EVALUATION OF UNIVERSAL NEWBORN HEARING SCREENING IN JAPAN: AN ANALYSIS OF THE LITERATURE

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Introduction To address delayed language development associated with severe-to-profound congenital hearing loss (CHL), universal newborn hearing screening (UNHS) has been implemented in many countries. In Japan, approximately 27,000 neonates (2.5% of newborn) are screened annually through public-funded programs. While foreign literature highlights the need for assessment, in Japan this has hitherto not been evaluated in detail.

Objective To investigate the efficacy of UNHS in Japan.

Methods We used two criteria to evaluate UNHS; accuracy of screening and the effectiveness of early detection, and searched the major medical and social research journal data bases for related research papers. Eleven articles were identified providing information on accuracy of screening tests and two on effectiveness of early detection.

Results 1) In two prefecture-based studies, 900 and 1,272 newborns were screened to find one case of bilateral CHL. In nine hospital-based programs, the number tested ranged from 313 to 1,910. None of the studies measured the sensitivity and specificity against a best practice standard. 2) The two studies suggested that early intervention might be beneficial, but neither provided conclusive evidence.

Conclusion The effectiveness of UNHS in Japan is still equivocal because of the difficulties associated with differential diagnosis of hearing loss and normal hearing at the early stage of life and the lack of evidence on effectiveness of early intervention. Before nationwide implementation of UNHS, these issues should be fully investigated and evaluated.

Key words: Newborn hearing screening, Congenital hearing loss, Deafness, Evidence based healthcare

I. Introduction

Severe-to-profound, bilateral congenital hearing loss (CHL) leads to the delays in language learning and speech development[1,2], and estimates of the prevalence range from 1/1,000 to 2/1,000[3~5]. Delays in hearing and language acquisition, however, are believed to be ameliorated or eliminated through early intervention such as speech and language therapy, employment of amplification aids, and family support[6,7]. Yoshinaga-Itano et al.[6] have highlighted the fact that early intervention during the first six months after birth is critical to the development of speech and language skills. In order to detect CHL and start intervention at an early age, hearing screening using Crib-O-Gram[8] and measurement of blood flow to the fingertips[9] have been applied to newborn infants. Because of the problems associated with the need for high operator skill, the time consuming nature and low sensitivity of the testing procedure, previous screening tests are not considered appropriate to universal newborn hearing screening (UNHS), but rather only for selective screening of high-risk cases[8~11]. Developments in testing technology since the 1970s, in particular regarding the automated auditory brainstem response (ABR) and automated otoacoustic emissions (OAE), has allowed the much hoped-for UNHS. The first trial was carried out in Hawaii in 1992[12] and UNHS has been implemented in almost all states in the United States since then. A number of other countries, including at the developing stage, have also implemented UNHS[13]. In Japan, in line with the recommendation of the Ministry of Health, Labour and Welfare in 2001, eight out of 47 prefectures and two cities have introduced UNHS as part of a model trial, up to 2003. Over 27,000 neonates

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(2.5% of newborns) have been screened through public programs[14~17]. In 2004, three prefectures and a city began UNHS[7], and the number is increasing. In addition, around 30% of obstetrical hospitals have been reported to have purchased automated ABR or automated OAE equipment[18], suggesting that a considerable number of infants are screened through privately-run practices.

In spite of this increase in UNHS testing, its effectiveness has not been evaluated in Japan. The foreign literature points to reduced effectiveness of the screening test being a waste of scarce resources, and also that testing has a number of unfavourable psychological consequences such as the labeling of infants as false-positives and false-negatives[19~24]. The idea that the implementation of universal testing should be based on effectiveness is gradually been recognised in Japan[25], and the present study aimed to investigate this question through evaluation of the literature in the area.

II. Evaluation Methods for the Effectiveness of UNHS

We used two criteria for evaluating the effectiveness of UNHS, as proposed by the United States Preventive Services Task Force (USPSTF)[26]: accuracy of the screening test and positive results with early detection. Appraisal of screening test is needed to determine how accurately diseases objectively detected. Generally, four indices are applied: sensitivity, specificity, positive predictive value, and negative predictive value. With early detection, whether intervention can lead to improvement in health outcomes is the question. Early intervention for CHL includes specific education for the deaf initiated after diagnosis, as well as medical treatment such as the fitting of hearing aids and cochlear implantation.

We searched two major data bases used in the area of medicine and social sciences in Japan (Japana Centra Revuo Medicina Web and FELIX) for relevant literature published in Japan up till April 2005, using the keywords listed in Table 1. We confined our search to original and review articles. We also used annual reports of the relevant study group set up by the Ministry of Health, Labour and Welfare in 1998.

In consideration of the methodological strength of the evidence, our review included controlled trials and observational studies on: (1) screening using OAE and/or ABR in the general newborn population, and (2) effects of intervention after early detection by UNHS.

(1) Of 129 articles identified in the first category shown in Table 1, 13 met the relevant criteria.

