PREVALENCE: NEWBORN PERIOD

REFERENCE	DESIGN	RECRUIT-MENT	CASE DEFINITION	SUBJECTS	PREVELANCE	AUTHORS' CONCLUSIONS
Dalzell L, Orlando M, MacDonald M, Berg A, Bradley M, Cacace A, Campbell D, DeCristofaro J, Gravel J, Greenberg E, Gross S, Pinheiro J, Regan J, Spivak L, Stevens F, Prieve B. 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; 21:118–130.	UNHS* was conducted for all infants in the NICUs* and WBNs*. Diagnostic evaluations were performed for infants who did not pass the screening. Ages at hearing loss identification, hearing aid fitting, and enrollment in EI* were investigated regarding nursery type, risk status, unilateral versus bilateral hearing loss, type of loss, loss severity, and state regions.	8 hospitals in the New York State. Screening: TEOAE* or ABR*, or both.	Hearing loss defined as >20dB* at any frequency from 500 to 4,000 Hz*. Severity was reported for the better hearing ear for bilateral losses and for the impaired ear for unilateral losses. Sensorineural, mixed, or structurally conductive hearing losses were covered.	All newborns at 8 hospitals in New York State who did not pass hearing screening in both ears before discharge. (1995 and 1996)	Bilateral and unilateral prevalence: 85/43,3115 = 2/1,000 Bilateral prevalence: 49/43,311 = 1.1/1,000 Unilateral prevalence: 36/43,311 = .8/1,000 Of the 85 infants with hearing loss, 61% were from NICUs and 67% were at risk for hearing loss.	Identification of hearing loss at early ages, hearing aid fitting, and enrollment in early intervention can be achieved for infants from NICUs and WBNs and for infants at risk and not at risk for hearing loss. If only high-risk infants were screened, many infants with hearing loss would be missed. NICU infants were typically diagnosed and fitted with hearing aids later than WBN babies. Median age at identification and enrollment in EI was 3 months.

*UNHS = universal newborn hearing screening; NICU = neonatal intensive care unit; WBN = well-baby nursery; EI = early intervention; TEOAE = transient evoked otoacoustic emissions; ABR = auditory brainstem response; dB = decibel; Hz = hertz

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De Capua B, Costantini D, Martufi C, Latini G, Gentile M, De Felice C: Universal neonatal hearing screening: The Siena (Italy) experience on 19,700 newborns. Early Hum Dev. 2007;83(9):601-6.	Aim was to test the value of a universal screening protocol, based on a two-stage strategy. All infants were screened with two-stage strategy of TEOAE*. This was followed by diagnostic ABR* for infants who did not pass the TEOAE. Additionally, all infants who met JCIH* 2004 risk criteria received a diagnostic ABR.	All hospital live births in Siena, Italy.	For screening: all live births. For diagnostic evaluation: infants who failed screening, plus high-risk infants.	19,700 infants (of 21,125 livebirths) in Siena, were tested during the period Apr 1, 1998–Jul 31, 2006.	Overall prevalence of hearing loss was 1.78/1,000 (35/19,700). For bilateral loss: 1.42/1,000 (28/19,700). For low-risk infants: 0.43/1,000 (14.9/19,700) For high-risk infants: 14.88/1,000 (20/1344)	The epidemiology of congenital hearing loss widely justifies universal newborn hearing screening. A two-stage TEOAE and diagnostic ABR screening for congenital hearing loss is feasible, minimally invasive, and accurate in the early detection of congenital hearing loss. A congenital hearing loss screening strategy based exclusively on the use of TEOAE should always consider the possibility of false-negative cases.

*TEOAE = transient evoked otoacoustic emissions; ABR = auditory brainstem response; JCIH = Joint Committee on Infant Hearing.

