-economic status was determined on the basis of the location of the family residence, according to the Australian census-based socio-economic indexes for areas (SEIFA) index for relative socioeconomic advantage and disadvantage; IRSAD (Pink, 2008). The index defines relative socioeconomic advantage and disadvantage in terms of “people’s access to material and social resources, and their ability to participate in society” (Pink, 2008). In this paper, the IRSAD was specified in terms of deciles, with a lower index associated with greater relative disadvantage. Maternal education was specified in terms of a three-point scale: less than or equal to 12 years of school attendance, diploma or certificate, and university qualification. Mode of communication at home and in early education was specified in terms of oral only, manual only, or a

combination of oral and manual methods. In addition, parents were asked to provide information about whether their child had a diagnosed disability in addition to hearing loss.

Outcome measures

Outcome measures included both direct assessments of children and written reports from parents or caregivers. Formal assessments included the Preschool Language Scale version 4 (Zimmerman et al, 2002) for assessing expressive and receptive language; the Peabody Picture Vocabulary Test version 4 (PPVT-4) (Dunn & Dunn, 2007) for assessing receptive vocabulary; and the Articulation and Phonology subtest of the Diagnostic Evaluation of Articulation and Phonology test (DEAP) (Dodd et al, 2002) for assessing speech production. For these measures, published normative data were used to derive z scores.

Written report tools included checklists from the Child Development Inventory (CDI) (Ireton, 2005) for soliciting information on expressive and receptive language, as well as social and motor skills development. The language comprehension, expressive language, social, self-help, gross motor, and fine motor subscale scores were reported in terms of quotients. Using published normative data, raw scores were converted into developmental ages, and quotients were calculated by dividing the child's developmental age by their chronological age, expressed as a percentage. Parents were also requested to complete the parents evaluation of aural/oral functional performance of children (PEACH), which is a measure of children's functional communicative performance in real-world situations (Ching & Hill, 2007). The PEACH also provided a measure of usage of hearing device and listening comfort in daily lives, as reported by parents. For this measure, the group mean score and standard deviation in children with normal hearing were used to derive z scores for the participants.

Procedure

A team of qualified speech pathologists administered tests directly to children in their homes or early education centres, after they turned three years of age. The mean age at evaluation was 36.8 months (interquartile: 36 – 38 months; min-max: 34 – 42 months). For children who used speech and sign to communicate, tests were administered by a qualified speech pathologist/sign language interpreter using the same communication mode as the child. The researchers who collected outcomes data were blinded to the aetiology, ANSD diagnosis, screening status, and severity of hearing loss of children. Direct assessments were video- and audio-recorded, and randomly selected samples constituting at least 10% of the total were subjected to a second, independent scoring. The inter-rater reliability was 97%. Written checklists and questionnaires were sent to parents prior to direct evaluation of children, and were collected either at or soon after the assessments.

Data analyses

The primary outcome measures were summarized in terms of means and standard deviations. Means were compared using independent sample t-tests with unequal variance, and proportions using χ^2 statistics. Effect sizes between the two samples for continuous scales were calculated by subtracting the mean of the ANSD sample from the mean of the SNHL sample and dividing the result by the standard deviation of the SNHL sample (Lydick & Epstein, 1993). An effect size of 0.2 is considered small, 0.5 moderate, and 0.8 large (Cohen, 1977).

We used multiple regression analysis to investigate the effect of ANSD, after controlling for a range of predictor variables. Factor analysis was first used to form a global outcomes score from test scores of nine measures (PLS-4 auditory comprehension score, PLS-4 expressive communication score, PPVT-4 receptive vocabulary score, DEAP consonant correct score,

