Audiological care following newborn hearing screening: a follow-up study on NICU infants and well-baby nursery infants

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Objective: We investigated outcomes in infants with bilateral hearing loss (BHL) diagnosed by newborn hearing screening (NHS) to demonstrate proper treatment for neonatal intensive care unit (NICU) infants and well-baby nursery (WBN) infants.

Methods: Fifty-three NICU infants and 66 WBN infants who failed the NHS at Kitasato University Hospital between 2003 and 2012 were enrolled in the study. NHS results, age at hearing evaluation, hearing loss severity, concurrent developmental disability, age at hearing aid fitting, hearing aid use, and rehabilitation facilities were studied.

Results: A total of 36 NICU (68%) and 49 WBN infants (74%) were diagnosed with BHL. Intellectual disability (ID) was diagnosed in 25 NICU infants (86.2%) and 14 WBN infants (43%). There were 4 NICU infants (13%) and 17 WBN infants (38%) enrolled in schools for children with hearing impairment. There were 19 NICU infants (61%) and 7 WBN infants (16%) who received special education for children with IDs, and 8 NICU infants (26%) and 21 WBN infants (47%) were enrolled in general preschools or elementary schools.

Conclusions: Many infants with BHL and IDs required special education, regardless of hearing loss severity. Otolaryngologists and speech-language-hearing therapists must regularly evaluate the infants' development and assess their hearing in order to select an appropriate rehabilitation facility for each individual.

Key words: newborn hearing screening, neonatal intensive care unit, well-baby nursery, rehabilitation, follow-up study

Introduction

Numerous reports have examined various aspects of the newborn hearing screening (NHS) system, including participation rates, pass/referral rates, and the prevalence of congenital hearing loss. 1-4 The introduction of the NHS system has made it possible to diagnose and treat infants with impaired hearing earlier than previously possible. 2-7 A combination of the NHS and appropriate therapy is reportedly beneficial in promoting the speech and language development of infants with impaired hearing. 6.7 Some reports have examined the parental response to the NHS system⁸ in cases of false-positive diagnosis and spontaneously improved hearing. 9 Several reports have also investigated the use of the NHS system in infants in the neonatal intensive care unit (NICU).

is recommended as a screening procedure because of the higher incidence of auditory neuropathy in NICU infants compared with non-NICU infants. ¹⁰⁻¹² To reduce the false-positive rate caused by brain immaturity and middle ear effusion in low-birth-weight infants, current protocols recommend performing the first screening test as late as possible. ^{11,13} Notably, hearing loss in NICU infants is significantly associated with a gestational age of less than 36 weeks, antibiotic therapy, and oxygen therapy. ¹⁴ The NHS failure rate and the frequency of improvement in the auditory brainstem response (ABR) threshold are reportedly higher in NICU infants than in well-baby nursery (WBN) infants, but few studies have evaluated the NHS and therapeutic outcomes in NICU infants.

From 1998 – 2001, the Japan Ministry of Health, Labor, and Welfare investigated effective methods for

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the NHS and rehabilitation, and several prefectures have since received public funding to perform the NHS. The NHS system includes facilities for screening (obstetrics and pediatrics clinics), advanced hearing evaluation (otorhinolaryngology departments at general hospitals), and intervention and rehabilitation. Children with impaired hearing are typically educated at schools for the hearing impaired or institutional daycare facilities with support for children with hearing loss. Depending on the severity of hearing loss, some children with hearing impairment may be educated at general nurseries, kindergartens, and schools while receiving rehabilitation at hospitals. Children with both hearing loss and intellectual disability (ID) may be educated at schools for ID or at institutional daycares with support for ID. According to The Oto-Rhino-Laryngological Society of Japan, as of February 2014, 162 facilities in Japan provide advanced hearing evaluation. We also identified 26 institutional daycare centers that provide support for children with hearing loss (age 0-6 years) and 106 schools that provide rehabilitation for children with hearing impairment. Several hospitals and institutional daycare centers provide rehabilitation for children with ID.

