Newborn hearing screening in Western Australia

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ABSTRACT

Aim: To report the preliminary findings of a pilot program to screen newborn babies for congenital bilateral permanent hearing loss.

Setting: The five largest maternity hospitals in Perth, Western Australia. Screening was gradually introduced over seven months from February to August 2000.

Participants: All babies born at these hospitals after the introduction of hearing screening until 30 June 2001.

Methods: One or both of two automated screening devices were used: one measuring transient evoked otoacoustic emissions (TEOAE) and the other automated auditory brainstem responses (AABR). If a "pass" was not obtained in both ears, screening was repeated. All babies who did not obtain a pass in either ear at follow-up were referred for audiological assessment.

Main outcome measures: Prevalence of permanent bilateral hearing loss.

Results: Of 13 214 eligible babies, 12 708 (96.2%) received screening. The main reason for missing screening was early hospital discharge (309; 2.3%). Of the screened babies, 99% had a pass response in both ears at either the initial or follow-up screen. Twenty-three babies were referred for audiological assessment, and nine were diagnosed with bilateral permanent hearing loss (0.68/1000; 95% CI, 0.31–1.28).

Conclusions: Despite our program meeting process quality indicators, our detection rate was low. Before extending the program to smaller hospitals, we need to validate our screening instruments and put in place a system to monitor false negative results.

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CONGENITAL BILATERAL permanent hearing loss has a major impact on speech and language development.[1] Estimates of the prevalence of such hearing loss range from 0.53–1.50 per 1000 live births[2-6] (Appendix 1, *page 185*). Studies have shown that children with hearing loss who receive early intervention have better language skills than those with later intervention.[1,7] The critical age by which intervention should commence may be as early as six months.[1] However, diagnosis is often delayed. In 1997, in Western Australia, the median age for fitting of hearing aids for congenital hearing loss was 25 months.[8]

Two approaches to newborn hearing screening have been used — targeted screening of babies with risk factors using criteria such as those of the Joint Committee on Infant Hearing 2000[9] (Appendix 2, *page 185*), or universal screening of all newborns. As only about 60% of children with congenital hearing loss can be identified by using high-risk criteria,[2,3,6] universal hearing screening has been recommended in the United States[9] and the United Kingdom.[10]

We report the preliminary findings of a pilot program of newborn hearing screening to detect congenital bilateral permanent hearing loss at five maternity hospitals in Perth, Western Australia. The program commenced in 2000 and our report includes results in babies born until 30 June 2001.

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METHODS

Participating hospitals

In Western Australia, about 25 000 babies are delivered every year; about 45% of these deliveries occur in the five largest maternity hospitals in the Perth metropolitan area. Hearing screening was gradually introduced at these hospitals during 2000: King Edward Memorial Hospital for Women (February), St John of God Health Care Subiaco and Woodside Maternity Hospital (May), and Joondalup Health Campus and Osborne Park Hospital (August). King Edward Memorial Hospital has a Level 3 nursery and St John of God Health Care and Joondalup Health Campus have Level 2 nurseries. In June 2001, hearing screening for all babies was introduced at the Level 3 nursery at Princess Margaret Hospital for Children (Western Australia's tertiary paediatric centre). Thus, newborn hearing screening was available at all Level 2 and 3 nurseries in Western Australia. Screening was offered at King Edward Memorial Hospital seven days a week, and at the other hospitals five to six days a week.

Hearing screeners

The hearing screeners were trained to use the screening equipment. They came from a variety of backgrounds (including qualifications in childcare or a health-related field) and had previous

1: Hearing loss screening instruments and flow diagram for newborn screening for well babies