DEAP vowel correct score, CDI language comprehension score, CDI expressive language score, CDI social score, and PEACH score). Multiple regression analysis was carried out with this global outcomes score as a dependent variable, and 15 predictor variables. The predictors included gender, device (hearing aids or cochlear implants), presence or absence of additional disability, presence or absence of ANSD, communication mode in early education (no intervention, oral mode, other [including oral and manual combined mode as well as manual only]), change in communication mode during educational intervention (not attending or no change, changed from oral mode to other, changed from other to oral mode), communication mode at home (oral mode or other), language used at home (English or other), maternal education (≤ 12 years of schooling, certificate or diploma, university), and hearing-aid prescription (DSL or NAL), age at first-fitting of hearing aids, age at switch-on of first cochlear implant, birth-weight, socio-economic status (expressed as IRSAD scores), and four-frequency average hearing loss in the better ear (4FA HL, average of hearing threshold levels at 0.5, 1, 2, and 4 kHz). Interaction terms of device × age at fitting, and device × 4FA HL were included. To investigate whether within each device group (HA or CI), children with ANSD perform significantly differently from those with SNHL, and whether the effect differs between device groups; an interaction term between device and ANSD was also added. To test whether children with ANSD exhibit greater variability in scores than children with SNHL, the Levene's test of equality of variances was applied to the residuals.

We used two-tailed tests for all analyses and set statistical significance at $p < 0.05$. The statistical analysis was performed using Statistica (Statsoft, 2005) and R (R Development Core Team, 2011), with the additional R packages ggplot2 (Wickham, 2010) and rms (Harrell, 2011).

Results

Characteristics of the cohort with ANSD

Table 1 summarizes demographic characteristics of children with ANSD. At three years of age, 19 children were using cochlear implants (CI) and 27 children were using hearing aids (HA). One child was overseas and the caregiver reported that the child was not using any hearing device. Thirty-five children (75%) were fitted with hearing aids before six months of age, ten (21%) between 7 and 12 months, and two (4%) between 13 and 16 months of age. Of the children who had cochlear implants, five received the implant before 12 months of age, twelve were implanted between 13 and 24 months, and two were implanted between 25 and 29 months of age.

Eighty percent ($n = 36$) of the children used an oral-only mode of communication at home and in early education. None reported using a manual-only mode of communication.

Fourteen children (30%) were reported to have at least one other disability in addition to hearing loss; which included autism spectrum disorder (n = 1), developmental delay (n = 3), motor or physical disabilities (n = 8), visual or tactile disabilities (n = 7), and cranio-facial abnormalities (n = 1). These disabilities were not mutually exclusive, with 10 of the 14 children reported to have more than one additional disability.

Thirty-six children (76.6%) had risk factors co-existing with ANSD (Table 2). Reports on computed tomography scans or magnetic resonance imaging were available for 26 children. Of these, four children had nerve defects: fused vestibulocochlear nerve unilaterally (two), and small, thin auditory nerves (one unilateral and one bilateral). Two children had enlarged vestibular aqueduct. Other medical conditions included congenital heart disease, chronic

lung disease, respiratory distress, hypoxi-ischaemic encephalopathy and convulsions, vision defect, kidney defect, sub-ependymal haemorrhage, atrial septal defect, and kernicterus.

Molecular testing of newborn blood spots was completed for 32 (68%) children (Dahl et al, 2013). Families of 15 children (32%) declined permission for testing. The results indicated that mutations were detected for *Connexin* (*n* = 5; 11%), *pendrin* (*n* = 4; 9%), *A1555G* mitochondrial (*n* = 1; 2%), and congenital cytomegalovirus (CMV) infection (*n* = 1; 2%). Twenty-one children (45%) with ANSD did not have detectable mutations or evidence of CMV infection.

Outcomes of children with ANSD

Of the 47 children with ANSD, 39 (83%) had outcomes data for at least two measures. Twenty-two of the children used hearing aids and 17 used cochlear implants. Eight children were not available for assessments, one of whom had additional disabilities. Table 3 shows comparisons between test scores of the SNHL group and the ANSD group on standardized assessment measures. On average, there were no significant differences between the two groups for any of the test measures.

