Characteristics and outcomes of children with Auditory Neuropathy Spectrum Disorder

Caractéristiques et évolution des enfants ayant un trouble du spectre de neuropathie auditive

KEY WORDS

AUDITORY NEUROPATHY SPECTRUM DISORDER

DIAGNOSIS

MANAGEMENT

HEARING AIDS

COCHLEAR IMPLANTS

REHABILITATION

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Abstract

Background. Auditory neuropathy spectrum disorder (ANSD) has been reported in up to 10% of all children with hearing loss. It is widely accepted that it can be difficult to identify and manage children with ANSD through typical clinical procedures, in comparison to children with sensorineural hearing loss (SNHL). An important limitation in managing children with ANSD is that behavioral pure-tone audiograms are less predictive of a child's intervention needs with respect to amplification and early rehabilitation. The objective of this study was to characterize the clinical profiles and audiological management of children with ANSD followed through a provincial universal newborn hearing screening (UNHS) program in one region of Canada from 2003-2013.

Methods. Population-based data were collected at the Children's Hospital of Eastern Ontario (CHEO), Ottawa, Canada between 2003 and 2013 for all children who were diagnosed with ANSD. Detailed characteristics were extracted including screening status, etiology, risk factors, and severity of hearing loss. Diagnostic, amplification, and intervention outcomes were also documented.

Results. Analyses of 418 children indicated that 22 (5.3%) children were identified with ANSD and more than 30% had other disabilities. Children were identified at a median age of 7.5 months (IQR: 4.7, 22.5), fitted with amplification at a median age of 17.3 months (IQR: 12.1, 26.6) and 12 underwent cochlear implantation at a median age 26.4 months (IQR: 18.0, 47.3).

Conclusion. More than 85.0% of children with ANSD are currently fitted with hearing aids or cochlear implants and have achieved some degree of open-set word recognition and communication development.

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Contexte : Les troubles du spectre de neuropathie auditive (TSNA) ont été rapportés chez des enfants ayant une perte auditive, avec une prévalence pouvant s'élever jusqu'à 10 %. Il est largement reconnu que l'identification et la prise en charge effectuées auprès des enfants ayant un TSNA peuvent être difficiles avec les procédures cliniques courantes, lorsque comparées aux enfants ayant une perte auditive neurosensorielle. Une limitation importante de la prise en charge des enfants ayant un TSNA est le caractère prédictif diminué des audiogrammes tonals pour les besoins d'intervention de ces enfants en ce qui concerne l'amplification et l'intervention précoce. L'objectif de cette étude était de décrire les profils cliniques des enfants avec un TSNA, ainsi que leur prise en charge en audiologie, suite à l'application du programme provincial de dépistage universel de l'audition chez les nouveau-nés dans une région du Canada, entre 2003 et 2013.

Méthodologie : Les données démographiques ont été recueillies au Centre hospitalier pour enfants de l'est de l'Ontario, Ottawa (Canada), et ce, pour tous les enfants diagnostiqués avec un TSNA entre 2003 et 2013. Des caractéristiques détaillées ont été extraites, ce qui inclut le statut de dépistage, l'étiologie, les facteurs de risque et la sévérité de la perte auditive. Le diagnostic, l'amplification et les résultats à l'intervention ont également été documentés.

Résultats : Les analyses effectuées auprès de 418 enfants ont montré que 22 des enfants (5,3 %) ont été identifiés avec un TSNA et plus de 30 % avaient d'autres difficultés. Les enfants ont été identifiés à un âge médian de 7,5 mois (EI : 4,7 – 22,5), ont été équipés avec une amplification à un âge médian de 17,3 mois (EI : 12,1- 26,6) et 12 ont reçu un implant cochléaire à un âge médian de 26,4 mois (EI : 18,0 - 47,3).

Conclusion : Plus de 85,0 % des enfants ayant un TSNA sont actuellement équipés d'appareils auditifs ou d'implants cochléaires, ont atteint un certain degré d'identification de mots en choix ouvert et ont développé un certain niveau de communication.

Introduction

Auditory Neuropathy Spectrum Disorder (ANSD) is used to describe a complex hearing disorder that involves normal cochlear function but abnormal auditory nerve function. There is a change in neural synchrony, characterized by auditory behaviors in which the function of outer hair cells is preserved, while the afferent neural transmission is altered. Sound processing in patients with ANSD is highly variable, and the relationship between hearing sensitivity and the ability to process speech cannot be predicted based on the level of residual hearing (Hood, 2011).

