REVIEW ARTICLE

Universal Newborn Hearing Screening: Systematic Review to Update the 2001 US Preventive Services Task Force Recommendation

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ABSTRACT

OBJECTIVE. This review is an update for the US Preventive Services Task Force on universal newborn hearing screening to detect moderate-to-severe permanent, bilateral congenital hearing loss. We focus on 3 key questions: (1) Among infants identified by universal screening who would not be identified by targeted screening, does initiating treatment before 6 months of age improve language and communication outcomes? (2) Compared with targeted screening, does universal screening increase the chance that treatment will be initiated by 6 months of age for infants at average risk or for those at high risk? (3) What are the adverse effects of screening and early treatment?

METHODS. Medline and Cochrane databases were searched to identify articles published since the 2002 recommendation. Data from studies that met inclusion criteria were abstracted, and studies were rated for quality with predetermined criteria.

RESULTS. A good-quality retrospective study of children with hearing loss indicates that those who had early versus late confirmation and those who had undergone universal newborn screening versus none had better receptive language at 8 years of age but not better expressive language or speech. A good-quality nonrandomized trial of a large birth cohort indicates that infants identified with hearing loss through universal newborn screening have earlier referral, diagnosis, and treatment than those not screened. These findings are corroborated by multiple descriptive studies of ages of referral, diagnosis, and treatment. Usual parental reactions to an initial nonpass on a hearing screen include worry, questioning, and distress that resolve for most parents. Cochlear implants have been associated with higher risks for bacterial meningitis in young children.

CONCLUSIONS. Children with hearing loss who had universal newborn hearing screening have better language outcomes at school age than those not screened. Infants identified with hearing loss through universal screening have significantly earlier referral, diagnosis, and treatment than those identified in other ways. *Pediatrics* 2008; 122:e266–e276

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Key Words
evidence-based medicine, hearing impairment, hearing screening, newborn screening, systematic reviews

Abbreviations
USPSTF—US Preventive Services Task Force
UNHS—universal newborn hearing screening
PCHL—permanent congenital hearing loss
JCIH—Joint Committee on Infant Hearing
OAE—otoacoustic emission
ABR—auditory brainstem response
CI—confidence interval
NNS—number needed to screen

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THIS SYSTEMATIC EVIDENCE review updates a previous review for the US Preventive Services Task Force (USPSTF) on universal newborn hearing screening (UNHS) to detect moderate-to-severe bilateral permanent congenital hearing loss (PCHL).[1,2] Based on research available in 2001, the USPSTF concluded that the evidence was insufficient to recommend for or against routine screening of newborns for PCHL during the postpartum hospitalization.[3]

In their previous recommendation, the USPSTF found that evidence was inconclusive regarding whether earlier treatment resulting from UNHS leads to clinically important improvement in speech and language skills at 3 years of age and beyond, because existing studies had important design limitations. Since then, additional studies with speech and language outcomes of children screened or not screened as newborns have been published. This update focuses on these new studies.

The Joint Committee on Infant Hearing (JCIH), composed of representatives from audiology, otolaryngology, pediatrics, education, and state speech and hearing programs, provides position statements and establishes practice standards for early identification, intervention, and follow-up care for infants and young children with hearing loss. According to the JCIH, hearing screening should identify newborns at risk for specifically defined hearing loss that interferes with development.[4,5] The aim for UNHS programs is detection of permanent sensory or conductive hearing

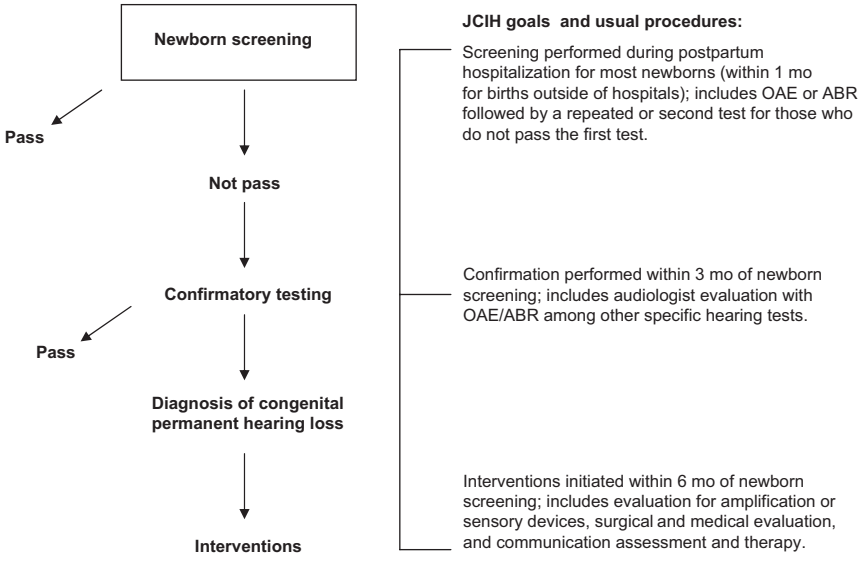

FIGURE 1
Process of screening and follow-up.

loss averaging 30 to 40 dB or more in the frequency region important for speech recognition (~500–4000 Hz). The focus of UNHS is on congenital as opposed to acquired or progressive hearing loss that may not be detected in the newborn period.