Direct assessments: Speech production

Data on the DEAP test were available from 19 children, 10 of whom achieved standard scores within one standard deviation of the normative mean score.

Direct assessments: Receptive and expressive language

Of the 29 children who were assessed using the PLS-4, 12 (41.4%) achieved scores that were within ± 1SD of the normative mean for both auditory comprehension and expressive communication scales. Data on PPVT-4 were available for 23 children, and scores of 12 children (52.2%) with ANSD were within the normal range. On average, the receptive vocabulary of children with ANSD was within the normal range.

Parent report

Data on the PEACH questionnaire on functional ability were available for 26 children, 17 (65.4%) of whom had scores within the range of their normal-hearing peers. The CDI reports were available for 33 children. The number of children *n* (%) obtaining quotients within the normal range of ≥ 80 varied with different subscales: Social subscale: 12 (36.4%); Self-help subscale: 18 (54.5%); Gross motor subscale: 16 (48.5%); Fine motor subscale: 20 (60.6%); Expressive language subscale: 9 (29%); and Language comprehension subscale: 7 (22.6%).

Comparing global outcomes: SNHL vs. ANSD

The regression model investigating predictors for global outcomes score showed that presence or absence of additional disabilities, gender, maternal education, and age at cochlear implant switch-on were significant predictors of three-year outcomes at the 5% level (Ching et al, 2013b). Of the 356 children with a factor score, 250 (21 with ANSD) had hearing aids and 106 (14 with ANSD) had a cochlear implant. Figure 1 shows factor scores as a function of hearing loss (4FA HL in the better ear) separately for children with HA and CI. The scores were adjusted for continuous predictors at their average values, with communication mode in early education set to “oral”, change in communication mode in early education set to “no change”, and maternal education set to “diploma or certificate”. Open symbols depict scores of children with ANSD.

Overall, the presence of ANSD was associated with an effect size of 1.02 (95% confidence interval: − 4.73, 6.78). When separating children according to device (HA or CI) into two groups (Figure 2), the effect size of ANSD for a child with hearing aids was 3.4 (95% confidence interval: − 3.9 to 10.7), p = 0.36; and the effect size of ANSD for a child with a cochlear implant was − 2.2 (95% confidence interval: − 10.7, 6.3), p = 0.61.

To examine whether prediction errors (variability) were greater for children with ANSD compared to children with SNHL, the residuals from the regression model (Table 4) were analysed using the Levene's test of equality of variance. Results indicated that the difference among groups did not reach statistical significance (p = 0.27). Combining children with HA and children with CI, the ratio of the variance of the ANSD vs. SNHL groups was 1.08 (95% confidence interval: 0.69, 1.90). An F test for comparison of two variances indicated that the difference did not reach statistical significance (p = 0.70). For children with HA, the ratio was 0.75, with 95% confidence interval (0.42, 1.58), and p = 0.45. For children with CI, the ratio was 1.76, with 95% confidence interval (0.86, 4.71), and p = 0.12.

Discussion

We described the characteristics of a population cohort of children who were identified with ANSD via newborn hearing screening programs in Australia. The present cohort of ANSD is unique in its early age of identification and its early age of receiving audiological and educational intervention. On average, the three-year outcomes of children with ANSD were not significantly different from children with SNHL, after allowing for the effects of multiple demographic characteristics. We found that the speech, language, and psychosocial outcomes of children with ANSD did not differ significantly from those with SNHL — both for children with hearing aids, and children with cochlear implants. The variability in outcomes of children with ANSD did not differ significantly from those with SNHL — both for children with hearing aids, and children with cochlear implants.