More than a decade has passed since the NHS system was introduced in Japan. Most WBN infants with hearing loss are treated according to the NHS protocol; however, infants in the NICU may possess a variety of characteristic features that differ from those of WBN infants and may not be treated according to the NHS protocol. The treatment of NICU infants may differ from that of WBN infants and should be done by examining the hearing examination results, median age at the time of hearing evaluation, diagnosis of hearing loss, age at hearing aid fitting, rehabilitation facilities, and cognitive abilities. In this study, we retrospectively investigated the things which are noted above of NICU and WBN infants who underwent the NHS at Kitasato University Hospital. From the data, we assessed the efficiency of the current NHS system and identified the appropriate method of rehabilitation for NICU infants by comparing the two groups.

Materials and Methods

This study was approved by the ethical review boards of Kitasato University School of Medicine and Kitasato University Hospital. Due the retrospective nature of this study, the requirement for informed consent was waived.

Setting

Kitasato University Hospital is located in Sagamihara,

which is a city of 700,000 people at the southern edge of Tokyo. The facility is used by people residing in northern Kanagawa Prefecture and southern Tokyo. The hospital has a Maternal Fetal Intensive Care Unit and NICU; therefore, many high-risk pregnancies and neonates with major disorders during the perinatal period are referred to the facility. The NHS was implemented at Kitasato University Hospital beginning in July 2002. Infants who failed the NHS were referred to the Department of Otolaryngology for additional hearing evaluation and diagnosis, hearing aid fitting, and rehabilitation by otolaryngologists and speech-language-hearing therapists.

Participants

The study was conducted between 2003 and 2012. During the study period, 178 infants were referred to the Department of Otorhinolaryngology for further hearing evaluation. Of these, 59 infants were excluded for the following reasons: 8 NICU infants who were diagnosed with genetic or cardiac disease died; 8 infants were transferred to another hospital; 3 discontinued the hospital visits; 8 were diagnosed with a malformed external ear (i.e., microtia and aural atresia); 28 infants were referred from maternity clinics with little information on their NHS results; and 4 infants relocated to the local area and were examined at our hospital after being diagnosed elsewhere.

The 119 participants were divided into two groups: 53 NICU infants (45%) and 66 WBN infants (55%). Among the NICU infants, 16 had extremely low-birthweight, 4 infants had birth weights of 1,000 g - 1,499 g, 23 had respiratory problems, 30 had cardiac disease, and 11 had birth abnormalities (overlapping data). The median follow-up period in the NICU infants was 43 months (range 1-115 months), and in WBN infants, the median follow-up was 47 months (1-118 months). There was no significant difference between the two groups (Wilcoxon rank-sum test, P = 0.97). At the time of this publication, 31 NICU infants (58%) and 45 WBN infants (68%) were continuing with treatment.

Newborn hearing screening protocol and diagnosis The age of the infants at the hearing evaluation, diagnosis, hearing aid fitting, and rehabilitation facilities were determined based on review of the pediatric and otorhinolaryngology department records.

1. Hearing examination

In NICU infants, very low-birth-weight infants (<1,500 g) were tested by AABR, and extremely low-birth-weight infants (<1,000 g) were tested by ABR, because

they are at high risk for hearing loss. Other infants were tested first by otoacoustic emissions (OAE) followed by AABR. The WBN infants were tested using only OAE, only AABR, or both OAE and AABR because there were differences in the availability of screening devices at maternity hospitals. In our hospital, the infants were tested first by OAE and then by AABR.

2. Diagnosis of hearing loss

The infants were diagnosed based on the ABR threshold and behavioral hearing testing of children. Hearing was evaluated based on behavioral observation audiometry, visual reinforcement audiometry, conditioned orientation response audiometry, peepshow test, and play audiometry as appropriate for the developmental maturity of the infant. In behavioral observation audiometry, the testing of infants and young children is accomplished without reinforcement of responses and is reliant on the subjective observation of responses under structured conditions. The auditory responses of infants and young children were also described in terms of reflexive or attentive behavior. During the conditioning period, both visual reinforcement audiometry and conditioned orientation response audiometry used TV monitors that were switched on simultaneously with the presentation of the auditory signal. During the testing phase, the monitors were switched on immediately following the response of the child looking toward the TV monitors. The visual reinforcement audiometry response required only detection of the auditory signal as a prerequisite to an appropriate head-turn. The conditioned orientation response audiometry response required that the child localize the test signal to determine which of two reinforcers (left or right) was the appropriate response. In the peep-show test, each child was conditioned during practice trials to quickly depress a button immediately following presentation of an auditory stimulus. A brief animated color video presentation followed each correct response. False responses produced no visual reinforcement. In play audiometry, the child was conditioned during practice trials to place marbles in a case immediately following presentation of an auditory stimulus.