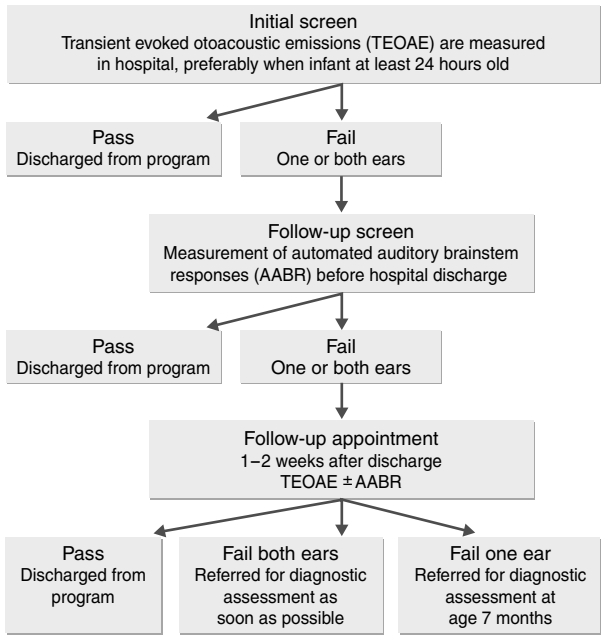
Measurement of transient evoked otoacoustic emissions (TEOAEs)

All well babies were screened using an Echocheck TEOAE hand-held screener (Otodynamics, Hatfield, UK), which involves placing a probe into the baby's outer ear. This test concentrates on the main speech frequency band range of 1.6–3.6 Hz. A "pass" response is based on detecting a non-linear TEOAE cross-correlated signal-to-noise ratio of at least 6 decibels (dB).

Automated auditory brainstem responses (AABR)

The detection of AABR is more specific, but the test takes more time and is more expensive than the TEOAE screen, requiring the placement of scalp electrodes. Until July 2000 the AABR instrument used was the Sabre system (SLE Ltd, South Croydon, UK). After July, the Algo 2e Color Newborn Hearing Screener (Natus Medical Inc, San Carlos, USA) was used. Auditory brainstem response is a modified electroencephalogram recording of brain activity in response to auditory stimuli presented in the form of brief clicks. By averaging techniques, the electrical potential can be detected and used to determine the hearing threshold. Both the Sabre and Algo 2e screening units use an automated detection of these responses and yield "pass"/"refer" criteria set at 35 dB normal hearing level scale.

Comments: Both the TEOAE and AABR screening instruments clearly display a "pass" response, so no interpretation is required. The TEOAE screen has limited specificity when used in the first few days of life. It only measures outer hair-cell function in the cochlea. Babies with central hearing loss can have normal cochlear function and so will pass the TEOAE screen. As the group most at risk of central hearing loss are babies who have received Level 3 care, most babies who have had long-term neonatal intensive care are screened initially using the AABR screen.



experience in handling babies. They were given a complete list of births from the previous day(s) and obtained written consent to perform the screening test from the parents. They explained the test and its results to parents, performed the screen, and recorded the results. An audiologist was available for consultation.

Screening protocol

All liveborn infants at the screening hospitals were eligible for screening, which was offered to well babies generally on the day after delivery. Babies admitted to a Level 2 or 3 nursery were screened when the baby was at least 34 weeks' gestation. If possible, the parents of babies who were discharged before screening were given an invitation by the visiting midwife to return for an outpatient screen.

We developed a combined data and consent form, and recorded for all babies (whether screened or not) delivery site, date and time of birth, and sex. If the baby was not screened, the reason for not screening was recorded. For screened babies, demographic information, birthweight, gestational age and screening details were collected.

We used a combined-technology screen to increase specificity and reduce costs. The well-baby screening flow diagram and a description of the two screening instruments used are given in Box 1. Babies with a significant family history of hearing loss who passed the hearing screen were referred for audiological assessment at age seven months.

After the hearing screen parents were given a results sheet which explained the results and reinforced the need for ongoing childhood hearing surveillance. The results of the screen were recorded in the baby's hospital records and personal health record.

Ethical approval

Approval for the program was obtained from the ethics committees at King Edward Memorial and Princess Margaret hospitals.

Data analysis

Completed data were entered into a Filemaker database.[11] We calculated 95% CIs for the prevalence of bilateral hearing loss using the Poisson distribution.[12]

RESULTS

There were 13 214 eligible babies. Of these, 12 708 (96.2%) were screened. The proportion of babies screened increased with time from 93.5% of those born before July 2000 to 97.1% of those born in the first six months of 2001 (Box 2). About 18% of babies required Level 2 or 3 nursery care.