ANSD is a hearing disorder, which presents distinct difficulties in speech understanding, particularly in competing noise. This audiological finding suggests disruption in processing time due to the lack of synchrony of the auditory pathways from the cochlea to the auditory cortex (Hayes, 2008; Sininger & Oba, 2001). Difficulties with the resolution of temporal processing (Rance, 2005) can have an impact on speech understanding and consequently on the development of oral language and communication. The term ANSD was adopted because there are a wide range of clinical manifestations, prognoses, and underlying etiologies associated with the disorder (Feirn, Sutton, Parker, Sirimanna, & Lightfoot, 2013).

Various studies have shown prevalence estimates ranging from 1.0% to 10.0% of children identified with permanent childhood hearing loss (Kirkim, Serbetcioglu, Erdag & Ceryan, 2008; Rance, 2005; Sininger and Oba, 2001) and 10.0% to 15.0% in schools for the deaf (Berlin et al, 2010). In population hearing screening studies, a prevalence of 24.1% to 40.0% has been reported in infants with hearing loss from the neonatal intensive care unit (NICU) (Berg, Spitzer, Towers, Bartosiewicz, & Diamond, 2005; Rea & Gibson, 2003).

Children with ANSD represent a subgroup of patients with hearing impairment with different pathologies most commonly associated with risk indicators for hearing loss related to admission to the NICU (Dowley et al., 2009). Children with histories of neonatal problems such as prematurity, low birth weight, anoxia, and hyperbilirubinemia, are at a greater risk for ANSD (Madden, Rutter, Hilbert, Greinwald, & Choo, 2002; Teagle at al., 2010). However, ANSD has also been reported in children in the absence of any clear risk factors (Sininger, 2002; Uhler, Heringer, Thompson, & Yoshinaga-Itano, 2012).

Children with sensorineural hearing loss (SNHL) or ANSD experience early deprivation of sensory input to the cortex and demonstrate changes in neuroplasticity and development upon introduction of auditory stimulation. Research regarding central auditory maturation suggests that plasticity can often be maximized via amplification and/or electrical stimulation to produce positive clinical outcomes in these patient populations (Kral, Hartmann, Tillein, Heid, & Klinke, 2001; Sharma, Dorman, & Spahr, 2002). For children with ANSD, auditory brainstem response (ABR) thresholds may differ significantly from behavioral hearing thresholds, therefore, hearing aid recommendations and adjustments should be based on behavioral audiological assessment results as soon as possible (Hayes, 2008), combined with careful observation of the child's hearing responses in everyday situations (Feirn et al., 2013). Cochlear implant (CI) surgery is also recommended for ANSD in children who are not making satisfactory auditory progress with conventional amplification; however, there is uncertainty about outcomes in cases of cochlear nerve deficiency (Breneman, Gifford, & DeJong, 2012; Buchman et al., 2006). Furthermore, it has been suggested that the site of lesion and the characteristics of pre and postsynaptic forms of ANSD may influence speech perception outcomes (Dimitrijevic et al., 2011). There is some evidence to suggest that electrophysiological tests such as auditory cortical measures might help differentiate between pre- and post-synaptic disorders and provide a better understanding of the differences in speech understanding in individuals with ANSD (Dimitrijevic et al., 2011; Santarelli, 2010).

There is considerable heterogeneity with respect to the severity of clinical manifestations and the benefit from hearing technology and different aural rehabilitation options in patients with ANSD (Pelosi et al., 2013; Roush, Frymark, Venediktov, & Wang, 2011). Given the prevalence and early diagnosis of ANSD in children, it is important to achieve a better understanding of management issues and potential outcomes for these children. The objective of this study was to characterize the clinical profiles and the audiological management and communication development outcomes of children with ANSD followed through a provincial universal newborn hearing screening (UNHS) program in one region of Canada from 2003-2013.

Methods

Population-based data were collected at the Children's Hospital of Eastern Ontario (CHEO), Ottawa, Canada between 2003 and 2013 for children diagnosed with ANSD. The hospital is the diagnostic centre for the region's provincially mandated UNHS program established in 2002. Children underwent newborn hearing screening and audiologic assessment following well-established provincial protocols. Screening protocols established by the province

involved a two-stage screen for well-babies where the initial screen typically carried out in hospital involved Automated Otoacoustic Emissions screening (AOAE) followed by Automated Auditory Brainstem Response screening (AABR) when there is a refer result from the AOAE. However, babies deemed to be at risk for hearing loss, such as those in the NICU were screened with AABR and then referred for audiological assessment when a refer result was obtained (Hyde, 2005). Consistent with provincial protocols, infants with known risk indicators for hearing loss were placed on a surveillance list and followed to age 30 months. The audiological assessment consisted of a comprehensive test battery with well-defined protocols for neonates and young children and included frequency-specific ABR, a click ABR sub-protocol when ANSD was suspected, OAEs, and immittance testing (Hyde, 2005). Children identified with permanent hearing loss were referred to otolaryngology for further medical evaluation.

