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**Universal Newborn Hearing Screening:  
Systematic Review to Update the 2001  
U.S. Preventive Services Task Force Recommendation**

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## Structured Abstract

**Objective:** This review is an update for the U.S. Preventive Services Task Force (USPSTF) on universal newborn hearing screening (UNHS) to detect moderate to severe permanent, bilateral congenital hearing loss. The review focuses on 3 key questions regarding the effectiveness of universal screening and early interventions in improving language and other outcomes in childhood, the effectiveness of universal screening in identifying infants with hearing loss and leading them to early interventions, and adverse effects of screening and early interventions.

**Methodology:** Literature searches of MEDLINE and Cochrane databases (2000-November 2007) were conducted to systematically identify articles addressing the 3 key questions published since the prior recommendation in 2001. Additional articles were obtained from reference lists of related reviews, studies, editorials, reports, websites, and by consulting experts. Articles were subjected to inclusion and exclusion criteria, data from included studies were abstracted, and studies were rated for quality with pre-determined criteria. Results were summarized descriptively in tables. An outcomes table estimating the number needed to screen was determined using estimates from the most relevant studies.

**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 UNHS versus none had better receptive language at age 8 years, 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 UNHS 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. Universal newborn hearing screening programs have low false-positive and referral rates and are generally well accepted and tolerated by parents of newborns. Studies indicate that usual parental reactions to an initial non-pass on a hearing screen include worry, questioning, and distress. These negative emotions resolve for most parents when a diagnostic test is provided with a normal result. Little information exists about the adverse effects of early interventions, although cochlear implants are associated with higher risks for bacterial meningitis in young children.

**Conclusions:** Children with hearing loss who had UNHS 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.

# Table of Contents

<b>I. Introduction</b> .....	1
Purpose of Review and Prior USPSTF Recommendation.....	1
Condition Definition.....	1
Prevalence and Burden of Disease.....	2
Risk Factors and High Risk Groups.....	2
Current Clinical Practice.....	2
Screening Tests and Diagnosis.....	3
Treatment.....	4
Recommendations of Other Groups.....	4
Analytic Framework and Key Questions.....	4
<b>II. Methods</b> .....	6
Literature Search and Strategy.....	6
Inclusion and Exclusion Criteria.....	6
Critical Appraisal.....	6
Size of Literature Reviewed.....	6
Data Synthesis.....	7
External Review Process.....	7
<b>III. Results</b> .....	8
KQ 1. Among infants identified by UNHS who would not be identified by targeted screening, does initiating treatment prior to age 6 months improve language and communication outcomes?.....	8
Summary.....	8
Evidence.....	8
KQ 2. Compared with targeted screening, does UNHS increase the chance that treatment will be initiated by age 6 months for average risk infants? For high risk?.....	11
Summary.....	11
Evidence.....	11
KQ 3. What are the adverse effects of universal screening and early treatment?.....	13
Adverse Effects of Screening.....	13
Adverse Effects of Early Treatment.....	15
Yield of Screening.....	15
<b>IV. Discussion</b> .....	17
Summary of Review Findings.....	17
Future Research.....	19
Conclusions.....	19
<b>References</b> .....	20

## **Figures**

Figure 1. Process of Screening and Follow-up

Figure 2. UNHS Analytic Framework and Key Questions

## **Summary Tables**

Table 1. Recommendations of Other Groups

Table 2. Cohort Studies Reporting Language Outcomes

Table 3. Descriptive Studies of UNHS Follow-up

Table 4. Studies of Potential Adverse Effects of Screening

Table 5. Yield of Screening in a Hypothetical Cohort of 10,000 Newborns for Moderate to Profound PCHL