Of the babies screened at Princess Margaret Hospital from the start of its newborn screening program in June 2001, only those transferred from a hospital where screening was available were included in the data analysis.

About 80% of all babies screened had measurement of transient evoked otoacoustic emissions (TEOAE) only, while about 16% also had automated auditory brainstem responses (AABR) measured (Box 1). Most of the 4% of babies screened using only AABR were in Level 2 or 3 nurseries.

Reasons for babies not being screened

Early discharge (309; 2.3%) was the main reason that babies were not screened (Box 2). By 2001, more women took up the offer of an outpatient screen for their infant, and the

proportion who missed screening decreased to 1.8%. Of the 129 (1.0%) babies not screened because of transfer to another hospital, 69 were screened as part of a high-risk screening program in the Level 3 nursery at Princess Margaret Hospital. All babies in the high-risk program were screened by an audiologist before hospital discharge. (Information was available on the outcome for these 69 babies only. They are included in the calculation of prevalence of hearing loss in the cohort, but not in the evaluation of the screening process.)

Initial hearing screening

Box 2 shows the results of the initial screening. The bilateral "pass" rate increased over time. A total of 351 babies failed the screen in one or both ears. In the first weeks of the program, an AABR instrument was not available, so babies were screened using only TEOAE, and follow-up was only offered to babies who failed the screen in both ears. Thus, 40 babies who failed the screen in one ear were not offered follow-up. Another 10 babies were referred directly for audiological assessment due to either repeated fail responses to hearing screening while in a Level 2 or 3 nursery, or a structural problem affecting the ears. Of the remaining 301 babies who were offered follow-up, 271 (90%) returned and 30 (10%) babies were lost to follow-up due to multiple missed appointments or parental refusal. When families did not attend the follow-up appointment, their child health nurse was notified and asked to encourage the family to arrange a repeat screen, or, failing this, to monitor the baby's hearing.

Follow-up screening

Of the 271 babies who attended for follow-up screening, 258 (95.2%) passed in both ears, 10 (3.7%) failed in one ear, and three (1.1%) failed in both ears. Therefore, 12 615 (99.3%) of the 12 708 babies screened passed either the initial or follow-up screen. Including those who were referred directly, a total of 23 babies were referred for diagnostic assessment because of failure in one or both ears. This is a referral rate of 1.81 per 1000 babies screened.

2: Uptake and results of the initial hearing screen and reasons initial screening was not completed

	Number (%) babies born			
	Before 01/07/00	01/07/00–31/12/00	01/01/01–30/06/01	Total
Uptake of screening*				
Eligible for screening	2513 (100%)	5325 (100%)	5376 (100%)	13 214 (100%)
Not screened	165 (6.6%)	186 (3.5%)	155 (2.9%)	506 (3.8%)
Discharged before screen	107 (4.3%)	103 (1.9%)	99 (1.8%)	309 (2.3%)
Transferred†	37 (1.5%)	53 (1.0%)	39 (0.7%)	129 (1.0%)
Refused screening	14 (0.6%)	23 (0.4%)	14 (0.3%)	51 (0.4%)
Missed screening‡	7 (0.3%)	7 (0.1%)	3 (0.1%)	17 (0.1%)
Screen completed	2348 (93.4%)	5139 (96.5%)	5221 (97.1%)	12 708 (96.2%)
Results of screening§				
"Pass" both ears	2200 (93.7%)	5027 (97.8%)	5130 (98.3%)	12 357 (97.2%)
"Pass" one ear	110 (4.7%)	72 (1.4%)	64 (1.2%)	246 (1.9%)
"Fail" both ears	38 (1.6%)	40 (0.8%)	27 (0.5%)	105 (0.8%)

* Excludes babies who died before screening.

† 69 of these babies, who were transferred to Princess Margaret Hospital, were screened as part of a high-risk hearing screening program at that hospital.

‡ Mostly babies who did not appear on the daily list of deliveries, and by the time the omission was discovered the local visiting midwife was no longer in contact with the family.

§ The denominator for these calculations was the number of babies screened.

Prevalence of hearing loss

- Of the babies referred after follow-up screening, five were diagnosed with bilateral hearing loss by the age of three months and three had unilateral loss.
- Of the 69 babies screened as part of the high-risk program at Princess Margaret Hospital, four were diagnosed with bilateral hearing loss.