If the right and left hearing thresholds could not be determined by behavioral hearing testing because of severe ID, then the infant was instead diagnosed based on the ABR threshold. Auditory brainstem response testing typically occurred during sleep induced by administration of triclofos sodium syrup. Recordings were made using click stimuli. The stimulus intensity began at 90 dBnHL and was reduced in 10 dB intervals.

The ABR threshold was considered the stimulus intensity at which the ABR was absent. When there was no ABR at 90 dBnHL, the stimulus intensity was increased to 105 dBnHL to confirm hearing loss. Stimulus delivery and response averaging were conducted using Neuropack (Nihon Koden).

Hearing loss was classified as mild (26-39 dBHL), moderate (40-69 dBHL), severe (70-89 dBHL), or profound (>90 dBHL). When the right and left ears differed in severity, the diagnosis was made according to the ear with better hearing.

3. Hearing aid fitting, rehabilitation facilities, and ID Children with bilateral hearing loss were fitted with hearing aids for both ears. The age at the time of hearing aid fitting was determined based on review of the pediatric and otorhinolaryngology department records. The rehabilitation facility, the school for children with hearing loss, special education for ID, or hospital (and educated by general preschools or elementary school) was also determined based on the records. There was a potential relationship between the presence of ID and the age at hearing aid fitting or the choice of rehabilitation facility; therefore, we investigated the presence or absence of ID.

To diagnose ID, the intellectual development of each infant was evaluated using the Japanese equivalent of the Stanford-Binet Intelligence scale, the Wechsler Preschool and Primary Scale of Intelligence, or the Wechsler Intelligence Scale for Children (third edition). If the infant was unable to complete the intelligence tests, then the infant's intellectual development was gauged based on medical records from the pediatrician.

Statistical analysis

The median age at hearing examination, hearing aid use, median age at hearing aid fitting, median follow-up period, prevalence of concurrent bilateral hearing loss and ID, and type of rehabilitation facility were subjected to statistical processing using the χ -squared test and Wilcoxon rank-sum test. Statistical significance was designated at P < 0.05.

Results

Hearing examination results

The diagnostic findings in each infant group are summarized in Table 1. Table 1 shows that the majority of infants in either the NICU or WBN groups had no response in either ear for the OAE and AABR tests. Two patients in the NICU group had clear responses in both ears. In 16 NICU infants, neither the OAE or AABR was

administered; these infants were tested by ABR. The ABR threshold was elevated by at least 30 dBHL in both ears. Nineteen patients in the WBN group had no response in one ear.

Median ages at hearing evaluation

Figure 1 shows the median ages at the time of hearing evaluation. The median age at the hearing examination in the NICU infants was 4 months (range, 0—29 months). Twenty-six infants were examined within 3 months of birth, 14 within 6 months, 9 within 12 months, and 4

more than 12 months after birth. The median age of WBN infants at hearing examination was 1 month (range 0-20 months). The examination in 51 infants was conducted within 3 months of birth, 7 within 6 months, 6 within 12 months, and two 2 more than 12 months after birth.

The median age at the time of hearing examination was significantly earlier in WBN infants than in NICU infants (Wilcoxon rank-sum test, P < 0.05). The percentage of infants who received a hearing examination within 3 months of birth was significantly higher among

Table 1. Hearing examination results in NICU infants $(n = 37)^1$ and WBN infants (n = 66)

	No response in either ear	Response in only one ear	Clear response in both ears
NICU infants			
OAE	7	1	1
AABR	3	1	0
OAE + AABR	18	5	1
WBN infants			
OAE	13	7	0
AABR	19	9	0
OAE + AABR	15	3	0

¹In 16 NICU infants, neither the OAE nor AABR testing was performed; these infants were tested by ABR.