REFERENCE	DESIGN	RECRUIT-MENT	CASE DEFINITION	SUBJECTS	PREVALENCE	AUTHORS' CONCLUSIONS
Flynn M, Austin N, Flynn TS, Ford R, and Buckland L: Universal Newborn Hearing Screening introduced to NICU infants in Canterbury Province, New Zealand. The New Zealand Medical Journal. 2004: 117(1206).	UNHS* was conducted for NICU* infants over a 12-month period. The purpose was to assess the feasibility of introducing UNHS over the 12-month period. If a baby failed initial screening, a rescreening was conducted before the baby was discharged. Testing was conducted once the baby was 34 weeks GA* and older than 48 hours.	Parents of all babies admitted from mid-July 2001 through mid-July 2003 to the NICU at Christchurch Women's Hospital were offered the opportunity to have their baby's hearing screened using the GSI-70 otoacoustic screener.	Infants who were admitted to the NICU and remained there at least 48 hours.	435 (64%) of 688 NICU infants born at Christchurch Women's Hospital were eligible by meeting the 48-hour stay required.	2 of 435 (4.6/1,000) babies had moderate-to-severe bilateral sensorineural hearing loss. Only one of the babies had risk factors for hearing loss.	UNHS was cost-effective and implemented efficiently and cost-effectively within the neonatal services of a New Zealand hospital. The study shows the benefits of UNHS against "at-risk" screening 27.1% of the babies screened had one or more risk factors. The positive predictive value of UNHS (10.53%) was significantly greater than that of "at-risk" (0.85%) screening.

*UNHS = universal newborn hearing screening; NICU = neonatal intensive care unit; GA = gestational age

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Hille ETM, van Straaten HLM, Verkerk, PH: Prevalence and independent risk factors for hearing loss in NICU infants. Acta Paediatr. 2007;96:1155-8.	Aim was to determine the prevalence and independent relationship between hearing loss and risk factors among a NICU* population. All infants meeting criteria were screened with AABR*. A second AABR was given to those who failed the first. Those who failed both were given a diagnostic ABR*. Screeners recorded the 2004 JCIH* risk factors on the screening form.	A nationwide sample of NICU infants meeting criteria from 11 NICUs in the Netherlands.	NICU infants: GA* <30 weeks, or BW* <1,000 grams, or both; born during the period Oct 1, 1998–Jan 1, 2002. Infants who died before 3 months of age were excluded.	2,186 Dutch NICU infants. Mean GA was 28.5 weeks. Mean BW was 1,039 grams.	Overall prevalence of unilateral or bilateral hearing loss was 3.2 % (71/2,186). Prevalence for infants with neither severe birth asphyxia nor assisted ventilation for ≥5 days was 1.3%. Prevalence for infants with severe birth asphyxia and assisted ventilation ≥5 days was 7.8%.	Severe birth asphyxia and assisted ventilation for ≥5 day are independent risk factors for hearing loss among infants born with a GA <30 weeks or a birthweight <1,000 grams, or both.

*NICU = neonatal intensive care unit; AABR = automated auditory brainstem response; ABR = auditory brainstem response; JCIH = Joint Committee on Infant Hearing; GA = gestational age; BW = birthweight.

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Lévêque M, Schmidt P, Leroux B, Danvin JB, Langagne T, Labrousse M, Chays A: Universal newborn hearing screening: a 27-month experience in the French region of Champagne-Ardenne. Acta Paediatr. 2007;96:1150-4.	Purpose was to report the results of the first 27 months of a UNHS* program in France. TEOAE* was used for the initial screen for infants in the WBN*; AABR* was used for NICU* infants. Infants who failed the first screen in both ears were rescreened with TEOAE or AABR 15 days after discharge. Infants who failed the second screen in both ears were referred for diagnostic testing.	Universal screening of all infants.	Infants born in the Champagne–Ardenne region of France during the period Jan 2004–Mar 2006.	36,652 infants were born in the region. 33,873 (92.4%) infants received the initial screen. 33,433 (98.7%) had negative bilateral or unilateral first screens. 440 (1.3%) had positive bilateral first screens 11/440 were lost to follow-up. 4 families refused screening; 5 families moved out of the region; 2 infants died. 429 infants had a second screening. 395/429 infants who were retested were negative for hearing loss in one or both ears. 34 were referred to diagnostics.	Unilateral losses were excluded. 27/34 (0.08% of all infants screened) had hearing loss. Mean age at diagnosis was 10 weeks.	The UNHS* program demonstrated its validity and feasibility.

*TEOAE = transient evoked otoacoustic emissions; WBN = well-baby nursery; AABR = automated auditory brainstem response; NICU = neonatal intensive care unit; UNHS = universal newborn hearing screening.