Thus, there were nine cases of permanent bilateral hearing loss (> 35 dB in the better ear) diagnosed in this cohort of babies, making the prevalence of bilateral permanent hearing loss 0.68 per 1000 eligible babies (95% CI, 0.31–1.28). Eight of these nine babies had at least one hearing-loss risk factor[9] (Appendix 2, *page 185*), including five who had received Level 2 or 3 nursery care. The rate of bilateral permanent hearing loss in well babies was 0.37 per 1000 (95% CI, 0.10–0.92). Two of the four well babies had a family history of hearing loss.

Of the nine babies with permanent bilateral hearing loss, six were fitted with hearing aids by the age of six months, and one was fitted with aids at 19 months because of parental delays. One baby who received long-term Level 3 nursery care is still in hospital and his condition has precluded fitting a hearing aid, and one baby has died.

DISCUSSION

During the first 17 months of this program, the prevalence of congenital bilateral hearing loss was 0.68 per 1000, with eight out of nine affected babies having a risk factor for hearing loss. A summary of other hearing screening programs is given in Box 3. However, the different definitions of hearing loss used make comparison between studies difficult.

Our program meets the recommendations of the Joint Committee on Infant Hearing[9] for process quality indicators, with 96.2% screening uptake, 90% return for follow-up and 0.18% referral for audiological assessment. However, our study has revealed a number of weaknesses: (i) the confidence intervals for the prevalence of bilateral hearing loss were wide; (ii) our cohort is not representative of all Western Australian births; (iii) the screening instrument we used has not been well validated in the medical literature; and (iv) we had lim-

3: Summary of the methods and results of published newborn hearing screening programs*

First author, country, years data collected	Definition of bilateral hearing loss	Population		No. of cases/ population	Rate per 1000 (95% CI)	Proportion with risk factors	Rate per 1000 well babies† (95% CI)
Barsky-Firsker,[13] USA, 1993–1995	Sensorineural, > 35–40 dB	One hospital with NICU; about 5000 births a year		46/15 749	2.92 (2.14–3.89)		2.07‡ (1.35–2.88)
Chapchap,[14] Brazil, 1996–1999	All, moderate or greater	One hospital with NICU; about 1500 births a year		7/4 196	1.67 (0.67–3.38)	71%	
Clemens,[15] USA, 1998–1999,	Permanent, > 35 dB§	One hospital; report of well baby screening only		6/5 010	1.20 (0.44–2.55)	67%	1.20 (0.44–2.55)
Clemens,[16] USA, 1999–2000	Permanent, > 35 dB§	One hospital; report of well baby screening only		2/3 142	0.64 (0.07–2.16)	38%‡	0.64 (0.07–2.16)
Dalzell,[17] Prieve,[18] USA, 1995–1996	Permanent, > 20 dB	Eight hospitals, all with NICUs; 2435–5474 births a year		49/43 311	1.13 (0.84–1.49)	80%	
Finitzo,[19] USA, 1996	Permanent, requiring intervention	Eleven sites; < 300–4540 births a year; four NICUs		32/17 105	1.87 (1.28–2.64)	47%‡	1.26‡ (0.77–1.93)
Isaacson,[20] USA, 1998–1999	⩾ 25 dB	One hospital with NICU; about 1000 births a year		6/2 031	2.95 (1.08–6.3)		
Kanne,[21] USA, 1995–1996	Permanent	One hospital with NICU; about 2000 births a year		1/2 289	0.44 (0.02–2.17)	100%	
Lim,[22] USA, 1999	> 35 dB§	46 sites in 11 States; no further information given		57/66 292	0.86 (0.65–1.11)	40%‡	
Mason,[23] USA, 1992–1997	Permanent, > 35 dB	One hospital with NICU; about 2000 births a year		15/10 372	1.45 (0.81–2.37)		0.89 (0.38–1.73)
Mehl,[24] USA, 1992–1996	Sensorineural	26/52 hospitals (40–3500 births a year); 60% of State's births		75/41 796	1.79 (1.41–2.25)	50%‡¶	
Sergi,[25] Italy, 1997–?		One hospital	Well babies	1/5 650	0.18 (0.01–0.88)		0.18 (0.01–0.88)
			NICU, no risk	3/749	4.01 (0.81–11.2)		
			At risk	14/118	118.64 (64.97–197.74)		
Stewart,[26] USA, 1996–1997	Sensorineural, > 35 dB§	Five sites; one NICU; two Level 2 nurseries		21/11 711	1.79 (1.11–2.73)		
Watkin,[27] UK, 1992–1995	Permanent, > 40 dB	District; 3500 births a year		23/11 606	1.98 (1.26–2.96)		
Watkin,[28] UK, 1992–1997	Permanent, > 40 dB	District; 3500 births a year		34/25 199	1.35 (0.93–1.88)		
Wessex Group,[29] UK, 1993–1996	Permanent, ⩾ 40 dB	Four hospitals; four SCN; 3300–5600 births a year		27/25 609**	1.05 (0.7–1.53)	74%	0.84†† (0.48–1.36)
White,[30] USA, 1990–1991	Sensorineural, > 25 dB	One hospital with NICU		6/1 850	3.24 (1.19–6.92)	83%	1.29 (0.15–4.38)