The rate of PCHL among newborns ranges from 1 to 3 per 1000 live births.[6–9] Compared with children with normal hearing, those with hearing loss have more difficulty learning vocabulary, grammar, word order, idiomatic expressions, and other aspects of verbal communication.[10] Hearing loss in children is also associated with delayed language, learning, and speech development and with low educational attainment.[4] Hearing disorders have also been associated with increased behavior problems, decreased psychosocial well-being, and poor adaptive skills.[11–13]

Risk factors associated with a higher incidence of PCHL include NICU admission for ≥2 days; syndromes associated with hearing loss; family history of hereditary childhood sensorineural hearing loss; craniofacial abnormalities; and congenital infections.[4] However, ~50% of infants with PCHL do not have any known risk factors.[9,14–18]

In the JCIH 2000 and 2007 position statements,[4,5] the JCIH endorsed integrated, interdisciplinary state and national systems of UNHS; evaluation; and family centered intervention outlined in Fig 1. In addition, the JCIH recommended that all infants with risk indicators should undergo periodic monitoring for 3 years. The 2007 statement expands screening protocols for NICU infants and provides additional guidance for the diagnostic audiology evaluation, medical evaluation, early intervention, surveillance, communication, and tracking.[5]

Newborn hearing screening involves the use of objective physiologic measures. Currently, otoacoustic emissions (OAEs) and/or auditory brainstem responses (ABRs) are most often used to detect sensory or conductive hearing loss.[19] Both technologies are noninvasive recordings of physiologic activities that are easily recorded in newborns and are highly correlated with the degree of peripheral hearing sensitivity. In UNHS programs, a 2-step process using OAEs followed by ABRs in those who fail the first test is often used to improve test performance. In a large trial using this approach, screening test sensitivity and specificity were 0.92 and 0.98, and the positive and negative likelihood ratios were 61.00 and 0.08, respectively.[15] Referral rates are lower in programs using dedicated technicians rather than volunteers and students.[20] Under ideal conditions, instruments designed specifically for newborns can test and record findings on sleeping newborns in <5 minutes.

With legislation for UNHS being enacted in 39 US states in recent years, screening practices and procedures have become routine in the postpartum hospital setting. Newborn hearing screening is generally well accepted and tolerated by parents. Rates of refusals in a US community-based health system were reported as 7 of 8707 during the first 10 months of 2007 (Christi Sperry, AuD, Providence Health and Services, written communication, 2007).

Infants not passing the newborn screening tests are referred for confirmatory testing for a diagnosis of PCHL. The American Academy of Pediatrics has set a referral standard of <4% of all screened newborns, and some hospitals use this measure to monitor quality of the screening program. Confirmation requires an evaluation by an audiologist using behavioral, as well as technological, methods. Although the American Academy of Pediatrics has set a standard of 95% for compliance with follow-up testing, this rate is typically much lower depending on tracking systems and local practices and services.[21–24]

The JCIH recommends that early intervention services should be designed to meet the individualized needs of the infant and family, including acquisition of communication competence, social skills, emotional well-being, and positive self-esteem.[4] Early intervention includes evaluation for amplification or sensory devices, surgical and medical eval-

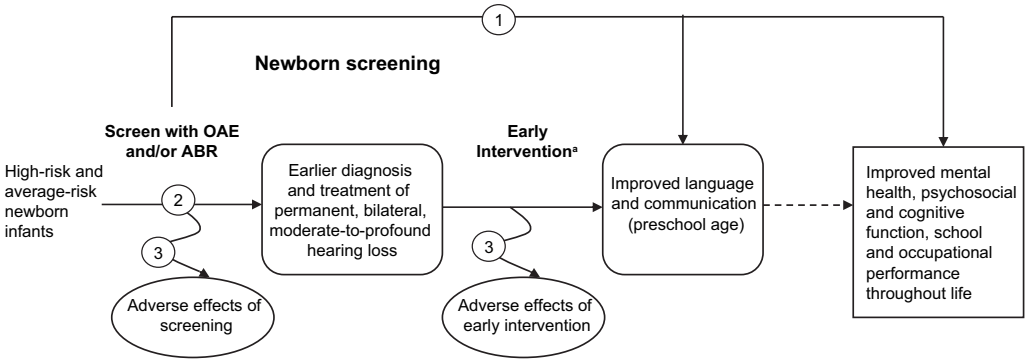

FIGURE 2
UNHS analytic framework and key questions. The key questions are: (1) Among infants identified by universal screening who would not be identified by targeted screening, does initiating treatment before 6 months of age improve language and communication outcomes? (2) Compared with targeted screening, does universal screening increase the chance that treatment will be initiated by 6 months of age for infants at average risk or for those at high risk? (3) What are the adverse effects of screening and early treatment? [a] Interventions include hearing aids or other amplification, cochlear implants, American Sign Language and/or English instruction, speech and language therapy, family education, and support.