NICU, neonatal intensive care unit; WBN, well-baby nursery; OAE, otoacoustic emission test; AABR, automated auditory brainstem response test; ABR, auditory brainstem response test

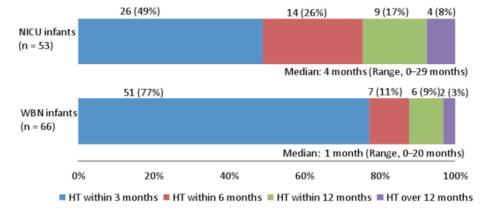


Figure 1. Age of NICU infants and WBN infants at initial hearing examination. The median age of WBN infants at the time of the hearing examination was significantly less than that of NICU infants (Wilcoxon rank-sum text, P < 0.05). The percentage of infants who received a hearing examination within 3 months of birth was significantly higher in WBN infants than in NICU infants (χ -square test, P < 0.05). NICU, neonatal intensive care unit; WBN, well-baby nursery; HT, hearing test

²One WBN infant who failed the OAE in one ear and could not be tested in the other ear was included in the "no response in either ear" group.

the WBN infants than the NICU infants (77% vs. 49%, respectively, χ -square test, P < 0.05).

Diagnosis of hearing loss

The right and left hearing thresholds were measured using behavioral hearing testing in 20 of 53 (38%) NICU infants. A total 33 of 53 (62%) NICU infants were diagnosed by ABR. As shown in Figure 2, of the 53 NICU infants: 8 (15%) were diagnosed with normal hearing, 9 (17%) with unilateral hearing loss, 15 (28%) with mild hearing loss, 12 (23%) with moderate hearing loss, 3 (6%) with severe hearing loss, and 6 (11%) with profound hearing loss.

Behavioral hearing testing was completed and the right and left ear hearing thresholds were measured in 44 of 66 (67%) WBN infants. A total of 22 of 66 (33%)

WBN infants were diagnosed by ABR. Of the 66 WBN infants, 10 (15%) were diagnosed with normal hearing, 7 (11%) with unilateral hearing loss, 13 (20%) with mild hearing loss, 18 (27%) with moderate hearing loss, 4 (6%) with severe hearing loss, and 14 (21%) with profound hearing loss.

The right and left hearing thresholds were measurable by behavioral hearing testing of children in significantly fewer NICU infants compared to WBN infants (χ -square test, P < 0.05). There were no significant differences between the NICU infants and WBN infants according to the severity of hearing loss (χ -square test, P = 0.49).

Age at hearing aid fitting

Figure 3 shows the use of hearing aids in 36 NICU infants and 49 WBN infants with binaural hearing loss. There

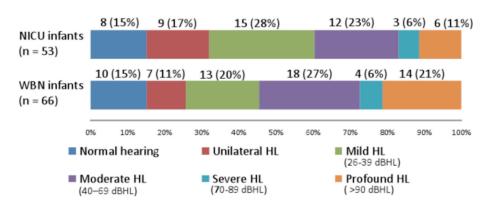


Figure 2. Prevalence of hearing loss in NICU infants and WBN infants. There was no significant difference between NICU infants and WBN infants according to the severity of hearing loss (χ -square test, P = 0.49). NICU, neonatal intensive care unit; WBN, well-baby nursery; HL, hearing loss

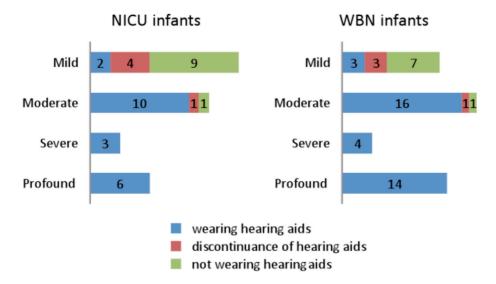


Figure 3. Hearing aid use in NICU infants and WBN infants with binaural hearing loss. There was no difference in hearing aid use between NICU infants and WBN infants (χ -square test, P = 0.20). NICU, neonatal intensive care unit; WBN, well-baby nursery

were 5 NICU infants and 2 WBN infants who received middle ear tube placement surgery for otitis media with effusion. None of them achieved normal hearing after surgery. Six of 15 NICU infants with mild hearing loss were fitted with a hearing aid, but 4 infants stopped wearing the devices. A total of 11 of 12 NICU infants with moderate hearing loss received a hearing aid, and 1 infant stopped wearing the device. All 3 NICU infants with severe and all 6 infants with profound hearing loss were fitted with hearing aids. Six of 13 WBN infants with mild hearing loss were fitted with hearing aids, however, 3 infants stopped wearing them. A total of 17 of 18 WBN infants with moderate hearing loss were fitted with hearing aids, but one infant stopped wearing the device. All 4 WBN infants with severe and 14 with profound hearing loss were fitted with hearing aids. Subsequently, 3 WBN infants with profound hearing loss received cochlear implant insertion surgery. There were no significant differences between the NICU infants and WBN infants in hearing aid use (χ -square test, P = 0.20).