All children identified with permanent hearing loss received audiological management services including amplification, if indicated, and auditory rehabilitation through the hospital. Children with ANSD were managed similarly to children with SNHL and were initially fitted with hearing aids once a hearing disorder was confirmed. Children with ANSD who derived little benefit from amplification, that is, they showed limited progress in auditory and communicative functioning, were considered for cochlear implants and underwent a comprehensive candidacy assessment. In the clinical program, children with ANSD are considered for cochlear implants even if their audiometric thresholds are outside the typical range of cochlear implant candidacy of severe to profound hearing loss.

Clinical audiological data related to age of diagnosis, etiology, risk indicators, and other clinical characteristics and hearing technology information were collected prospectively as part of a database on all children identified since the implementation of UNHS in 2002. A total of 22 patients included in this database met the diagnostic criteria based on well-defined clinical profiles of ANSD (Feirn et al., 2013; Northern, 2008). Detailed audiological information (e.g. audiograms) and communication outcomes were extracted retrospectively through chart reviews, specifically for this study. This study was approved by the institutional review boards of the CHEO and University of Ottawa.

Clinical Assessment Procedures. Typical clinical protocols for these children were followed and included regular audiological assessments of speech perception and communication development testing. Because speech perception abilities are age dependent and influenced by vocabulary and language levels, tests were selected clinically, based on the child's developmental level. Formal assessment tools are listed below and included speech perception measures, parent questionnaires, and language tests:

Speech perception measures

- Phonetically Balanced Kindergarten-PBK (Haskins, 1949) – open-set monosyllabic word test.
- Hearing in Noise Test for Children-HINT-C (Nilsson, Soli, & Gelnett, 1996) – open-set sentence test.
- Early Speech Perception Test-ESP (Moog & Geers, 1990) – closed-set test; child points to picture from a set of 12 pictures.

Parent auditory questionnaires

- Infant Toddler Meaningful Auditory Integration Scale-IT-MAIS (Zimmerman-Phillips, Robbins, & Osberger, 2001) – a 10-item questionnaire administered in interview format to parents to probe the child's auditory function in his/her everyday environment in three areas: 1) vocalization behavior, 2) alerting to sound, and 3) deriving meaning from sound.
- Parents' Evaluation of Aural/Oral Performance of Children-PEACH (Ching & Hill, 2007) – a parent questionnaire that measures functional communication in real-world environments.

Language Measures

- Peabody Picture Vocabulary Test-PPVT (Dunn & Dunn, 2007) – a measure of receptive vocabulary.
- Preschool Language Scale-PLS-4, (Zimmerman, Steiner, & Pond, 2002) – a measure of both auditory comprehension (receptive language) and expressive communication.

Results

Figure 1 shows the study selection process for the children with ANSD. A total of 418 children were identified with permanent hearing loss from 2003 to 2013, 22 (5.3%) of whom presented with ANSD.

Clinical profile of children with ANSD. Table 1 shows details of the clinical characteristics for the children with ANSD. The majority (90.9%) underwent neonatal hearing screening and 15 (75.0%) of these patients received a 'refer' result while the remaining 5 (25.0%) had a 'pass' status. The sample was predominantly male (77.3%). As shown, more than two-thirds (68.2%) of the children were diagnosed with