uation, and communication assessment and therapy. Cochlear implants are often considered in infants with severe-to-profound hearing loss after inadequate response to hearing aids.[25–28] Research in neurologic and auditory cortical development suggests that early verses late implantation may be linked to more normal cortical auditory pathway development.[29–31]

METHODS

Evidence reviews for the USPSTF follow a specific methodology.[32] The analytic framework outlines the patient population, interventions, outcomes, and adverse effects of the screening process (Fig 2). Corresponding key questions examine a chain of evidence about the effectiveness and potential adverse effects of UNHS and subsequent early intervention. Key questions include: (1) Among infants identified by universal screening who would not be identified by targeted screening, does initiating treatment before 6 months of age improve language and communication outcomes? (2) Compared with targeted screening, does universal screening increase the chance that treatment will be initiated by 6 months of age for infants at average risk or for those at high risk? (3) What are the adverse effects of screening and early treatment?

For this review, targeted screening indicates selective screening of newborns based on the presence of risk factors or associated conditions. Newborns at high risk are those with risk factors known to be associated with PCHL and/or newborns admitted to the NICU. Additional key questions included in the previous report, such as sensitivity and specificity of testing procedures, were not updated in this report, because they were adequately addressed by existing evidence.[1,2]

We conducted literature searches to systematically identify articles relevant to the key questions.[33] Databases included the Cochrane Central Register of Controlled Trials, Cochrane Database of Systematic Reviews, Database of Abstracts of Reviews of Effects (through the fourth quarter of 2007), and Ovid Medline (2000 to November 2007 for key questions 1 and 2 and 1996 to November 2007 for key question 3). Additional articles were obtained from reference lists of related reviews, studies, editorials, reports, Web sites, and by consulting experts. Figure 3 describes results of the searches.

We reviewed abstracts and selected full-text articles

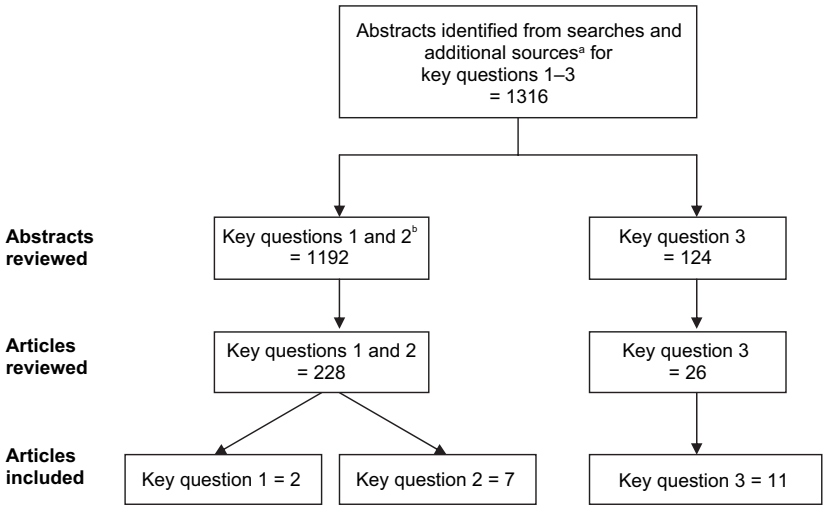

FIGURE 3
Yields from searches, abstract review, and article review. [a]Abstracts were identified from reference lists, experts, and so forth. [b]Some studies overlap between key questions.

based on inclusion and exclusion criteria specific to each key question. Eligible studies addressed key questions and were English language; conducted in the United States or a comparable location; and, for screening studies, included infants screened before 6 months of age. For key questions 1 and 2, we included controlled trials and observational studies. For key question 3 on adverse effects, we included descriptive, as well as comparative, studies. Data from the full text of the original articles were abstracted to evidence tables.

Study quality was rated by using design-specific criteria developed by the USPSTF.[32] The overall rating of each study considers internal validity and applicability. Descriptive studies without quality criteria are summarized. An outcomes table estimating the number needed to screen under various assumptions was determined using estimates from the most relevant studies.

The USPSTF advised the Oregon Evidence-based Practice Center in formulating and reporting this review. Additional experts provided comments on an earlier draft.