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Mehl A, Thomson V: Newborn hearing screening: The great omission. Pediatrics. 1998;101(1) e4.	Universal screening was implemented in 26 of Colorado's 52 hospitals. This study was designed to assess the feasibility, accuracy, and cost-effectiveness of a hospital-based hearing screening program. Screening methods: 19 hospitals primarily used A-ABR* 1 hospital use OAE* 6 used conventional ABR* Hospitals reported all screening results to the Colorado State Newborn Hearing Screening Project.	All infants who were screened in Colorado from 1992 through 1996.	All infants born in hospitals with UNHS*.	41,796 screened babies. 2,709 (6.5%) failed initial screening. 1,296 completed diagnostic evaluation (52% lost to the system).	126 (3/1,000) infants were identified with sensorineural or conductive hearing loss. 94 (2.2/1,000) were identified with sensorineural loss. 75 (59.5%) had bilateral sensorineural loss. 19 (15.1%) had unilateral sensorineural loss. 32 (25.4%) had conductive hearing loss; 13 had structural or ossicular malformations; and 19 had persistent neonatal middle ear effusions.	Since the screening project began, 17 children have been identified with hearing loss by 18 or more months of age. All were born at non-UNHS hospitals 50% of children identified with hearing loss had no risk factors. Cost of screening: $18.30 when screened by volunteers; $25.60 when screened by technician; and $33.30 when screened by an audiologist.

*A-ABR = automated auditory brainstem response; OAE = otoacoustic emissions; ABR = auditory brainstem response; UNHS = universal newborn hearing screening

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Nagapoornima P, Ramesh A, Srilakshm, Rao S, Patricia PL, Gore M, Dominic M, Swarnarekha: Universal Hearing Screening. Indian J Pediatr. 2007;74:545-9.	Purpose was to determine the weighted incidence of hearing loss in a standardized population of at-risk and not-at-risk infants seeking care at a tertiary level hospital in India. Prospective study of a nonrandomized cohort. From Sep 2002–Sep 2004 screening was done once a week for only high-risk infants using automated equipment Echo screen. Beginning in Sept,. 2004, all infants were screened on all working days using OAE*. ABR* and BOA* were used to confirm hearing loss if infant failed the OAE screen 2 times. Follow-up was done using REELS* and BOA.	Infants born at St. John's Medical College hospital, Bangalore, India, during the period Sep..1, 2002–Mar 31, 2006.	All infants screened.	8,192 infants were born during the recruitment period. 1,769 infants were screened. Not at risk: 1,490. At risk: 279 Risk criteria were adapted from the JCIH* 2000 position statement.	All infants screened: 5.65/1,000 (10/1,769) Not-at-risk infants: 4.70/1,000 (7/1,490) At-risk infants: 10.75/1,000 (3/279) Of the 3 at-risk infants with hearing loss, 2 had a family history of childhood onset sensorineural loss and 1 had severe birth asphyxia.	A high incidence of hearing loss of 5.60/1,000 among a standardized neonatal population of at-risk and not-at–risk infants warrants the urgent implementation of universal newborn hearing screening. Screening only at-risk infants can miss up to 70% of all infants with hearing loss in a typical tertiary care hospital.

*OAE = otoacoustic emissions; ABR = auditory brainstem response; BOA = behavioral observation audiometry; REELS = Receptive Expressive Emergent Language Scale; JCIH = Joint Committee on Infant Hearing.

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Vohr, B: The Rhode Island Hearing Assessment Program. Rhode Island Medicine. 1995;78:11–13.	Demonstration project: This was a demonstration project to assess the feasibility of using TEOAE* as a screener. Infants were screened with TEOAE and confirmed with BAER* and Behavioral Audiometry. Statewide screening: Success of demonstration project helped to influence the July 1993 amendment to mandate universal screening. Protocol was: TEOAE before discharge. If infant did not pass, screening was repeated 4–6 weeks after discharge. Infant who failed rescreening and screening BAER was referred for diagnostic BEAR or BOA at 6–8 months of age.	Demonstration project: All infants born at Women and Infants Hospital in Rhode Island. Statewide screening: Infants born in the 8 Rhode Island hospitals that had completed staff training.	Demonstration project: All infants born at Women and Infants Hospital in Rhode Island. Statewide screening: All infants born in Rhode Island's 8 maternity hospitals since the July 1993 mandate.	Demonstration project: 3,303 infants born 8/15/90–12/22/91 at Women and Infants Hospital in Rhode Island. Statewide screening: 13,307 infants screened in 8 Rhode Island hospitals 7/1/93–6/30/94.	Demonstration project: 18 infants with unilateral and bilateral sensorineural hearing loss were identified, for a prevalence rate of 5.4/1,000. Statewide screening: 29 (02%) infants were referred for diagnostic BAER and 29 (2.1%) for BOA. Did not indicate the prevalence rate.	2-stage sensitivity of 100% and specificity of 99%.