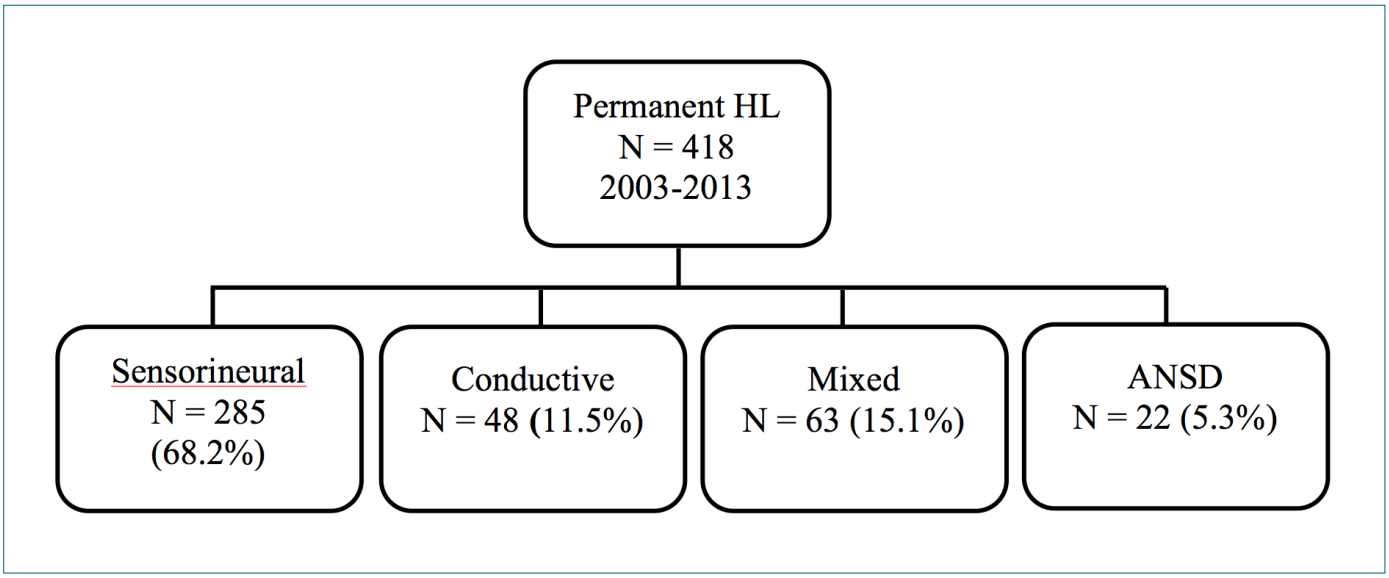

Figure 1. election of study participants
HL: hearing loss; ANSD: auditory neuropathy spectrum disorder

congenital or early onset (< 6 months) ANSD. More than 60% showed at least one risk factor, as defined by the Joint Committee on Infant Hearing (JCIH, 2007). Notably, 86.7% of these children were NICU graduates and the gestational age was less than 36 weeks for 54.5% of children. Table 1 shows the severity of hearing loss at diagnosis, which was calculated based on the three-frequency (500, 1000, 2000 Hz) pure tone average (PTA). Nineteen (86.4%) children showed moderate or greater hearing loss (>40dB HL) with 59.1% of them having severe or profound hearing levels. All but one child had bilateral ANSD. The majority of the children (86.4%) had symmetrical hearing loss (defined as <20dB HL difference between thresholds in each ear). Some children experienced fluctuating hearing loss (5/22) or deterioration in hearing thresholds over time (6/22).

Hearing technology management. The median age of diagnosis of ANSD for the 22 children was 7.5 months (Interquartile range [IQR]: 4.7, 22.5). Nineteen children (86.4%) were initially fitted with hearing aids and one patient was fit with a Frequency Modulation (FM) system for a trial period. Two children were not fitted with hearing aids due to complex medical issues and developmental delays. Initial fitting of amplification occurred at a median age of 17.3 months (IQR: 12.1, 26.6). The status of current hearing technology for the 22 children is shown in Figure 2. Seven children with bilateral ANSD who were fitted with hearing aids are currently using them. Although two of these children had audiometric thresholds consistent with profound hearing loss, they achieved open-set monosyllabic word scores of 56% and 84% (Table 2), placing them outside the typical criteria for cochlear implants. One child with unilateral profound hearing loss discontinued using amplification (FM system).

More than half of the group underwent CI surgery (n=12) at a median age of 26.4 months (IQR: 18.0, 47.3). Six of these children received cochlear implants within 12 months of hearing loss identification, while three showed deterioration in audiometric levels over time and the remaining three likely experienced delays in cochlear implantation associated with other medical/developmental issues.

Auditory and communication development. Table 2 shows the best score achieved on clinical speech perception evaluations for the seven children with hearing aids. Age-appropriate speech perception testing was performed preoperatively and repeated at multiple intervals postoperatively. The evaluations reported, based on best score in the medical chart, were completed between 11 months and 60 months of amplification use. As noted previously, outcome measures included direct assessments of children and questionnaires administered to parents when children were unable to complete formal speech perception tests. As shown, five of seven were able to complete open set tests (PBK or HINT) and one of seven completed the IT-MAIS. One child using hearing aids could not complete formal speech perception tests, therefore only aided thresholds were available. As detailed in Table 2, the variability in children's auditory abilities is reflected in the scores, which ranged from 80% on the IT-MAIS to scores between 50% and 96% on the open-set PBK word test.