RESULTS

Key Question 1: Among Infants Identified by Universal Screening Who Would Not Be Identified by Targeted Screening, Does Initiating Treatment Before 6 Months of Age Improve Language and Communication Outcomes?

A Cochrane review comparing the long-term effectiveness of UNHS and early treatment with high-risk or opportunistic screening was updated in February 2005.[34] No randomized, controlled trials were identified that fulfilled inclusion criteria. No additional trials comparing screening approaches were identified by our searches.

A good-quality retrospective cohort study evaluated the effect of UNHS on speech and language outcomes of children with PCHL.[35] A total of 120 children with PCHL were identified from a cohort of 157 000 children born in 8 districts of southern England between 1992 and 1997 and underwent speech and language assessment at school age (mean: 7.9 years; range: 5.4–11.7 years). Included children were either part of the Wessex Universal Neonatal Hearing Screening Trial,[16] constituting 34% of the birth cohort in this study, or from districts in greater London providing UNHS or not at the time of birth. Seventy-one percent of children with PCHL in the cohort who were eligible for the study participated. Participants were similar to nonparticipants in age, gender, and severity of hearing loss.

Protocols for screening and confirmation of hearing impairment were similar at all of the sites, and all of the children had bilateral impairment of a ≥40-dB hearing level.[16] All of the children were also screened using the Health Visitor Distraction Test at 7 to 8 months of age as usual care in the United Kingdom. Therapy was provided for all of the children as a public health service and included education and audiology services with access to hearing aids. Sixty-three age-matched children with normal hearing underwent testing to derive *z* scores for outcome measures.

Baseline characteristics were similar between comparison groups. Outcome measures were adjusted for degree of hearing loss, maternal education, and age-adjusted nonverbal ability. Receptive language was evaluated by the Test for Reception of Grammar, British Picture Vocabulary Scale, and aggregate scores. Expressive language was evaluated by the Renfrew Bus Story Test sentence information and 5 longest sentences and aggregate scores. Speech was evaluated by the Children's Communication Checklist speech scale. Evaluators were blinded to the children's histories.

Children with hearing impairment confirmed by ≤9 months of age had significantly better age-adjusted scores than those confirmed later on 2 tests of receptive language and 1 of 2 tests of expressive language but not on the speech scale. All of the aggregate scores for receptive and expressive language were significantly better for the early confirmation group. Differences in higher scores for early versus late confirmation are equivalent to an increase of 10 to 12 points in the verbal compared with nonverbal IQ.

Children who underwent UNHS had better scores than those who did not on 2 tests of receptive language but not on 2 tests of expressive language or speech. Aggregate scores for receptive language were better for the UNHS group.

Limitations of the study include the potential for underestimation of the size of benefit because the system of screening and follow-up has improved since the study birth cohort underwent these processes. Also, it is not clear whether children not undergoing UNHS had the onset of hearing impairment after birth or not. A sensitivity analysis indicated that benefit for the UNHS group would have been higher if all of the cases in the study were truly congenital. Speech was assessed on the basis of parental or professional report, which may lack sensitivity as an outcome measure, rather than by direct measurement. This study does not report the proportions of hearing-impaired children that would have been considered high versus average risk for hearing impairment at birth. However, the proportion of children with other disabilities (13%–26%), a possible surrogate for risk at birth, was similar between early versus late confirmation and UNHS versus not-screened comparison groups.

A fair-quality retrospective cohort study conducted in Australia provides speech and language outcomes for a birth cohort exposed to targeted newborn hearing screening.[36] This study examined the relationship of age at diagnosis of PCHL and severity of impairment on several language, speech, and reading measures in children 7.0 to 8.0 years of age who were fitted with hearing aids by age 4.5 years. All of the children born in the state of Victoria who were identified with PCHL through risk-based screening of infants, universally available behavioral hearing screening at 8 to 10 months of age, and other referral mechanisms were included. The government provided services for all of the eligible children and data obtained at the time that services were used in the study.

Several outcome measures were examined using validated methods, including receptive and expressive language, receptive vocabulary, cognition, articulation, read-

ing comprehension, intelligibility, and family functioning. Evaluators were blinded to the children's history and hearing status. Regression models were constructed that controlled for confounders, including nonverbal IQ, maternal education, paternal occupational prestige, and family functioning.

Few children in the cohort were diagnosed with PCHL at <6 months of age (n = 11) or 12 months (n = 28). The mean age of diagnosis was 21.6 months, and the mean age of hearing aid fitting was 23.2 months. Comparisons of characteristics between early versus late-diagnosed children were not reported, except that the age at diagnosis was negatively correlated with severity at diagnosis. Age at diagnosis did not contribute significantly to the variance on any measures except receptive vocabulary; however, the small number of children diagnosed at young ages may provide inadequate power to evaluate an age effect. The severity of impairment contributed significantly to the variance on all of the measures except reading comprehension. Language outcomes were >25 points lower than expected from IQ scores.