The median age when infants with bilateral hearing loss were first fitted with hearing aids is shown in Table 2. The median age at the time of hearing aid fitting was significantly lower in WBN infants (10 months) than in NICU infants (15 months, Wilcoxon rank-sum test, P < 0.05). In WBN infants, the median age at the time of hearing aid fitting for those with severe to profound hearing loss was significantly earlier than in infants with mild to moderate hearing loss (Wilcoxon rank-sum test, P < 0.05). However, in NICU infants, there was no significant difference in the time of hearing aid fitting according to hearing loss severity (Wilcoxon rank-sum test, P = 0.79). In cases of severe to profound hearing loss, the median age at the time of hearing aid fitting in WBN infants was significantly earlier than in NICU infants (Wilcoxon rank-sum test, P < 0.05). There was a

Table 2. Median age of first hearing aid fitting in infants with bilateral hearing loss

	All infants	Mild to moderate hearing loss	Severe to profound hearing loss	P-value*
NICU infants	15 (3-74)	15 (3-74)	15 (4-27)	0.79
WBN infants	10 (3-26)	13 (5-26)	5 (3-10)	< 0.05
P-value**	< 0.05	0.78	< 0.05	

All data are presented as the median (range) and measured in months.

NICU, neonatal intensive care unit; WBN, well-baby nursery

Table 3. Rehabilitation facilities for NICU and WBN infants according to severity of hearing loss

		Hearing loss severity		D 1 4
	n	Mild to moderate	Severe to profound	P-value*
NICU infants	31			0.62
School for children with hearing impaired		0 (0%)	4 (44%)	
Special education for ID		14 (64%)	5 (56%)	
General preschool or elementary school		8 (36%)	0 (0%)	
WBN infants				< 0.05
School for children with hearing impaired		3 (10%)	14 (88%)	
Special education for ID		7 (24%)	0 (0%)	
General preschool or elementary school		19 (66%)	2 (13%)	

Data are presented as the number (percentage) of infants. NICU, neonatal intensive care unit; WBN, well-baby nursery

^{*}Comparison between infants with mild to moderate vs severe to profound hearing loss in NICU and WBN using the Wilcoxon rank sum test

^{**}Comparison between NICU infants and WBN infants in mild/moderate, severe/profound, and whole population groups using the Wilcoxon rank sum test

^{*}Comparison of hearing loss severity within NICU and WBN infants who were enrolled at a facility specializing in intellectual disability using the chi-square test

difference, but there was not a statistically significant difference between WBN infants and NICU infants with mild to moderate hearing loss (Wilcoxon rank-sum test, P = 0.78).

Rehabilitation and developmental delay

Table 3 lists the distribution of rehabilitation facilities for each of the infant groups. Intellectual disability was diagnosed in 25 of 29 (86.2%) NICU infants with bilateral hearing loss. Their median age was 76 months (range 5 — 123 months), and the grades of ID were: severe 16, moderate 4, mild 4, and unclear 1. None of the 22 NICU infants with mild or moderate hearing loss was enrolled in a school for children with hearing impairment. None of the 9 NICU infants with severe or profound hearing loss was enrolled in a general preschool or elementary school.

Intellectual disability was diagnosed in 14 of 33 (43%) WBN infants with bilateral hearing loss. The median age of WBN infants with ID was 78 months (range 17—114 months), and the ID grades were: severe 2, moderate 5, mild 6, and unclear 1. Nineteen of 29 (66%) WBN infants with mild or moderate hearing loss were enrolled in general preschools and elementary schools. Fourteen of 16 (88%) WBN infants with severe or profound hearing loss were enrolled in schools for children with hearing impairment.

The prevalence of concurrent bilateral hearing loss and ID was significantly higher in NICU infants than in WBN infants (χ -square test, P < 0.05). In the NICU infants with bilateral hearing loss, the type of rehabilitation facility was not associated with the severity of hearing loss (χ -square test, P = 0.62). By contrast, in the WBN infants with bilateral hearing loss, enrollment in a school for children with hearing impairment was associated with severe hearing loss (χ -square test, P < 0.05).