Table 1. Clinical characteristics of 22 children

Characteristic		n (%)
Sex		
	Male	17 (77.3)
	Female	5 (22.7)
Screening outcome		
Not screened		2 (9.1)
Screened		20 (90.9)
	Refer	15 (75.0)
	Pass	5 (25.0)
Gestational status		
	Premature	12 (54.5)
	Full-term	10 (45.5)
Onset of hearing loss		
	Congenital	7 (31.8)
	Early	8 (36.4)
	Late	7 (31.8)
Risk indicators		
No risks		7 (31.8)
Risk		15 (68.2)
	NICU	13 (59.2)
	Family risk	1 (4.5)
	Syndromes	1 (4.5)
Degree HL - PTA (better ear)		
	Within normal limits	1 (4.5)
	Mild	2 (9.1)
	Moderate	2 (9.1)
	Moderate-severe	4 (18.2)
	Severe	2 (9.1)
	Profound	11 (50.0)
Hearing loss profile		
	Bilateral	21 (95.5)
	Unilateral	1 (4.5)
	Stable	11 (50.0)
	Fluctuation/ inconsistent	5 (22.7)
	Deterioration ≥20dB	6 (27.3)
	Symmetric	19 (86.4)
	Asymmetric	3 (13.6)

NICU: neonatal intensive care unit; PTA: pure-tone average; HL: hearing loss

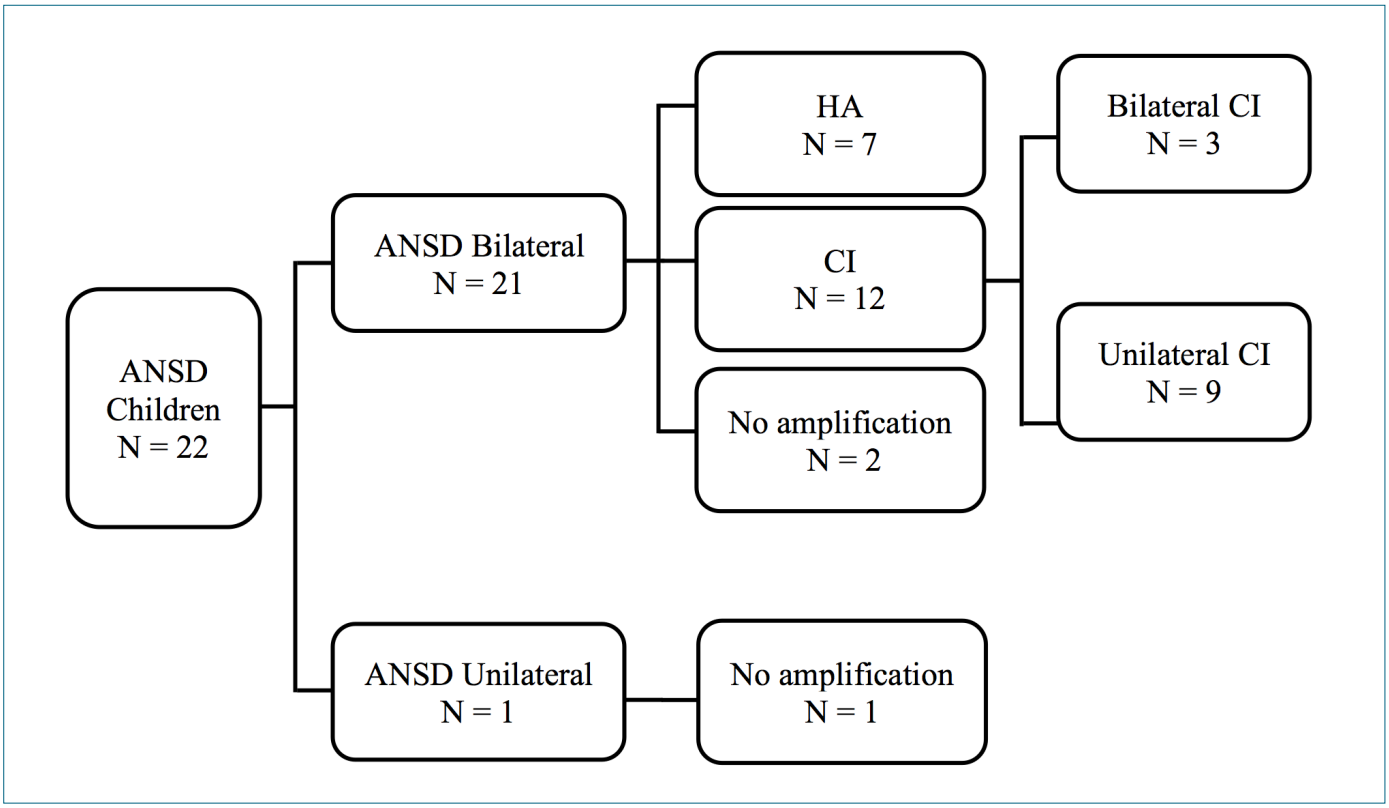

Figure 2. Hearing technology management for children with ANSD
ANSD: auditory neuropathy spectrum disorder; CI: cochlear implant HA: hearing aid

Table 2. Speech perception and audiological outcomes for hearing aid group

Child number	PTA unaided (R/L, dB)	Duration HA use (months)	Last PTA aided (R/L, dB)	Test	Best score (%)
1	40/60	55	25/25	PBK w	96
4	35/45	11	15/15	PBK w	92
16	95/NR	NA	CNT	PBK w	50
22	95/95	84	CNT	PBK w	84
19	40/45	60	20/20	HINT	88
9	NR/NR	96	30/30	IT-MAIS	80
10	75/NR	NA	50/30	CNT	CNT

PTA: threshold at 0.5, 1 and 2kHz; HA: hearing aid; dB: decibel; PKA: Phonetically-Balanced Kindergaten (PBK words); HINT: Hearing in Noise Test; IT-MAIS: Infant-Toddler Meaningful Auditory Integration Scale; CNT: could not test; R: right; L: left; NR: No response; NA: not available

Table 3. Speech perception outcomes for cochlear implant group

Child number	PTA unaided[1] (R/L, dB)	Age at CI	Duration of CI use (months)	Bilateral CI / Unilateral CI	Test	Best score[2] (%)
6	113/113	25m	36m	UCI	HINT	100
5	100/102	11m	48m	UCI	HINT	98
20	110/102	31m	3m	UCI	HINT	98
8	92/87	55m	34m	UCI	PBK w	92
14	85/80	51m	3m	BCI	PBK w	96
15	75/78	15m	36m	UCI	PBK w	88
18	95/88	18m	48m	BCI	PBK w	88
11	95/95	54m	41m	BCI	ESP	100
12	87/90	19m	17m	UCI	ESP	88
2	120/120	11m	1m	UCI	PEACH	100
7	80/110	29m	7m	UCI	PEACH	100
17	115/110	18m	NA	UCI	CNT	CNT

PTA: threshold at 0.5, 1 and 2kHz; CI: cochlear implant; dB: decibel; PKA: Phonetically-Balanced Kindergaten (PBK words); HINT: Hearing in Noise Test; IT-MAIS: Infant-Toddler Meaningful Auditory Integration Scale; CNT: could not test; R: right; L: left; NR: No response; NA: not available