*TEOAE = transient evoked otoacoustic emissions; BAER = brainstem auditory evoked response; BOA = behavioral observation audiometry

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Vohr BR, Carty LM, Moore PE, Letourneau K: The Rhode Island Hearing Assessment Program: experience with statewide hearing screening (1992-1996). J Pediatr. 1998;133:353–357.	Retrospective analysis of hearing screening and rescreening, number of infants refered data collected prospectively for 53,121 live births in Rhode Island. Objective was to evaluate key outcomes of a universal hearing screening program.	All livebirths in 8 birthing hospitals in Rhode Island born 1/1/93–12/31/96 Parents were given a brochure about the screening process in their preadmission packet, a hearing screen video on the hospital teaching channel, childbirth classes, and a brochure in their baby's crib after the screening.	All livebirths in Rhode Island that were screened with TEOE* in 8 maternity hospitals 1/1/93–12/31/96.	53,121 livebirths in the 8 hospitals. 9.7% were in the NICU*. 52,659 (86%) were screened 677 (1.3%) did not pass 2-stage screening. An additional 1% referred for risk factors.	111 (2.1/1,000) infants identified with hearing loss. 9.7/1,000 were from the NICU; 1.3/1,000 were from WBN*. 79 (71%) had bilateral hearing loss. 32 (29%) had unilateral hearing loss.	Time and experiences are important factors. A steady improvement in screening outcome was noted over the 4 years. 44 (39.6%) of identified babies had no risk factors 60.3% had one or more risk factors Most common risk factors were family history and craniofacial anomalies Sensitivity was 95%; specificity was 87%; and positive predictive value improved from 2% to 16%. In 1993, 26% were lost to the system. In 1996, 12% were lost to the system.

* TEOAE = transient evoked otoacoustic emissions; NICU = neonatal intensive care unit; WBN = well-baby nursery.

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Vohr, B, Simon P, McDermot, C, Kurtzer-White E, Johnson MJ, Topol D: Early Hearing Screening, Detection and Intervention (EHDI) in Rhode Island. Medicine and Health/Rhode Island. 2002; 85(12):369–372.	Authors describe the changes in screening, re-screening protocol 1993–2000. Screening protocol 1993–1996 was: OAE/OAE*. To reduce referral rates, in 1997 the Women and Infants Hospital in Rhode Island piloted a new protocol: OAE/AABR*. By 2000, all Rhode Island used the OAE/AABR protocol.	1/1/93–3/8/95	For screening: Those who didn't pass OAE were re-screened with AABR at 35dB*.	108,154 infants were eligible for screening; 10.4% were NICU* babies. 99.7% of the babies were screened. 6,299 (5.9%) failed 1st screen; 735 (0.68%) failed 2nd screen; 735 (0.68%) were referred for evaluation. 2,173 (1.8%) passed 1st screen, but were referred because of risk factors.	231 (1.9/1,000) were identified with hearing loss: NICU = 9.4/1,000 WBN*=1.1/1,000 Of total infants identified, 46% were from NICU and 65% from WBN. 151 (65%) had a bilateral loss. 80 (35%) had a unilateral loss.	By 2001, referral rate reduced to 1.6%. Age of identification decreased from 9 months in 1993 to 1.7 months in 2001. Average age of amplification declined from 13 months in 1993 to 3.3 months in 2001.

*OAE/OAE = otoacoustic emissions followed by otoacoustic emissions; OAE/AABR = otoacoustic emissions followed by automated auditory brainstem response; dB = decibel; NICU = neonatal intensive care unit; WBN = well-baby nursery.