Discussion

Hearing evaluation and diagnosis following the NHS In this study, the median age of NICU infants at the first hearing examination was significantly greater than that of WBN infants. Uus et al. 15 reported similar results and found that the age at the first audiologic assessment was significantly higher in NICU infants than in WBN infants (10.8 ± 3.0 and 4.7 ± 3.0 weeks, respectively). The authors attributed this trend to health differences between the two infant populations. At our hospital, the initial hearing evaluation was conducted in most of the NICU infants using ABR or behavioral hearing testing immediately before discharge and when their weight reached 2,500 g (5.5 lbs). Until that time, the NICU

infants received intense medical care and struggled with life-threatening conditions. The long duration of hospitalization for NICU infants is likely an essential factor contributing to delayed hearing evaluations.

Our follow-up study clearly indicated that the prevalence of each form of hearing loss did not differ significantly between NICU infants and WBN infants. Russ and Kuo¹⁶ reported that additional difficulties were also experienced in obtaining accurate audiological evaluations when the child was developmentally delayed. In some infants with multiple disabilities, it was difficult to measure the hearing threshold correctly, but we were able to successfully measure the right and left hearing thresholds in 38% of NICU infants and 67% of WBN infants using behavioral hearing testing. In order to accurately evaluate the hearing threshold, the selection of an appropriate audiometric method corresponding to the development and regular follow-up examination of the infant are essential. Our results are consistent with van Straaten et al.¹⁷ who found that 18.2% of the infants in their NICU population had normal hearing, 19.5% had unilateral hearing loss, and 62.3% had bilateral hearing loss. However, hearing loss severity differed in this previous study, which reported mild hearing loss in 7% of the NICU infants, moderate to severe loss in 51%, and profound hearing loss in 41% of the NICU infants. However, we observed mild hearing loss in 42% of the NICU infants, moderate to severe hearing loss in 42%, and profound hearing loss in 17% of the NICU infants. The prevalence of profound hearing loss in the NICU infants was lower in the present study than in the previous studies. In a report by Dalzell et al., 18 56% of the NICU infants were diagnosed with mild to moderate hearing loss and severe to profound loss in 44%. Among the WBN infants, 52% infants were diagnosed with mild to moderate hearing loss, and 48% were diagnosed with severe to profound loss. There were no significant differences in the severity of hearing loss between the two groups in their study. The rate of severe to profound hearing loss was notably higher in a report by Dalzell et al. 18 compared to the present study.

Kang et al.¹⁹ observed that the hearing thresholds of infants with congenital sensorineural hearing loss can change during the first year of life, and in severely premature babies, delayed maturation of the auditory pathway may contribute to spontaneous hearing threshold recovery. van Straaten et al.¹⁷ performed ABR hearing evaluations within 3 months of birth in all infants. This variation in the time of hearing examination likely contributes to the variable prevalence of hearing loss severity between the studies.

Hearing aid fitting in infants with bilateral hearing loss Remarkably, in the present study, there was no significant difference in the use of hearing aids between the NICU infants and the WBN infants. However, the median age of hearing aid fitting was significantly greater in the NICU infants than in the WBN infants. Further, the age of hearing aid fitting was delayed in both groups (10 months in WBN infants and 15 months in NICU infants), although the median age of hearing evaluation was 1 month in WBN infants and 4 months in NICU infants. One reason for the delay was that parents resisted using hearing aids. Yee-Arellano et al.²⁰ indicated that rehabilitation and the use of hearing aids began after 6 months, and parental denial that their child could have hearing loss was the primary reason. Sjoblad et al.21 also noted that families may respond negatively during the early stage of identification and diagnosis, or they may have been unaware of the benefit of early amplification.

The factors contributing to delayed hearing aid fitting in NICU infants include the following: delayed hearing evaluation due to their longer duration of hospitalization; inability to undergo periodic hearing examination due to recurrent hospitalization after discharge from the NICU; and the higher prevalence of severe ID, which delays hearing examination. Uus et al., ¹⁵ Dalzell et al., ¹⁸ Sjoblad et al.,21 and Spivak et al.22 observed that the median age of hearing aid fitting in WBN infants was significantly earlier than in NICU infants. Dalzell et al.18 attributed the later ages of hearing aid fitting in NICU infants to illness and developmental delay, and Uus et al. 15 attributed the trend in NICU infants to their poorer health and longer duration of hospitalization. Spivak et al.²² reported that the history of treatment in the NICU was related to delays in follow-up services because these infants were likely to have significant health and developmental issues that could limit their availability for follow-up. Sjoblad et al.²¹ described that waiting until the child is old enough to perform reliably on behavioral measures is certain to impose considerable delays, particularly in infants with developmental delay or disabilities.