Table 3 summarizes auditory performance for children with CI and shows the best score achieved on speech understanding (direct measurement or parent questionnaire). Cochlear implant use ranged from 1 to 48 months. These children obtained detection levels after implantation between 15 and 30 dB HL. Speech perception outcomes varied greatly from one child to another. As shown, 7 of 12 children were able to complete open-set speech perception tests (HINT or PBK word), achieving scores ranging from 88% to 100%. Two children (#11 and #12) could only complete closed-set testing and two others (#2 and #7) had only parent questionnaire scores. Child #2 was too young to complete speech perception testing (< 1 year of age) and child #7 was unable to complete testing due to severity of disabilities.

Communication development outcomes are presented in Table 4 for all children. As indicated previously, children had received various speech-language assessments depending on their level of functioning, therefore scores were summarized to reflect the child's overall functioning. Test results were not available for seven children because they received rehabilitation in other centers. For the 15 children followed locally, 6 (35.3%) obtained scores on standardized assessments comparable to those expected for typically developing children and another seven had severe language delays. The remaining two children were within the moderate range of delay in language. Of the seven with severe delay, six presented complex medical issues and developmental delay.

Table 4. Communication development outcomes for children with ANSD

Outcomes	HA	CI	No amplification
Within normal limits	3	3	-
Moderate delay	-	2	-
Severe delay	2	3	2
No local follow up	2	4	1

HA: hearing aid: CI: cochlear implant

Discussion

We described the characteristics of a cohort of children who were identified with ANSD since the implementation of a UNHS program. Prevalence rates of ANSD in this study were 5.3% of children with permanent hearing loss, which is consistent with international reports (Bielecki, Horbulewicz, & Wolan, 2012; Madden et al., 2002; Rance, 2005). It is important to note that prevalence rate can be underestimated in clinical practice because children can be missed when otoacoustic emissions screening is carried out.