Several other observational studies report the effects of early intervention programs.[37–45] These studies were reviewed for the previous USPSTF recommendation. All of these studies have important methodologic limitations, including the use of convenience samples, nonblinded assessments, and lack of information on attrition and follow-up, among others.[1,2] All of these studies report better outcomes for children with hearing impairment identified and/or treated early versus late.

Key Question 2: Compared With Targeted Screening, Does Universal Screening Increase the Chance That Treatment Will Be Initiated by 6 Months of Age for Infants at Average Risk or for Those at High Risk?

The Wessex Universal Neonatal Hearing Screening Trial is a good-quality nonrandomized, controlled trial investigating whether the addition of UNHS to usual care screening at 7 to 8 months of age versus usual care screening alone increases detection and improves the early management of infants with PCHL in the United Kingdom.[16] The trial included all of the newborns born in 4 participating hospitals from 1993 to 1996, including 25 609 born during periods of UNHS and 28 172 not screened as newborns. Newborns screening positive using OAE followed by ABR for those who failed the first test were referred for audiologic assessment to determine their hearing levels. All of the infants were also subjected to screening using the Health Visitor Distraction Test at 7 to 8 months of age as usual care. Children with abnormal newborn screening tests, abnormal Health Visitor Distraction Tests, or additional concern for impairment were referred to audiology services. These are public health services available to all children.

All of the newborns enrolled in the Wessex Trial were included in an 8-year follow-up study.[15] Information about diagnoses and management was obtained from multiple sources (records, therapists, etc). Children with postnatal causes of hearing impairment were excluded from the study.

In this analysis, 1 additional case of PCHL was referred before the age of 6 months for every 1969 (1011–12 896) infants in the UNHS population. More infants with true PCHL were referred to audiology services before 6 months of age if they were born during periods with UNHS rather than during periods without (74% vs 31%; difference: 43% [95% confidence interval (CI): 19%–60%]; P = .001). Adjustment for the effect of severity of hearing impairment on age of referral increased the odds ratio between newborn screening and early referral from 6.3 to 6.9 (95% CI: 2.2–22.0; P = .001). The age at referral was lower for children undergoing UNHS versus not (0 vs 8 months; $P <$ .001).

Results may have been limited by the effects of initiating a new clinical service as part of the trial. Parents of 7 children with subsequently diagnosed hearing impairment initially refused newborn screening. Also, 23% of all of the cases included in the analysis may actually have had progressive hearing losses that would not be detected at birth. Although referrals were early in the study, management was often initiated later than desired (48% after 18 months).

Several descriptive studies report follow-up data from UNHS programs (Table 1).[9,16–18,46,47] In the most recent study of a national UNHS program in the United Kingdom, referred infants who were not admitted to the NICU had their first follow-up visits at a median age of 4 weeks, were diagnosed at 10 weeks, enrolled in education services at 10 weeks, and were fitted with hearing aids at 14 weeks.[17] Infants from the NICU used these services at slightly older ages and had their first follow-up at 9 weeks, were diagnosed at 13 weeks, and received hearing aids at 24 weeks.

In a national survey of 151 parents with children <6 years old with hearing impairment in the United States, children screened as newborns were diagnosed and received hearing aids at younger ages than those not screened.[48] For children with unknown causes for hearing impairment, the median ages of confirmation for screened versus nonscreened children were 4.0 vs 25.0 months for mild or moderate impairment and 2.0 vs 15.0 months for severe or profound impairment. The median ages for hearing aid fitting for screened versus nonscreened children were 6.0 vs 30.5 months for mild or moderate impairment and 4.0 vs 16.0 months for severe or profound impairment.

Key Question 3: What Are the Adverse Effects of Screening and Early Treatment?

Two fair-quality cohort studies,[49,50] 1 poor-quality case-control study,[51] and 5 survey studies with >40% response rates[52–56] provide relevant information on adverse effects of newborn hearing screening (Table 2). In a case-control study, parents expressed anxiety when their infants did not pass the initial screening test that resolved for most after a confirmatory test indicated normal results.[51] Additional studies reported no differences in concern and anxiety between parents with newborns who passed or did not pass screening tests,[49,50] although parents with newborns with confirmed hear-