Table 1. Keywords used in the literature search

1.	("hearing tests" and "newborn screening") or "newborn hearing test" or "newborn hearing screening"
2.	("hearing loss" or "deaf") and ("early detection" and "effectiveness")
3.	("hearing loss" or "deaf") and ("early intervention" or "onset of intervention" or "early education") and "effectiveness"

Two articles were excluded because of the screening procedures employed were insufficient or because the size of the sample was too small.

(2) Of 11 articles identified in the second or third category in Table 1, two met the relevant criteria.

III. Accuracy of the Screening Test

Two prefecture-based programs[27,28] and nine hospital-based programs[29~37] provided information about the extent of UNHS testing (Table 2). The number of neonates screened as a proportion of the testing population ranged from 70% to 100%. All programs listed in Table 2 used a two, three or five stage protocol, in which an infant who fails the initial test is repeatedly tested in the hospital or at an outpatient clinic, and one who fails the final screening test is referred for complete diagnostic evaluation.

In the two prefecture-based studies, 900 and 1,272 newborns were screened to find each bilateral CHL case. In the eight hospital-based programs, the number of neonates tested ranged from 313 to 1,910. Some of the values are lower than previously reported estimates of the prevalence of severe-to-profound CHL among newborns (1/1000–2/1000)[3~5]. In three studies (Yamamoto et al.[30], Matsuo et al.[33], Kawashima et al.[35]), small sample sizes might have accounted for these low estimates.

The positive predictive value varied considerably between programs. In a prefecture-based study which included about 20,000 infants, PPV after the final-stage screening test was 18%, with two of 11 infants referred for diagnostic test being true-positives, and nine false-positives[27]. In other studies involving smaller numbers of subjects, the PPV ranged from 0% to 50%.

Between 0.2% and 1.6% of newborns were referred for audiological assessment and over 50% of those referred were false-positives. According to the review by USPSTF, 1% to 3% of newborns were referred and over 90 % of those were

Table 2. Studies of Universal Newborn Hearing Screening

Study	Region	Period	Device	Stages	Total subjects	Screened subjects (%)[a]	Cases[b]	Yield[c]	Referred subjects[d]	Referral rate	PPV
Mimaki et al, 2003[27)]	Prefecture (41 facilities)	7/01–1/03	A-ABR	2	19,467	19,078 (98)	15	1,272	85	0.5	18
Asamura et al, 2004[28)]	Prefecture[e]	10/02–7/03	A-ABR A-OAE	2 2	7,345	6,301 (86)	7	900	19	0.3	37
Mimaki et al, 2000[29)]	One hospital	11/97–2/00	A-ABR	2	2,962	2,843 (96)	7	406	23	0.8	30
Yamamoto et al, 2001[30)]	One hospital	1/2/99–31/1/01	A-ABR	3	747	747 (100)	1	747	4	0.5	25
Fujita et al, 2001[31)]	One hospital	8/98–4/00	A-ABR	2	3,427	2,731 (80)	2	1,366	13	0.5	15
Matsumoto et al, 2002[32)]	One hospital	9/99–8/01	A-ABR	5	NR	916 (NA)	1	916	3	0.3	33
Matsuo et al, 2003[33)]	One hospital	1/4/00–31/3/01	A-OAE	—[f]	313	313 (100)	1	313	5	1.6	20
Iwasaki et al, 2003[34)]	Two hospitals	1/00–12/01	A-ABR	2	4,092	4,085 (99.8)	8	511	29	0.7	28
Kawashima et al, 2004[35)]	One hospital	2/02–4/03	A-ABR	2	NR	826 (NA)	2	413	4	0.5	50
Yamagishi et al, 2004[36)]	One hospital	1/7/01–30/6/03	A-OAE	3	542	527 (97)	0	NA	8	1.5	0
Shimizu et al, 2004[37)]	One hospital	12/99–5/03	A-ABR	3	5,450	3,821 (70)	2	1,910	8	0.2	25

NNS indicates number needed to screen; CHL, congenital hearing loss; PPV, positive predictive value; A-ABR, automated auditory brainstem response; A-OAE, automated otoacoustic emissions; NR, not reported; NA, not applicable

[a] The proportion of the number screened to number offered.

[b] Cases of children having diagnosed as bilateral severe or profound congenital hearing loss.

[c] The number screened to identify one case.

[d] The number of subjects who failed the final screening test and were referred for complete diagnostic evaluation.

[e] Number of facilities unknown.

[f] Referred infants were repeatedly screened in the hospital as many times as possible.

false-positives[38)]. The results might be due to the two-stage protocol prevailing in Japan. Longer hospitalization after delivery in Japan than in other countries makes it possible to screen infants failing the initial test before discharge.