In our cohort, we found that there were substantially more males with ANSD, however, Sininger and Starr (2001) state that ANSD shows no difference in the number of boys and girls affected. In our study, more than 50.0% of children were <36 weeks gestation age, a finding consistent with Teagle et al. (2010) who reported prematurity in over 40% of cases with ANSD.

Our findings showed primarily early onset ANSD, corroborating the findings of several other investigators (Berlin, Morlet, & Hood, 2003; Sininger & Oba, 2001). Both genetic factors and conditions in the neonatal period including asphyxia, ototoxic drug exposure, hyperbilirubinemia, neuroinfections, and intraventricular hemorrhage have been reported to be associated with ANSD (Bielecki, Horbulewicz, & Wolan, 2011; Martınez-Cruz, Poblano, & Fernandez-Carrocera, 2008). A close examination of these specific neonatal indicators was beyond the scope of this study. However, based on the JCIH (2007) risk indicators for hearing loss recorded in our database for these children, we documented that 68.2% of patients showed risk indicators, with NICU being the most frequent risk condition. This finding is consistent with several other reports (JCIH, 2007; Rance, 2005; Teagle et al., 2010). ANSD may also occur in association with other syndromes or neurologic pathologies (Raveh, Buller, Badrana, & Attias, 2007) and family history of hearing loss (JCIH, 2007).

The majority (59.2%) of ANSD occurred in babies from the NICU with the remaining in well-babies. These findings are similar to other reports that also found that a substantial number of children with ANSD had risk factors related to NICU admission (Dowley et al., 2009; Sanyelbhaa Talaat, Kabel, Samy, & Elbadry, 2009). Korver, van Zanten, Meuwese-Jongejeugd, van Straaten, and Oudesluys-Murphy (2012) concluded that ANSD is likely more common in the well-baby population but that with current screening techniques, some children go undiagnosed. In the absence of screening using ABR, it is likely that some infants in the well-baby nursery will be missed through screening (Hayes, 2008).

Because of the many possible sites of dysfunction resulting in a diagnosis of ANSD (e.g., inner hair cells, synapse, auditory nerve), audiological profiles have been reported to be highly variable (Norrix & Velenovsky, 2014). Patients with auditory dyssynchrony have a wide range of hearing losses from normal to profound, as documented in several studies (Rance et al., 2007; Sininger & Oba, 2001). Similar to the conclusion from 16 studies reviewed by Roush et al. (2011), our study found that the majority of children had audiological results in the severe-profound range. Consistent with other research (Humphriss et al., 2013), our study showed that more than half of children showed variation in audiometric thresholds over time. All but one child in our cohort had bilateral ANSD, which is aligned with

the results reported in larger studies (Berlin et al., 2010; Bielecki et al., 2012; Sanyelbhaa Talaat et al., 2009).

Previous research has shown that ANSD in many children can be associated with co-morbidities such as developmental delays, learning disabilities, attention deficit disorder, autism spectrum disorders, blindness, cerebral palsy, and motor disorders (Ching et al., 2013; Kirkim et al., 2008; Pelosi et al., 2013; Shallop, 2008; Uhler et al., 2012). These complexities can delay early diagnosis. More than 30.0% (7/22) of children in our study cohort had other disabilities which likely accounted for the average age of diagnosis of over one year (range 0.16 - 49.2 months). This is in contrast to the Ching et al. (2013) study that showed an early diagnosis of ANSD at an average age of 3.3 months.

There is increasing evidence that a substantial number of children with ANSD benefit from hearing aid fitting. ANSD guidelines (Feirn et al., 2013; Northern, 2008) recommend that amplification should be fitted as soon as ear-specific elevated pure-tone and speech detection thresholds are demonstrated by behavioral test procedures. By definition, the presence of otoacoustic emissions in ANSD suggests normal cochlear outer hair cell function and a subset of ANSD patients may exhibit neuromaturation, whereby ABR develops with age (Madden et al., 2002), thus a trial with hearing aids has been recommended as a primary management strategy (Clarin, 2014; Feirn et al., 2013). This study found that there was considerable improvement in aided thresholds and that the majority of children developed auditory skills. However, the literature is quite mixed with some studies showing improvement in speech perception (Roush et al., 2011) and others showing limited improvement in speech perception ability with hearing aids (Berlin et al, 2010; Rance, 2005). Like pure-tone thresholds, speech perception ability can be variable in this population and is difficult to predict from the pure-tone audiogram (Starr, Picton, Sininger, Hood, & Berlin, 1996). Our results for the group of children with hearing aids were not markedly different from those with CIs, pointing to the importance of a well-managed trial period with hearing aids for these children as recommended by Clarin (2014). In our study, the majority of children without other severe disabilities developed their auditory abilities, a finding that is supported by recent research by Ching et al. (2013) in a population-based study examining outcomes in children with hearing loss. That study showed that the presence of additional disabilities was a significant predictor of outcomes at age 3 years for children with hearing loss.