The Joint Committee on Infant Hearing (JCIH) guidelines recommend performing NHS within 1 month of birth and a hearing examination within 3 months; if required, hearing aid fitting and rehabilitation should begin within 6 months JCIH.²³ However, in the present study, this standard was not met in a significant proportion of infants, particularly among NICU infants. Spivak et al.²² reported that only 39% of infants diagnosed with hearing loss requiring amplification were fit for hearing aids within the recommended time. Further, Durieux-Smith, et al.²⁴ reported that few children in their study

met the JCIH²³ recommendations of diagnosis at 3 months and underwent intervention at 6 months. As indicated by our data, hearing aid fitting can be individually tailored not only in WBN infants but also in NICU infants. While it is ideal to fit hearing aids in children with hearing loss early, it is not always possible in specific cases where there are concomitant medical issues.

Selection of rehabilitation facilities

We found that 86.2% of NICU infants with bilateral hearing loss also had ID. There have been several reports of NICU infants with bilateral hearing loss and concurrent ID. Robertson et al.²⁵ observed that 75% of children with severe or profound hearing loss and 50% of children with mild or moderate hearing loss also had ID. Dalzell et al.18 found that 52% of NICU infants who were fitted with hearing aids also had ID. The prevalence of concurrent bilateral hearing loss and ID in NICU infants in the present report was higher than that in earlier studies. In addition, the prevalence in the present report varied according to the severity of systemic disease in the NICU infants. Our data suggest that in ID infants, particularly NICU infants who have additional disabilities, the rehabilitation facility was selected not only according to the severity of hearing loss but also according to general health and intellectual development. All infants require appropriate rehabilitation corresponding to their individual needs.

Otolaryngologists and speech-language-hearing therapists can advise families on the most appropriate rehabilitation facility for their infants based on the results of several different assessments. Some infants may need to be enrolled in schools for children with hearing impairment because their primary disability is hearing loss, while others may require enrollment in a center for ID because their primary disability is developmental impairment. Otolaryngologists and speech-languagehearing therapists should coordinate their care with that of pediatricians and other clinicians to ensure that the rehabilitation and family support accommodates all relevant factors including the general health, mental development, and family environment of the infant. Based on the collective diagnostic findings, comprehensive decisions can be made on the best time to incorporate hearing aids and the best rehabilitation facility.

Study limitations

The ages of the infants at the NHS was not investigated in the present study. In many cases, the ages of the infants at NHS was not even recorded; rather, only the test results were included in the medical record. The ID status could not be compared between NICU infants and WBN infants; therefore, no conclusions could be drawn on this point. In addition, the data were obtained from only one location and may not be generalized to other locations.

Conclusions

The ages of infants at their hearing examinations and at their hearing aid fittings were significantly later in the NICU infants than in the WBN infants. However, there was no significant difference in the use of hearing aids between the NICU infants and the WBN infants. Otolaryngologists and speech-language-hearing therapists must regularly evaluate the developmental ability of their infants in addition to assessing hearing. Based on these evaluations, clinicians can select an appropriate rehabilitation facility that accommodates the hearing and developmental abilities of each individual infant.

References

- 1. Mehl AL, Thomson V. Newborn hearing screening: the great omission. *Pediatrics* 1998; 101: E4.
- 2. Russ SA, Dougherty D, Jagadish P. Accelerating evidence into practice for the benefit of children with early hearing loss. *Pediatrics* 2010; 126 (Suppl 1): S7-18.
- 3. Pourarian S, Khademi B, Pishva N, et al. Prevalence of hearing loss in newborns admitted to neonatal intensive care unit. *Iran J Otorhinolaryngol* 2012; 24: 129-34.
- 4. Schnell-Inderst P, Kunze S, Hessel F, et al. Screening of the hearing of newborns Update. *GMS Health Technol Assess* 2006; 2: Doc20.
- 5. Vohr B, Jodoin-Krauzyk J, Tucker R, et al. Early language outcomes of early-identified infants with permanent hearing loss at 12 to 16 months of age. *Pediatrics* 2008; 122: 535-44.
- 6. Clemens CJ, Davis SA, Bailey AR. The false-positive in universal newborn hearing screening. *Pediatrics* 2000; 106: E7.
- 7. Lim HW, Kim EA, Chung JW. Audiological follow-up results after newborn hearing screening program. *Clin Exp Otorhinolaryngol* 2012; 5: 57-61.
- 8. Colella-Santos MF, Hein TA, de Souza GL, et al. Newborn hearing screening and early diagnostic in the NICU. *Biomed Res Int* 2014. doi: 10.1155/2014/845308.