Characteristics of the ANSD cohort

Audiological treatment—The present cohort is unique in its early age at audiological intervention, 60% (28) of whom were fitted with hearing aids before six months of age and the remaining before 16 months of age. By three years of age, 19 children (40%) used cochlear implants, including five who were implanted before 12 months of age, and the remaining children between 14 and 29 months. Previous reports on children with ANSD generally indicated amplification at older ages and low uptake of cochlear implantation (e.g. four of the 37 children (10%) in the Colorado cohort got a cochlear implant, 28 (76%) used hearing aids, and five (14%) did not use a hearing device (Uhler et al, 2012). As shown in Table 1, 64% (30 of 47 children) of children with ANSD have hearing loss ranging from mild to severe degrees. This is consistent with reports on other cohorts (Sininger & Oba, 2001). Children with moderate to severe degrees of hearing sensitivity loss rely on hearing-aid amplification to gain access to sounds. Early age of amplification provides auditory stimulation, and earlier implantation has been linked to better outcomes (Dettman et al, 2007; Ching et al, 2013b).

Communication mode—The majority of families (80%) in the present ANSD cohort chose to use an oral-only mode to communicate with their children, and to attend early education that used the same mode. The remaining children used a combination of oral and manual methods of communication, but none used a manual-only mode. The desire for oral communication has been related to the potential for broader education opportunities, better speech and language development, and opportunities for socialization (Li et al, 2003; Hyde et al, 2010; Crowe et al, submitted). The cohort in Australia is unlike the one in Colorado

(Uhler et al, 2012), where 84% (31 of the 37 families of children with bilateral ANSD) of families chose to learn and use a sign language for communication. This may be because access to sign language instruction may not be as readily available in Australia as it is in Colorado. This may also be related to the earlier age at amplification or cochlear implantation for the Australian cohort, a factor that was identified by parents to have influenced their decision on the use of speech rather than sign for communication (Crowe et al, submitted).

Language in the home—More than one in three children with ANSD was born in a family where a spoken language other than English was used. The languages included Africaan, Arabic, Caldean, Assyrian, Farsi, Japanese, and Turkish. Where a combination of oral and manual communication modes was reported, oral English was used with Auslan (Australian sign language), signed English, or keyword signing (Makaton). The use of manual methods of communication was largely driven by practical communication needs, in instances where children have difficulties that compromised their ability to use an oral-only mode of communication (Uhler et al, 2012; Crowe et al, 2013). The linguistic diversity of the cohort revealed the need for information about this disorder to be available in languages other than English to better support families.

Additional disabilities and developmental characteristics—Twenty-two children (47%) in the present cohort of 47 were reported to have disabilities in addition to hearing loss. This rate is higher than the 32% (12 of 37 children) reported for the Colorado cohort (Uhler et al, 2012); and the 25% in the general population with sensorineural hearing loss (Ching et al, 2013b).

Assessments of nonverbal cognitive ability using the Wechsler non-verbal scale of ability (Wechsler & Naglieri, 2006) were completed by qualified psychologists for 26 children at five years of age. Of the 21 who were not tested, seven were deemed unable to cope with the assessment, others were not available. Of the 26 who had nonverbal cognitive test scores, 24 were within one standard deviation of the normative mean. However, only seven of these children attained age-appropriate language ability when assessed by using the PLS-4. This is unlike the Colorado cohort (Uhler et al, 2012) in which 21 of the 25 children with normal cognitive levels attained normal language levels when assessed by using parent reports at an older age than the present cohort. Thus, factors other than cognitive ability must contribute to the difficulties in speech and language development of the Australian cohort, including but not limited to maternal education, socio-economic status, and severity of hearing loss, all of which are known to affect child outcomes (Ching et al, 2013b). Whether the Australian cohort with normal cognitive levels would catch up with their normal-hearing peers at an older age remains to be measured.

Do children with ANSD perform differently from children with SNHL?

We found that children with ANSD did not perform better or poorer than children with SNHL, on average (Table 2). The range of measures adopted in this study enabled monitoring of progress for a higher proportion of children with ANSD (80%) than was reported for other cohorts (60%) (Uhler et al, 2012).