Two important problems exist for evaluation of accuracy of screening test. Firstly, none of the studies reported sensitivity and specificity against a best practice standard. Behavioral tests appropriate for best practice standard are performed in Japan with one or two year olds[39)]. Calculation of indices against best practice standards needs follow-up research, but no follow-up data are available on infants who were given true positive and false negative diagnoses after screening. It is difficult to establish best practice standards for the following two reasons. (1) Some hearing impairment in infants improves naturally during infancy[40)]. This may be due to delays in neurological development, but the prevalence and responsible risk factors remain uncertain. (2) Some infants suffer progressive hearing loss[41)], the prevalence of which is also unknown. The uncertainties regarding hearing among infants prevent practitioners from determining a best practice standard for objective calculation of sensitivity and specificity.

Secondly, few articles distinguished between results relating to low-risk and high-risk newborn. Only four studies[29,35~37)] distinguished between high-risk and low-risk newborn within the samples, but the sample sizes of the high-risk newborn in three of them[35~37)] was small. With Mimaki et al.[29)], 203 newborn had high risk factors and four of them were identified as having severe-to-profound CHL. Selective screening of newborn that have risk factors as listed by the Joint Committee on Infant Hearing 1994[42)] has been considered as an alternative to UNHS[43)]. However, the scarcity of information on data relating to high-risk newborn prevents analysis to determine the effectiveness of this strategy.

IV. Effectiveness of Early Detection

In other countries, several cohort studies which didnot comply with guidelines rating 'good' quality according to USPSTF[38)], nevertheless found improvements in language and communication with early detection[38,44]. In the Japanese literature, no studies have been published on effectiveness of early detection and only two on effectiveness of early intervention, both by Uchiyama et al. in 2000[45] and in 2004[46].

Uchiyama et al. evaluated the effectiveness of an auditory-oral intervention program using the Wechsler Preschool and Primary Scale of Intelligence (WPPSI) Test. Data analysis revealed that verbal IQ correlated closely with performance IQ and the time of intervention inception. They concluded that 50 of the 72 hearing-impaired children were able to acquire a language ability comparable to that of normal hearing children of similar age (verbal IQ≧80) if they completed an early intervention program.

In the later study, the same authors evaluated the effectiveness of early intervention in hearing-impaired children under 12 months of age using the WPPSI Test and at six years of age using the Wechsler Intelligence Scale for Children Third Edition (WISC III) (range: 7～15 years). They divided 39 congenital hearing-impaired children into three groups according to the age at entry into the treatment: a group of six early-identified children (five～eleven months), 19 intermediate group children (15～22 months), and 14 late-identified children (24～34 months). They concluded that the results of their study provided clear evidence of the benefits of early intervention on the development of language ability in hearing-impaired children under 12 months of age.

Both of these studies had similar limitations. First, there was considerable sampling bias. All children were sampled from one private institution, but in Japan public schools for the deaf play an important role in early intervention and it has also been suggested that the quality of intervention for hearing-impaired children varies largely among institutions[47]. Second, the length of intervention was not controlled. It is difficult to determine whether the documented differences in IQ were influenced by the start-age of the intervention or by the length of the intervention. Third, early intervention was only evaluated using an auditory-oral intervention program. Sign-language-based interventions were not taken into consideration. Given these limitations, it is not necessarily a simple matter to conclude that early intervention is effective.

Regarding future research on the effectiveness of early detection and intervention, further discussion is needed on the variables appropriate for measurement of development outcomes for children with CHL. While a number of foreign studies have attempted to address this question[6,32,33,48～52], evaluation studies in Japan have simply assessed development regarding communication ability, intelligence level, or developmental quotient. These variables do not assess the wider aspects of child development. The effectiveness of early detection needs to be assessed from a wider point of view and should also include research on psychological consequences, the extent of social activities, lifestyle in adulthood, and so on.

In addition, the adverse effects of early detection, especially parental attitudes to false-positive results should be taken into consideration. A number of studies[19～24] regarding unrelated newborn screening tests have shown that false-positive results can lead to a lasting emotional impact on the mother and infant relationship. Such an impact has also been suggested in a few studies on newborn hearing screening[53], but this question has not been addressed in Japan.

Although a number of difficulties exist in evaluation of UNHS, the necessity to make decision based on evidence is clear. In July of 2003, after 18 years of implementation, the MHLW recommended the cessation of universal infant screening for neuroblastoma on the grounds that the evidence of reduced mortality with early detection was poor[54,55]. We could argue that the same situation could occur with UNHS and that it is undesirable or too early to implement UNHS nationally in Japan. While the parents of children with CHL have voiced hopes for early detection and intervention of CHL, screening tests should not be systematically implemented without evidence of effectiveness.

V. Conclusion

The utility of UNHS in Japan is still inconclusive because of the lack of best practice standards to assess the clinical markers for distinction between impaired and normal hearing. Our results highlight the need for assessment of the accuracy of screening and the effectiveness of early detection before nationwide implementation of UNHS with the findings discussed amongst pediatricians, otorhinolaryngologists, obstetricians, speech therapists, teachers, parents and the deaf themselves.

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