TABLE 1 **Descriptive Studies of UNHS Follow-up**

Source	Program Description	Age Screened	No. of Cases	Proportion High Risk	Age Referred	Age at First Follow-up	Age Diagnosed	Age Enrolled in Programs	Age of Hearing Aid Fitting
Uus and Bamford,[17] 2006, United Kingdom	169 487 infants at 23 sites in the first phase of a national UNHS program in 2001–2003. Well newborns had OAE, then ABR if needed; NICU newborns had both tests. Referred ≥40 dB hearing level	Before hospital discharge	169	54% with risk factors[a]	At screening	Well infants: median age 4 wk; NICU: 9 wk	Well infants: median age 10 wk; NICU: 13 wk	Median age 10 wk for education services	Well infants: median age 14 wk; NICU: 24 wk
Joseph et al,[46] 2003, Singapore	UNHS of 4387 newborns in 1999–2001 at 1 hospital using OAE; positives were rescreened at ~2 wk, and again at 6 wk if needed; referred if specific criteria not met	Most within 24 h	8	38% high risk	Those who tested positive at 6 wk were referred for formal evaluation	NR	7 of 8 by 7 mo	Interventions in place by 9 mo for 4	NR
Bailey et al,[47] 2002, Australia	UNHS of 12 708 newborns in 5 hospitals in 2000–2001 using OAE and ABR if needed; referred at ≥35 dB hearing level	Before hospital discharge	9	5 NICU; 8 with risk factors	NR	NR	NR	NR	6 had hearing aids by 6 mo; 1 at 19 mo
Mehl and Thomson,[9] 2002, Colorado	Colorado Newborn Hearing Screening Project screened 148 240 newborns in 1992–1999; ABR in 52 hospitals; OAE in 3 hospitals; 2-stage screening in 2 hospitals; referred at >35-dB hearing level in 1 or both ears	Before hospital discharge	291 (71% bilateral)	47% with risk factors	Median age: 2.1 mo	NR	NR	NR	NR
Dalzell et al,[18] 2000, New York	UNHS of 43 311 newborns in 8 hospitals in New York state in 1995–1996 with OAE and ABR if needed; referred >20-dB hearing level	Before hospital discharge	85	61% NICU; 67% with risk factors	NR	NR	Median: 3 mo of age: younger for well infants and for severe or profound impairment	Median: 3 mo	Median: age 7.5 mo
Wessex UNHS Trial Group,[16] 1998, United Kingdom	UNHS arm of the Wessex Trial including 25 609 newborns screened in 1993–1996 with OAE and ABR if needed; referred at ≥40-dB hearing level	Within 48 h	23	41% special care units; 74% had risk factors	All by 6 mo of age	NR	67% by 10 mo	63% by 10 mo	NR

[a] Risk factors include NICU for >48 hours, family history of hearing impairment, and craniofacial anomaly.

TABLE 2 **Studies of Potential Adverse Effects of Screening**

Source (Quality)	Study Design	Subjects	Screening	Setting	Results
Kennedy et al,[49] 1999 (fair)	Retrospective cohort	Parents of average-risk newborns: 100 passed UNHS and 100 did not pass; subset of Wessex Trial	OAE and ABR	United Kingdom; 2–12 mo after UNHS	No differences in scores on the Spielberger State-Trait Anxiety Inventory and Attitudes Toward the Baby Scale between parents of newborns who passed UNHS versus not passed
Weichbold and Welzl-Mueller,[50] 2001 (fair)	Prospective cohort	85 mothers whose newborns failed first and/or second screening tests	OAE; 2 times	Innsbruck, Austria	59% of mothers whose newborns failed the first screen were not concerned, 27% were slightly concerned, and 14% were highly concerned; in an additional sample of 43 mothers whose newborns failed the second screening, 42% were not concerned, 37% were slightly concerned, and 21% were highly concerned; differences in proportions between groups were not statistically significant
Poulakis et al,[51] 2003 (poor)	Case control	Parents of infants: 108 at risk for hearing impairment; 64 control subjects for at risk infants; 103 failed distraction test; 53 control subjects for infants failing the test	Distraction test	Australia	Parent concerns about language development, general development, and perceived vulnerability to ill health did not differ among the 4 groups; ~18% of parents continued to feel worried 6 mo after the definitive hearing testing; 6% rated the test procedures as somewhat difficult or unpleasant; parents of children who failed the distraction test reported more negative emotions (anger, sadness, upset, worry, and confusion) after their child's definitive hearing test than parents of children considered at risk ($P < .05$)
Crockett et al,[52] 2006	Survey (questionnaire)	Parents of 722 screened newborns (53% response rate); 103 with 1 or 2 negative tests; 81 with third negative test; 105 with third test positive in 1 ear; 55 with third test positive in both ears	OAE; ABR final test	United Kingdom; 3 wk and 6 mo postscreening	Significant trends for increased anxiety ($P < .05$), increased worry ($P < .001$), and decreased certainty ($P < .001$) as the number of tests increased; parents in group 4 who understood test implications had lower anxiety ($P = .01$) and lower worry ($P < .01$) versus those who did not
de Uzcategui and Yoshinaga-Itano, 1997[53]	Survey (questionnaire)	Parents of 201 screened newborns who were referred for further testing (51% response rate)	Not reported	Colorado; 2 university hospitals	78% of parents were not angry, 81% felt informed, 38% did not feel comforted by hospital staff; 14% had negative emotions, half had a child with a confirmed hearing loss; parents of children with confirmed hearing losses had a higher level of frustration, anger, depression, and confusion versus other parents; 25% of the sample did not return for follow-up testing after a referral was indicated
Hergils and Hergils,[54] 2000	Survey (questionnaire)	Parents of 83 screened newborns (95% response rate)	OAE	Linkoping, Sweden; well-infant visit, 5–6 mo of age	76 were satisfied with screening, 3 neutral, 3 dissatisfied, 1 did not know; screening raised questions for 28 and no questions for 44; 79 were positive about the test and 4 were negative; information on the test was sufficient for 64 and insufficient for 9; the majority of parents were positive about the screening; most felt early detection was good, test was easy and did not bother their infant; negative comments included the test being too demanding, test took too long, clearing the ear canal would be difficult for newborns; complaints included getting information about the test earlier and test methodology; parents of 6 of the 10 infants needing retesting reported anxiety
Russ et al,[55] 2004	Survey (questionnaire)	Parents of 134 hearing impaired children after hearing aid fitting (61% response rate)	ABR; distraction test	Victoria, Australia	Themes analysis showed parents had generally positive responses to ABR screening and mixed response to the distraction test; denial and shock at diagnosis; frustrations in delays in diagnosis; and communication difficulties with providers; difficulty testing children with other medical and development problems was also reported
Vohr et al,[56] 2001	Survey (interview)	Mothers of 307 screened newborns (85% response rate); mothers of 40 newborns needing rescreening (90% response rate)	OAE	Rhode Island	Significantly more mothers with infants who were rescreened worried about the test results compared with mothers with infants undergoing only 1 screening ($P < .001$); for mothers of infants requiring rescreening, the degree of worry at the time of the rescreening was significantly greater than at the first screen ($P < .001$); greater worry at the initial screening was seen in mothers with less than high school education ($P = .003$) and who were bilingual ($P = .006$), nonmarried ($P = .02$), and nonwhite race ($P = .005$); learning about screening during hospitalization versus before arriving was also associated with greater worry ($P = .012$)