The effect size of the presence of auditory neuropathy is small and statistically not significant at the 5% level. Within the group of children who used HA, the mean effect size is only 0.23 SD (3.4 points, 95% confidence interval is ± 7.3 points). We can conclude from the current data that the true mean effect of auditory neuropathy lies within the range − 3.9 points to + 10.7 points. Within the group of children who used CI, the mean effect size is − 0.15 SD (− 2.2 points, 95% confidence interval was ± 8.5 points). The true mean effect of

auditory neuropathy on children with cochlear implants lies within the range of − 10.7 and + 6.3 points. The effect size is effectively too small to be of clinical significance. Our finding with young children is consistent with that reported in recent studies on school-aged children, which showed that those with ANSD who used HA demonstrated speech perception and abilities and spoken language development at levels expected for children with SNHL (Rance et al, 2007; Rance & Barker, 2008, 2009) and similarly for children who used CI (Leigh et al, 2009; Rance & Barker, 2009; Breneman et al, 2012). In addition, Rance and Barker (2009) showed that school-aged children with ANSD who used HA achieved speech and language outcomes that were comparable to their counterparts who used CI. The present study extended previous findings by allowing for the effects of variations in basic characteristics between children with ANSD and those with SNHL in quantifying the impact of ANSD. The present study is the first to report on speech and language development outcomes of children with ANSD at a young age. The present evidence does not support the need to provide differential management strategies for children with ANSD, relative to children with SNHL. Our findings imply that audiological management including acoustic amplification and cochlear implantation should be provided to children with ANSD, as with children with SNHL.

Do children with ANSD display greater variability than children without ANSD?

We found that children with ANSD did not demonstrate greater variability in outcomes than children without ANSD (Table 4), contrary to clinical observations in previous studies and subjective impressions of clinicians. The higher incidence of additional disabilities and delays in audiological management for children with ANSD compared to children with SNHL may have contributed to observed variability reported in previous studies (Berlin et al, 2010). Unlike the present study, none of the previous reports controlled for the effect of demographic characteristics that are known to affect child outcomes when comparing children with and without ANSD. In this study, the standard deviations of residuals were comparable between the ANSD and the SNHL groups even though the sample size for the former was small compared to the latter group. Nonetheless, we could not rule out the possibility that a larger sample size may reveal significant differences in variability.

Implications for audiological management

The present study revealed no significant difference between children with ANSD and children with SNHL, both for those who use hearing aids and those who use cochlear implants. Even though electrical stimulation via cochlear implantation bypasses the peripheral site of lesion, the signal would still need to reach the brain via a disordered neural system. The present findings support the provision of amplification to children with ANSD as a treatment option. About 60% of children with ANSD have hearing sensitivity loss in the mild to severe range. For these children, early amplification allows early access to sounds. Our findings demonstrate that some children benefit from acoustic amplification.

The question of how to estimate hearing thresholds for early fitting is in need of further research. We have used an approach whereby cortical auditory evoked potentials elicited using speech stimuli were used to complement subjective observations of auditory behaviour to sounds for estimating hearing thresholds. After aiding, audibility was confirmed by measuring CAEP (Golding et al, 2007; Carter et al, 2010; Van Dun et al, 2012). This approach is consistent with previous studies showing a link between the presence of aided CAEP at normal latencies and speech perceptual and functional benefits with amplification (Rance et al, 2002; Sharma et al, 2005, 2011; Golding et al, 2007). Absence of aided CAEP despite hearing-aid adjustment would indicate that amplification might not be an effective treatment option. In these cases the child may benefit from additional non-auditory communication strategies such as cued speech or sign language. Referral for cochlear

implantation should also be considered even if the sensitivity loss may not be profound. The efficacy of using CAEP and parental reports to guide early amplification or cochlear implantation remains to be confirmed in future research.