In our study, while almost all children were initially fitted with amplification, an examination of age of fitting showed that almost all received hearing devices after the typical recommended age of 6 months (JCIH, 2007). While this is partly due to a later age of diagnosis, the delay in amplification may also be explained by the characteristics of ANSD. Because ABR results for children with ANSD do not provide a valid estimate of behavioral thresholds, amplification decisions for these children can be challenging. Early hearing aid fitting for these children is highly dependent on reliable behavioral thresholds (Feirn et al., 2013; Norrix & Velenovsky, 2014). Some children with ANSD require more time for threshold determination (Cardon & Sharma, 2011) due to disabilities or medical conditions that include developmental delay. There is also evidence that, like the general population of children with permanent hearing loss, hearing may not initially be the first priority for parents of children who present other medical issues and/or a stormy neonatal course (Uus, Young, & Day, 2012).

Although conventional hearing aids improve sound audibility, there is research showing that they may not resolve temporal processing deficits in some children with ANSD (Rance & Barker, 2009). Some children may not experience the same benefits from hearing aids expected from those with typical sensorineural loss in whom temporal processing is relatively unaffected. This fundamental difference has implications for management and has led to discussions about the best option for affected children (Roush et al., 2011). In our cohort, all children who received CIs had severe to profound hearing loss, however, some authors (Berlin et al., 2010; Breneman et al., 2012) have recommended that children with ANSD should be considered for CI regardless of pure-tone thresholds, particularly when there is poor progress in speech and language development despite amplification. It has been proposed that CI can improve auditory temporal processing by stimulating the synchronous discharge of the auditory nerve (Humphriss et al., 2013; Rance, 2005; Shallop, 2008). In a recent systematic review (Fernandes, Morettin, Yamaguti, Costa, & Bevilacqua, 2015), the authors concluded that there is no difference in the hearing skills of children with ANSD and SNHL who use CIs with respect to speech detection, discrimination, and recognition of words and sentences.

Our findings related to communication development of the 17 children followed in rehabilitation showed that six had communication outcomes within the range expected for typically developing children, with another two children showing moderate language delays. Budenz et al. (2013)

studied children with isolated auditory neuropathy who developed speech and language skills at a level commensurate with their peers who had SNHL and CIs. It is important to note that our study included all children with ANSD including those with an additional diagnosis of cognitive or developmental disorders. Other studies of children with CIs (Cruz et al., 2012; Edwards, 2007) who have SNHL with additional developmental disabilities have typically reported poorer speech and language development compared to children with hearing loss alone. Rance and Barker (2009) compared speech and language outcomes between three groups of children: 1) children with ANSD after CI implantation, 2) children with SNHL after cochlear implantation, and 3) children with ANSD with amplification alone. They found no significant difference between any of the groups on any measure of language development. More recently, in a group of 47 early-identified children, Ching et al. (2013) found there was no significant difference in performance levels or variability between children with and without ANSD at age 3 years, both for children who used hearing aids and those who used CIs.

Our study adds to the growing understanding of the communication outcomes that can be expected from children with ANSD. Our work shows that more than 85.0% of children with ANSD were fitted with current hearing technology and achieved some degree of open-set speech recognition, while almost half either developed spoken language skills similar to their hearing peers or with moderate delays. Undoubtedly, there remain many questions about the management of these children, including the approaches that favor speech-language development, and how soon cochlear implantation should be recommended. Recent research suggests that electrophysiological measurements including electrocochleography (Santarelli, Starr, Michalewski, & Arslan, 2008) and cortical auditory evoked potentials may assist in characterizing auditory function and differences in speech recognition. These techniques may provide insight into the severity of the disorder and whether hearing aids are likely to be helpful (Cardon & Sharma, 2011; Neary & Lightfoot, 2012), which should assist with clinical decision-making for these children.

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