ing loss had higher levels of distress than those with confirmed normal hearing.[51,53]

No studies addressed the potential adverse effects of early treatment using hearing aids, American Sign Language, English instruction, speech and language therapy, or family education and support. Case series reports of cochlear implantation indicate few surgical complications in children.[57–60] However, among children who had

cochlear implantation from 1997 to 2002 at <6 years of age, 41 episodes of postimplantation bacterial meningitis occurred among 38 children.[61,62] None of the children with meningitis received their implants at <12 months of age, and rates of infection did not indicate age-related risks. The highest risks were among those using implants with positioners,[63] a wedge inserted next to the implanted electrode to facilitate transmission. Implants with the positioner were voluntarily recalled in the United States in 2002.

Few studies of psychological issues associated with treatment specifically focus on young children or the adverse effects of early treatment. Parents of 123 children with hearing loss (average: 38 months of age) were asked about their stress and quality of life related to 3 types of treatments.[64] All had cochlear implants, hearing aids, or switched treatments (hearing aid first then cochlear implants). Parents of children with switched treatments had significantly reduced quality of life and increased stress, whereas parents of children using only cochlear implants had improved quality of life and normal stress.[64]

Parents of 28 children 12 to 30 months old undergoing cochlear implantation in Turkey noted on questionnaires that making the decision for cochlear implantation was stressful.[65] Families were anxious about possible device failure and maintenance of the equipment and acknowledged that their children needed more support from the family after the implantation. Most parents reported benefits of implantation, including improved communication, self-confidence, well-being, and social relationships.

Yield of Screening

Although no studies directly compare the yields of universal versus targeted screening approaches, estimates can be determined by applying results of relevant studies in an outcome table model (Table 3). Assumptions for the model include the proportion of newborns considered high risk,[15] the prevalence of PCHL in populations at high and average risk,[1] the proportion not screened in the hospital,[66] the sensitivity of 2-stage screening,[15] the compliance with follow-up testing (estimated), the accuracy of diagnostic tests,[1] and the proportion of newborns at average risk diagnosed with PCHL by 3 months (estimated). Assumptions in this model differ from those in the previous evidence review because they draw from recently published data of universal and targeted screening.[15,66] Using these assumptions, if 10 000 newborns underwent UNHS, there would be 11 to 12 diagnosed cases by 3 months of age, 86 false-positive screening test results, and possibly 1 missed case. The number needed to screen (NNS) to diagnose 1 case would be 878. If only newborns at high risk underwent screening, there would be 4 or 5 diagnosed cases, 6 false-positive screening test results, and 8 or 9 missed cases. The NNS to diagnose 1 case would be 178.