Table 6. Summary of Evidence

## **Appendices**

Appendix A. Abbreviations

*Appendix B. Literature Retrieval and Review Process*

Appendix B1. Search Strategies

Appendix B2. Inclusion and Exclusion Criteria

Appendix B3. Studies Excluded from Results of Review

Appendix B4. USPSTF Quality Rating Criteria

Appendix B5. Yield from Searches, Abstract Review, and Article Review

Appendix C. Evidence Table of Key Screening Studies

Appendix D. Expert Reviewers of the Draft Report

# I. INTRODUCTION

## Purpose of Review and Prior USPSTF Recommendation

This systematic evidence review updates a prior review for the U.S. Preventive Services Task Force (USPSTF) on universal newborn hearing screening (UNHS) to detect moderate to severe permanent, bilateral congenital hearing loss (PCHL).<sup>1, 2</sup> In 2001, based on results of a prior systematic evidence review,<sup>3, 4</sup> the USPSTF concluded that the evidence was insufficient to recommend for or against routine screening of newborns for PCHL during the postpartum hospitalization (I Recommendation).

The USPSTF provided additional specific conclusions about the evidence. They determined that methods of screening using otoacoustic emissions (OAEs) and auditory brainstem response (ABR) are highly accurate for identifying PCHL in newborns, and UNHS leads to earlier identification and treatment of infants with PCHL. They found that evidence was inconclusive regarding whether earlier treatment resulting from newborn screening leads to clinically important improvement in speech and language skills at age 3 years and beyond because existing studies had design limitations. The USPSTF considered that earlier identification and intervention may improve the quality of life during the first year of life, and prevent regret by the family over delayed diagnosis of PCHL, but limited data addressing these benefits existed. They were not able to determine whether potential benefits outweighed the potential harms of false-positive tests.

This update focuses on critical evidence gaps that were unresolved at the time of the 2001 recommendation, and utilizes the format and methods of the prior systematic evidence review.<sup>3, 4</sup>

## Condition Definition

The Joint Committee on Infant Hearing (JNIH), comprised 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 infants at risk for specifically defined hearing loss that interferes with development.<sup>5, 6</sup> The targeted hearing loss for UNHS programs is permanent sensory or conductive hearing loss averaging 30 to 40 decibels (dB) or more in the frequency region important for speech recognition (approximately 500 through 4000 Hertz [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 term “hearing impairment” is commonly used outside the U.S. instead of “hearing loss.” These terms refer to the same condition and are both used in this review. (A list of all abbreviations is located in Appendix A.)

## **Prevalence and Burden of Disease**

The rate of PCHL among newborns ranges from 1 to 3 per 1,000 live births.<sup>7-10</sup> Hearing loss occurs more frequently than other newborn conditions for which newborns are routinely screened.<sup>11</sup> Compared to children with normal hearing, those with hearing loss have more difficulty learning vocabulary, grammar, word order, idiomatic expressions, and other aspects of verbal communication.<sup>12</sup> Hearing loss in children is also associated with delayed language, learning, and speech development, and with low educational attainment.<sup>5</sup> Hearing disorders have also been associated with increased behavior problems, decreased psychosocial well-being, and poor adaptive skills.<sup>13-15</sup>

## **Risk Factors and High Risk Groups**

Risk factors associated with a higher incidence of PCHL include neonatal intensive care unit (NICU) admission for 2 or more days; Usher's syndrome, Waardenburg's syndrome, or other syndromes associated with hearing loss; family history of hereditary childhood sensorineural hearing loss; craniofacial abnormalities; and congenital infections such as cytomegalovirus, toxoplasmosis, bacterial meningitis, syphilis, herpes, or rubella.<sup>5</sup> However, approximately 50% of infants with PCHL do not have any known risk factors.<sup>10, 16-20</sup>

## **Current Clinical Practice**

Practice standards were set in the U.S. by the JCIH in their 2000 and 2007 position statements.<sup>5, 6</sup> In their statements, the JCIH endorsed integrated, interdisciplinary state and national systems of UNHS, evaluation, and family-centered intervention. They recommended that all infants should have access to UNHS and be screened before age 1 month. Infants not passing the screening test should undergo audiologic and medical evaluations before age 3 months, and infants with confirmed hearing loss should receive appropriate intervention before age 6 months (Figure 1). In addition, 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, the medical evaluation, early intervention, surveillance, communication, and tracking.<sup>6</sup>