Anecdotal observations suggest that some children with ANSD may have a restricted dynamic range for amplification, compared to children with SNHL. This could occur because of a deficient number of low-spontaneous rate, high-threshold auditory nerve fibres functioning (for whatever reason) for some individuals with ANSD. There is also indication in previous reports that some children with ANSD may require slower stimulation rate in their cochlear implants, relative to children with SNHL (Peterson et al, 2003). Whether the optimal range of auditory stimulation for children with ANSD may differ from that of children with SNHL remains to be investigated in future research.

Limitations

The current findings apply only to children at three years of age, and cannot be generalized to other age groups. The longitudinal design of the LOCHI study means that the participants will be followed through to early adulthood. Evaluations at an older age will allow the investigation of other significant outcomes, including speech perception, literacy and academic attainment. Future follow-through of the cohort will also enable quantification of growth trajectories for children with ANSD and those with SNHL. As radiological examination of children diagnosed with ANSD are not routine clinical practice in Australia, the incidence of auditory nerve deficits in this population is not known.

Summary and Conclusion

The strengths of the present study include its population base, its cohort of children diagnosed with ANSD through newborn hearing screening programs, its prospective measurement of a range of outcomes including language, speech and functional performance, its inclusion of standardized tests administered directly to participants as well as parent reports, its assessment of early developmental outcomes at three years of age; and its use of blinded assessments. The present study also included comprehensive information about hearing-aid fitting and hearing threshold levels, thereby allowing for a more accurate estimation of the effects of amplification characteristics on developmental outcomes.

In summary, the present findings are:

1. There was no significant difference in outcomes between children with ANSD and children with SNHL for a given hearing loss, after allowing for the effects of demographic characteristics.
2. There was no significant difference in the effect of the presence of ANSD between those who use HA and those who use CI.
3. Children’s development of language and functional performance, by three years of age, is susceptible to influences of social disadvantage and maternal educational level. On average, performance was slightly poorer than 1 SD below normative mean.

Our current findings cannot be generalized to children at other age groups. It remains to be seen whether the presence of ANSD has differential impacts on children’s language development and speech perception at an older age. Data collection for participants at five years is currently under way, and we have also begun testing at nine years of age using additional measures of literacy and related skills.

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Abbreviations

4FA	Four-frequency average, across 0.5, 1, 2, and 4 kHz
ABR	Auditory brainstem responses
ANSD	Auditory neuropathy spectrum disorder
ASHA	American Speech-Language-Hearing Association
CAEP	Cortical auditory evoked potentials
CDI	Child development inventory
CI	Cochlear implants
DEAP	Diagnostic evaluation of articulation and phonology
DSL	Desired sensation level prescription
HA	Hearing aids
HTL	Hearing threshold level LOCHI Longitudinal outcomes of children with hearing impairment study
NAL	National Acoustic Laboratories
NAL-NL1	National Acoustic Laboratories' prescription for non-linear hearing aids, version 1
NCEBPCD	National Centre for Evidence-Based Practice in Communication Disorders
PEACH	Parents' evaluation of aural/oral performance of children
PLS-4	Preschool language scale, 4 th ed.
PPVT	Peabody picture vocabulary test
SNHL	Sensorineural hearing loss
UNHS	Universal newborn hearing screening