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Watkin PM, Baldwin M: Confirmation of deafness in infancy. Arch Dis Child. 1999; 81:380–389.	Purpose was to assess reasons for the delay between failed hearing screen and confirmation of PCHI*. A 2-stage protocol was used: TEOAE* was followed by ABR* if infant failed initial hearing screen. The PPV* of the ABR test and measures of delay between identification and diagnosis were analyzed. Prevalence estimates were calculated for children with mild, moderate, severe, and profound hearing loss.	All infants who had ABR thresholds of >40dB* nHL* in the better ear were referred to diagnostic assessment clinics. Medical and birth histories were obtained; physical and audiological examinations were conducted. Children with ABR thresholds of ≤40 dB were not referred for habilitation but were included in the prevalence estimates for mild PCHI.	Infants identified January 1992–December 1997 with permanent congenital hearing loss of >40dB nHL in the better ear. Mild hearing loss = 20dB–40dB nHL. Moderate hearing loss = 41dB–70dB nHL. Severe hearing loss = 71dB–95dB nHL. Profound hearing loss ≥95dB nHL.	25,199 newborns were screened with TEOAE. 596 failed bilaterally; 271 failed unilaterally. 34 had targeted bilateral PCHL; 9 of those referred for ABR were subsequently confirmed with a mild permanent congenital hearing loss.	1.35/1,000 bilateral >40 dB. 0.12/1,000 bilateral >40dB were directly referred to ABR (no TEOAE done). 0.36/1,000 had mild hearing loss 0.36/1,000 had unilateral hearing loss	For infants with thresholds of 80dB or greater, the PPV was 100%. For infants with thresholds of 70dB and 50dB the PPVs were 60% and 8%, respectively. The identification of hearing loss was delayed in all infants except those with severe or profound bilateral PCHI.

*PCHL = permanent congenital hearing impairment; TEOAE = transient evoked otoacoustic emissions; ABR = auditory brainstem response; PPV = positive predictive value; dB = decibel; nHL = normalized hearing level.

REFERENCE	DESIGN	RECRUIT-MENT	CASE DEFINITION	SUBJECTS	PREVALENCE	AUTHORS' CONCLUSIONS (other findings)
White KR, Vohr BR, Maxon AB, Behrens TR, McPherson MG, Mauk GW: Screening all newborns for hearing loss using transient evoked otoacoustic emissions. Int J Pediatr Otorhinolaryngol. 1994; 29:203–217.	Goal of project was to evaluate whether TEOAE* could be used accurately and cost efficiently screen all infants. 2 samples of randomly selected infants were screened for hearing loss and rescreened at 4–6 weeks if they failed either test. Those who did not pass 2nd screening were referred for complete audiological assessment using diagnostic ABR* or behavioral audiological evaluation techniques, or both. Risk factors and medical and demographic characteristics were collected.	1,850 infants were randomly selected from Women and Infants Hospital in Rhode Island. Infants were rescreened 4–6 weeks later with TEOAE or ABR, or both.	Infants born 8/15/90–2/28/91 with a bilateral or unilateral hearing loss of >25 dB* at 500 Hz* or 4 kHz* or both.	1,850 newborns: 304 (16.4%) were NICU* babies; 1,546 (83.6%) were from WBN*. 2 samples were recruited: (1) 464 who were screened with both TEOAE and ABR regardless of TEOAE results and (2) 1,386 who were screened with ABR only if they failed TEOAE.	Of 1,850 infants screened, 115 were referred for diagnostic evaluation. 11 (5/9/1,000)were identified with hearing loss. 6 infants had bilateral hearing loss: none had mild or moderate loss, all had severe to profound loss. 5 infants had unilateral hearing loss: none had mild loss; 1 had moderate loss; and 4 severe to profound loss.	37 (20/1,000) additional infants were identified with recurrent conductive hearing losses 5 (45%) of the 11infants identified would have been missed if only NICU and high-risk infants were tested. Cost of TEOAE screening was estimated to be $25 per baby.

*TEOAE/OAE = transient evoked otoacoustic emissions; ABR = auditory brainstem response; dB = decibel; Hz = hertz; kHz = kilohertz; NICU = neonatal intensive care unit; WBN = well-baby nursery.