* Some values have been calculated from the data provided in the studies. Where possible, all cases of temporary or unilateral hearing loss have been excluded.
† Well babies are those who were not admitted to a neonatal intensive care unit (NICU) or a special care nursery (SCN).
‡ It was not possible to separate out cases of bilateral hearing loss from cases of unilateral and bilateral hearing loss from this calculation.
§ A definition of hearing loss was not specifically defined. As an AABR with a pass response of 35 dB normal hearing level was used, > 35 dB was assumed to be the definition of hearing loss.
¶ It was not possible to separate cases of permanent hearing loss from the total number of cases of temporary and permanent hearing loss in this calculation.
** The rate was calculated from the total population of babies born in the time period, not the screened population.
†† In the well group, the rate for those with a risk factor was 0.99/1000. In the well group without risk factors the rate was 0.28/1000.

ited time for diagnosis of hearing loss in babies who passed the hearing screening ("false negatives").

Wide confidence intervals

Despite our large sample size, the 95% CIs for the prevalence of bilateral congenital hearing loss are wide (0.31–1.28). However, most other screening programs have equally wide CIs (Box 3). There is a strong case to continue screening, so that, with a larger sample, there will a more precise estimate of prevalence.

Unrepresentative cohort

In our cohort of babies those requiring special care are over-represented, as screening is offered at all delivery sites with Level 2 or 3 nursery facilities. Since these babies are more likely to have hearing loss, the prevalence we calculated is likely be higher than in the State as a whole. There are few reports of screening programs covering the whole population of a defined geographical area.[6,27,28]

Echocheck screener not well validated

In most reports, the instrument used to measure TEOAE was the Otoacoustic Analyzer ILO 88 (Otodynamics, Hatfield, UK) on which the Echocheck screener is based. We plan to conduct a comparison study using both the Echocheck and the Otoacoustic Analyzer ILO 88.

CONCLUSIONS

We have detected a prevalence of bilateral congenital hearing loss that is at the lower end of those found in population-based studies (Appendix 1, *page 185*). We found that eight of the nine babies with bilateral hearing loss had a risk factor for hearing loss, which is a higher proportion than others have reported.[2,3,6] Little is known about the prevalence and aetiology of newborn hearing loss in Western Australia. According to the report of the Birth Defects Registry of Western Australia, 1980–2000,[31] the prevalence of congenital deafness in Western Australia was 0.7 per 1000 births between 1980 and 1994, although under-reporting is suspected. By manipulating the data provided by Australian Hearing,[32] the hearing-aid fitting rate for all causes of hearing loss greater than 30 dB in Western Australian children born in 1996 was 0.95 per 1000. However, from the information provided, the proportion of hearing loss thought to be congenital in origin cannot be calculated. To monitor both for false negative results and provide population-based prevalence data for Western Australia, a database is being set up to record all Western Australian children born in 1999 and later who are diagnosed with bilateral hearing loss before the age of five years.