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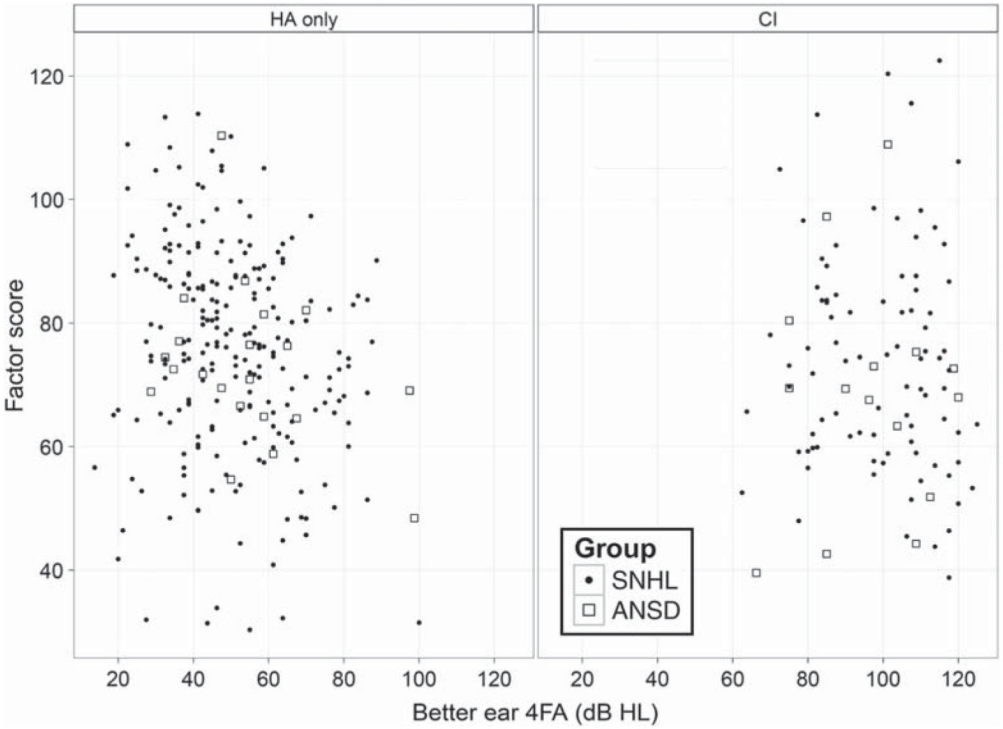

Figure 1.
Global outcomes factor scores as a function of hearing loss (defined as average of hearing thresholds at octave frequencies between 0.5 and 4 kHz in the better ear, 4FA HL), separately for children with HA and CI. Filled symbols depict scores for individual children with sensorineural hearing loss (SNHL), and open symbols depict scores for children with auditory neuropathy spectrum disorder (ANSD).

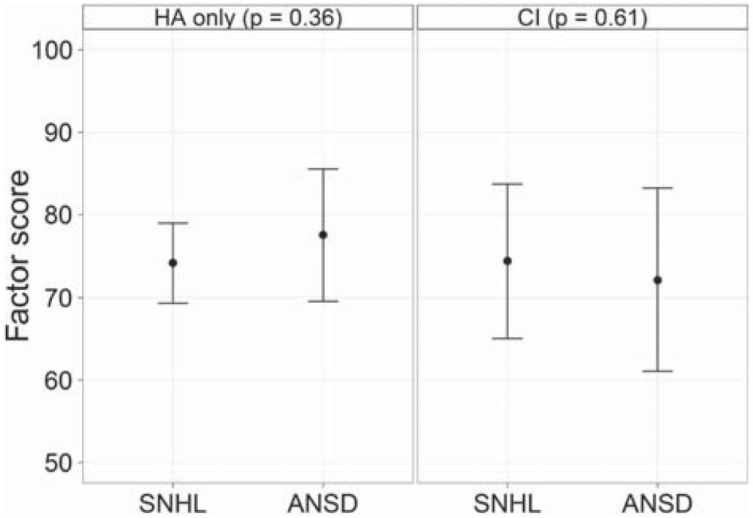

Figure 2.
Mean factor scores (filled circles) and 95% confidence intervals for children with hearing aids (HA) or cochlear implants (CI); separately for children with sensorineural hearing loss (SNHL) and children with auditory neuropathy spectrum disorder (ANSD).