- 9. Slaughter JL, Meinzen-Derr J, Rose SR, et al. The effects of gestational age and birth weight on false-positive newborn-screening rates. *Pediatrics* 2010; 126: 910-6.
- 10. Lim SY, Simser J. Auditory-verbal therapy for children with hearing impairment. *Ann Acad Med Singapore* 2005; 34: 307-12.
- 11. Patel H, Feldman M. Universal newborn hearing screening. *Paediatr Child Health* 2011; 16: 301-10.
- van Dommelen P, van Straaten HL, Verkerk, PH; Dutch NICU Neonatal Hearing Screening Working Group. Ten-year quality assurance of the nationwide hearing screening programme in Dutch neonatal intensive care units. *Acta Paediatr* 2011; 100: 1097-103
- 13. Freitas VS, Alvarenga Kde F, Bevilacqua MC, et al. Critical analysis of three newborn hearing screening protocols. *Pro Fono* 2009; 21: 201-6.
- 14. Cristobal R, Oghalai JS. Hearing loss in children with very low birth weight: current review of epidemiology and pathophysiology. *Arch Dis Child Fetal Neonatal Ed* 2008; 93: F462-8.
- 15. Uus K, Bamford J. Effectiveness of population-based newborn hearing screening in England: ages of interventions and profile of cases. *Pediatrics* 2006; 117: e887-93.
- 16. Russ SA, Kuo AA, Poulakis Z, et al. Qualitative analysis of parents' experience with early detection of hearing loss. *Arch Dis Child* 2004; 89: 353-8.
- 17. van Straaten HL, Hille ET, Kok JH, et al.; Dutch NICU Neonatal Hearing Screening Working Group. Implementation of a nation-wide automated auditory brainstem response hearing screening programme in neonatal intensive care units. *Acta Paediatr* 2003; 92: 332-8.
- 18. Dalzell L, Orlando M, MacDonald M, et al. The New York State universal newborn hearing screening demonstration project: ages of hearing loss identification, hearing aid fitting, and enrollment in early intervention. *Ear Hear* 2000; 21: 118-30.
- 19. Kang MY, Jeong SW, Kim LS. Changes in the hearing thresholds of infants who failed the newborn hearing screening test and in infants treated in the neonatal intensive care unit. *Clin Exp Otorhinolaryngol* 2012; 5 (Suppl 1): S32-6.
- 20. Yee-Arellano HM, Leal-Garza F, Pauli-Muller K. Universal newborn hearing screening in Mexico: results of the first 2 years. *Int J Pediatr Otorhinolaryngol* 2006; 70: 1863-70.
- 21. Sjoblad S, Harrison M, Rouch J, et al. Parents' reactions and recommendations after diagnosis and hearing aid fitting. *Am J Audiol* 2001; 10: 24-31.
- 22. Spivak L, Sokol H, Auerbach C, et al. Newborn hearing screening follow-up: factors affecting hearing aid fitting by 6 months of age. *Am J Audiol* 2009; 18: 24-33.

- 23. Joint Committee on Infant Hearing, et al. Year 2000 position statement: principles and guidelines for early detection and intervention programs. *Pediatrics* 2000; 106: 798-817.
- 24. Durieux-Smith A, Fitzpatrick E, Whittingham J. Universal newborn hearing screening: a question of evidence. *Int J Audiol* 2008; 47: 110.
- 25. Robertson CM, Howarth TM, Bork DL, et al. Permanent bilateral sensory and neural hearing loss of children after neonatal intensive care because of extreme prematurity: a thirty-year study. *Pediatrics* 2009; 123: e797-807.