Our program has been successful in screening a high percentage of eligible babies, with a low referral rate. However, the prevalence of bilateral permanent hearing loss detected is low (0.68/1000). Several aspects of the program are being evaluated and data about permanent hearing loss in Western Australian children will be obtained before deciding whether to extend the program to other hospitals.

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COMPETING INTERESTS

None identified. The Department of Health, Western Australia, and King Edward Memorial and Princess Margaret hospitals were involved in the planning of the pilot program. The funding bodies had no input into the analysis or interpretation of the data, the writing of the article or the decision to submit for publication.

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Appendix 1: Summary of population-based studies* on the prevalence of congenital, permanent bilateral hearing loss

Study	Definition of bilateral hearing loss	Characteristics of population	No. of cases/ population	Rate per 1000 (95% CI)	Proportion with risk factors	Rate per 1000 well babies† (95% CI)
Fortnum and Davis[2] 1997	⩾ 40 dB, permanent	Born 1985–1990, living in Trent region, UK, in 1994–1995	487/366 480	1.33 (1.21–1.45)		
	⩾ 40 dB, congenital	16% of hearing loss thought to be acquired later	409/366 480	1.12 (1.01–1.23)	58.9%	0.84‡ (0.75–0.95)
Parving[3] 1993	⩾ 25 dB, requiring hearing aid	Born 1980–1990, living in Copenhagen city or county in 1992. Those with hearing loss had a hearing aid fitted by January 1992	181/95 912	1.89 (1.62–2.18)		
	⩾ 25 dB, congenital/ early acquired	20% of hearing loss thought to be acquired later	144/95 912	1.50 (1.27–1.77)	59.7%	
Van Naarden et al[4] 1999	⩾ 40 dB, permanent	Born 1981–1990, living in five Atlanta counties, USA, in 1991–1993	862/790 200§	1.09 (1.02–1.17)		
	⩾ 40 dB, congenital (sensorineural cases with no postneonatal event recorded)	50% of sensorineural hearing loss thought to be acquired later	173/324 327¶	0.53 (0.46–0.62)		
Vartiainen et al[5] 1997	> 25 dB, sensorineural	Born 1974–1987 in Kuopio region, Finland	98/46 240	2.12 (1.72–2.58)		
	> 25 dB, congenital	34% of hearing loss > 25 dB thought to be acquired later	65/46 240	1.41 (1.08–1.79)		
	> 40 dB, sensorineural		52/46 240	1.12 (0.84–1.47)		
	> 40 dB, congenital	17% of hearing loss > 40 dB thought to be acquired later	41/46 240	0.89 (0.67–1.20)		
Vohr et al[6] 1998	> 30 dB, permanent hearing loss, all presumed congenital	Born 1993–1996, Rhode Island, USA, population-based screening program, no mention of later acquired hearing loss	79/53 121	1.49 (1.18–1.85)	60%**††	1.27** (0.97–1.63)

* Some values have been calculated from the data provided in the reports.
† Well babies are those who were not admitted to a neonatal intensive care unit.
‡ Excluding those with a family history of hearing loss, the rate of bilateral hearing loss in well babies was 0.54/1000.
§ Cross-sectional analyses using the sum of the annual total number of 3–10-year-olds living in area, 1991–1993, as the denominator.
¶ Birth-cohort analyses using total livebirths in the area between 1981 and 1990 as the denominator.
** It was not possible to separate out babies with bilateral hearing loss from those with unilateral and bilateral hearing loss for this calculation.
†† Excluding those in whom the family history was noted only after diagnosis of hearing loss, 50% had a known risk factor.

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Appendix 2: Indicators associated with sensorineural and/or conductive hearing loss for use in neonates when universal screening is not available*

- An illness or condition requiring admission of 48 hours or more to a neonatal intensive care unit
- Stigmata or other findings associated with a syndrome known to include a sensorineural and/or conductive hearing loss.
- Family history of permanent childhood sensorineural hearing loss.
- Craniofacial anomalies, including those with morphological abnormalities of the pinna and ear canal.
- In-utero infection, such as cytomegalovirus, herpes and toxoplasmosis, or rubella.

* Joint Committee on Infant Hearing.[9]