Table 1

Demographic characteristics of participants with ANSD.

	n (%)	*Mean (SD)*	*Interquartile range*
Gender			
Male	28 (60)		
Female	19 (40)		
Age at diagnosis (months)		3.3 (2.2)	1.6 to 4.3
Age at hearing aid fitting (months)		6.2 (3.5)	4.0 to 7.5
Age at cochlear implantation (months)		18.2 (6.6)	14.3 to 20.3
Birthweight (gm)		1842.8 (1139.6)	877.0 to 2700.0
Device			
Unilateral cochlear implant	1 (2)		
Cochlear implant and hearing aid in contralateral ear	9 (19)		
Bilateral cochlear implants	9 (19)		
Unilateral bone conductor	1 (2)		
Bilateral hearing aids	26 (55)		
None	1 (2)		
Additional disabilities			
None	33 (70)		
≥ 1 additional disability	14 (30)		
Screened at birth			
Yes	46 (98)		
No	1 (2)		
Severity of hearing loss at outcome[#]			
Mild (20–40 dB HL)	6 (13)		
Moderate (41–60 dB HL)	18 (38)		
Severe (61–80 dB HL)	6 (13)		
Profound (> 80 dB HL)	17 (36)		
Communication mode at home and in early education[†]			
Aural/oral only	36 (80)		
Oral and sign	9 (20)		
Sign only	0 (0)		
Language used at home[*]			
English	28 (64)		
Other	16 (36)		
Maternal education			
University	15 (33)		
Diploma or certificate	14 (31)		
School	16 (35)		
Socio-economic status (Decile)		6.8 (2.9)	5.0 to 10.0
Sign only	0 (0)		

[#] Averaged thresholds at 0.5, 1, 2, and 4 kHz in the better ear.

[†] Missing data from two participants.

[*] Missing data from three participants.

Table 2

Risk factors reported for children with ANSD.

Risk factors	*n*	*%*
Prematurity*	33	70.2
Mechanical ventilation	18	38.3
Jaundice	14	29.8
Exposure to ototoxic drugs	8	17.2
Exchange transfusion	7	14.9
Family history of hearing loss	3	6.4
Birth trauma	2	4.3
Cerebral palsy	2	4.3
Gentamycin exposure	2	4.3
Sepsis	2	4.3

* Defined as < 37 gestation weeks at birth.

Table 3

Comparison of ANSD group with SNHL group for standardized outcome measures. Descriptive characteristics of children with sensorineural hearing loss (SNHL), and children with auditory neuropathy spectrum disorder (ANSD). Means ± SD, effect size, and p value for test of significance between groups are shown.

	SNL			*ANSD*			*Effect size (SD)*	*P*
	n	***mean***	***SD***	***n***	***mean***	***SD***		
PLS-AC	281	78.6	24.6	29	75.9	35.9	0.1	0.573
PLS-EC	281	85	19.4	29	81.8	17.8	0.2	0.388
PPVT	203	87.5	16.7	23	88.0	16.4	0.0	0.895
DEAP_C	185	6.1	2.6	19	6.1	2.5	0.0	0.978
DEAP_V	185	6.6	2.69	19	5.9	2.1	0.2	0.336
CSQ	269	87.5	36.3	33	77.7	34.3	0.3	0.140
CSHQ	269	89.6	32.6	33	82.4	38.9	0.2	0.247
CGMQ	269	86.1	35.5	33	75.9	34.9	0.3	0.122
CFMQ	269	93.8	28.3	33	86.1	33.0	0.3	0.149
CELQ	253	76.9	31.1	31	67.0	25.0	0.3	0.919
CLCQ	253	73.2	28.2	31	64.4	26.3	0.3	0.099
PEACH	226	71.1	21.0	26	70.5	23.2	0.0	0.889

Table 4

The standard deviation of residuals for each of four groups: Children using hearing aids with sensorineural hearing loss (HA: SNHL) or auditory neuropathy spectrum disorder (HA: ANSD); and children using cochlear implants with sensorineural hearing loss (CI: SNHL) or auditory neuropathy spectrum disorder (CI: ANSD). The average of the residuals was zero for each group.

Group	*n*	*Standard deviation of residuals*
HA		
SNHL	229	13.1
ANSD	21	11.3
CI		
SNHL	92	12.6
ANSD	14	16.7