TABLE 3 **Yield of Screening in a Hypothetical Cohort of 10 000 Newborns for Moderate-to-Profound PCHL**

Relevant Factors	Proportions or Rates	Hypothetical Model	
		UNHS	High-Risk Screening[a]
Assumptions			
Proportion high risk[15]	0.0800	—	—
Prevalence[1]	—	—	—
High-risk group	0.0080	—	—
Average risk group	0.0008	—	—
Miss rate for UNHS (proportion not screened in hospital; estimate)	—	—	—
In high risk	0.0500	—	—
In average risk	0.0500	—	—
Follow-up rate for misses	0.9000	—	—
Miss rate for high-risk screening[66]	0.2300	—	—
Follow-up rate for misses	0.0000	—	—
Sensitivity of 2-stage screening[15]	0.9200	—	—
Specificity of 2-stage screening[15]	0.9900	—	—
Compliance with follow-up (estimate)	0.9000	—	—
Accuracy of diagnostic ABR[1]	—	—	—
Sensitivity	1.0000	—	—
Specificity	0.9950	—	—
Proportion of average-risk diagnosed by 3 mo without screening (estimate)	0.1000	—	—
Results			
Target group for screening	—	10 000	800
No. of infants screened	—	9500	616
High risk	—	760	616
Average risk	—	8740	0
High-risk cases in screened group	—	6	5
Average-risk cases in screened group	—	7	0
Cases diagnosed by 3 mo	—	11 to 12	4 to 5
High-risk cases missed by screening	—	<1	1 to 2
Average-risk cases missed by screening	—	<1	7
Total No. of cases	—	13	13
False-positive screening test results	—	86	6
Normal infants incorrectly diagnosed to have PCHL at first posthospital audiologic examination	—	<1	<1
NNS to diagnose 1 case	—	878	178
NNS to diagnose 1 additional case by 3 mo	—	1333	NA

NA indicates not applicable.

[a] High risk was defined by risk factors (family history of hearing impairment, perinatal infection, low birth weight, anatomical deformity, birth asphyxia, chromosomal abnormality, and exchange transfusion).

DISCUSSION

Results of a community-based cohort study of children with PCHL indicated that those who had early versus late confirmation and those who had UNHS versus none had better language outcomes at 8 years of age.[35] In contrast, a community-based cohort study of children with hearing impairment who were exposed to risk-based newborn screening indicated no relationship between age at diagnosis and language, speech, and reading measures at 7 to 8 years of age.[36] In this cohort, few children were diagnosed by 6 months of age and had the opportunity for early treatment.

No trials directly compare targeted screening with UNHS and report data about the initiation of early treatment for infants at average risk or those at high risk. Data from a large nonrandomized trial and descriptive studies indicate that infants at average and high risk with PCHL born in hospitals with UNHS have earlier referral and initiation of treatment than those born in hospitals

without UNHS. The most impressive follow-up measures come from the most recently published studies, potentially reflecting refinements in screening techniques, system and process improvements, incorporation of UNHS as a routine practice, and increasing commitment to implementing successful programs in response to practice and policy changes.

Studies of adverse effects of screening indicate that usual parental reactions to an initial nonpass include worry, questioning, and distress. Negative emotions resolve for most parents when a diagnostic test is provided with a normal result. No studies addressed the adverse effects of delaying screening of PCHL. Now that practice standards exist, it would be difficult to conduct adequate comparison studies of early versus late initiation of treatment to evaluate both benefits and adverse effects.

A major limitation of the application of the key studies in this update is that they were conducted outside of the United States. Although the methods of screening and the inpatient maternity experiences are likely similar, the processes of referral, follow-up, and treatment would be expected to differ. Currently, there is no standard method in the United States to track children through these processes to ultimately obtain language outcomes from a birth cohort, as was done in the United Kingdom study,[35] although approaches to do so are being piloted. Factors influencing follow-up and treatment in the United States would need to be considered, as well as exposure to UNHS when determining long-term outcomes.

CONCLUSIONS

More studies of long-term functional outcomes related to UNHS are needed to support the findings of the United Kingdom study.[35] Although the United Kingdom study reported important long-term benefits with UNHS for receptive language, expressive language and speech outcomes were not better. Other functional outcomes, such as school performance, social interactions, and quality of life, may be more relevant to children and their families. Future research should include these as well.

Results of this review indicate that infants identified with PCHL through UNHS have significantly earlier referral, diagnosis, and treatment than those identified in other ways. Although the clinical community has acknowledged the significance of early treatment for many years, evidence of its effect on long-term functional outcomes has been limited. New data on improved language outcomes at school age strengthen the case for UNHS but are also dependent on effective methods of referral, follow-up, and treatment. As these needs are being addressed with ongoing projects, further research will be required to demonstrate effectiveness for the entire process that UNHS initiates.

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