According to the American Speech-Language-Hearing Association, 39 U.S. states have enacted legislation related to UNHS.<sup>21</sup> The laws are similar in their intent to promote the early identification of hearing loss, but differ in their requirements. These differences include whether screening is mandated, and in what hospitals, or encouraged by the state, how and if the results are required to be reported, and funding mechanisms.<sup>22</sup> In addition to state legislation regarding screening, the federal Individuals with Disabilities Education Act (IDEA) requires states to develop and implement statewide systems of early intervention services for infants and toddlers. This Act requires that infants and toddlers with disabilities be identified and evaluated using risk criteria and appropriate audiologic screening techniques.<sup>21</sup>



Thirty states and territories receive funding from the Early Hearing Detection and Intervention (EHDI) program at the CDC. Some of the funded states have state UNHS legislation while others do not. All states, Puerto Rico, Guam, and the Commonwealth of the Northern Marianas and Palau have received funds to develop and implement UNHS and intervention programs through HRSA's Maternal and Child Health Bureau beginning in 2000.

## Screening Tests and Diagnosis

Hearing screening of newborns involves use of objective physiologic measures. Currently, otoacoustic emissions (OAEs) and/or auditory brainstem response (ABR) are most often used to detect sensory or conductive hearing loss.<sup>21</sup> 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 OAE followed by ABR in those who failed 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 and 0.08, respectively.<sup>17</sup>

Otoacoustic emissions are low intensity sounds from the cochlea resulting from stimulation by audible sounds. The outer hair cells of the cochlea vibrate, and the vibration produces a low intensity sound that echoes back into the middle ear. This sound can be measured with a small probe inserted into the ear canal. The presence of transient middle ear fluid can affect the performance of this test resulting in a non pass result. The ABR is an auditory evoked potential that originates from the cochlea and is not affected by middle ear fluid. It consists of a series of peaks corresponding to the neural response to an auditory stimulus along the auditory pathway between the auditory nerve and the rostral brainstem. Electrodes are placed on the head, and brain wave activity in response to sound is recorded.<sup>21</sup>

Newborn hearing screening is generally well accepted and tolerated by parents. Rates of refusals in a U.S. community-based health system were reported as 7 of 8,707 during the first 10 months of 2007 (Personal communication Providence Health & Services [written] December 6, 2007). Under ideal conditions, instruments designed specifically for newborns can test and record findings on sleeping infants in under 5 minutes.

Infants not passing the newborn screening tests are referred for confirmatory testing for diagnosis of PCHL. Referral rates are lower in programs using dedicated technicians rather than volunteers and students.<sup>23</sup> 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 a more extensive evaluation by an audiologist using behavioral as well as technologic (AOE/ABR) methods. Although the American Academy of Pediatrics has set a standard of 95% for compliance with follow up testing, this rate varies depending on tracking systems and local services.

The procedure at one state with a tracking system includes the following (Personal communication [written] October 6, 2005).<sup>24</sup> At the diagnostic follow-up, the child receive at least a diagnostic OAE evaluation bilaterally and high frequency (1000 Hz) tympanometry. If tests are within normal

limits, the child is no longer followed by the state tracking system unless there are risk factors and/or remarkable history (such as family history, syndromes, eventful NICU stay, etc). If the child does not test within normal limits at the diagnostic follow-up, it is recommended that they received a threshold ABR evaluation. This includes click thresholds and frequency specific information at least at 500 and 4000 Hz, and bone conduction thresholds if necessary (normal range on diagnostic ABR is considered 30 dB hearing level and below). This test should be scheduled and completed by 3 months of age.

## **Treatment**

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.<sup>5</sup> Early intervention includes evaluation for amplification or sensory devices, surgical and medical evaluation, and communication assessment and therapy. Use of cochlear implants in infants has become more available for appropriate candidates in recent years, and is usually considered in those with severe to profound hearing loss after inadequate response to hearing aids.<sup>25-28</sup> Research in neurological and auditory cortical development suggests that early versus late implantation may be linked to more normal cortical auditory pathway development.<sup>29-31</sup>

## **Recommendations of Other Groups**

Recommendations of other groups are summarized in Table 1. Most recommendations support the JCIH 2000 statement<sup>5</sup> specifying UNHS for all newborns, diagnostic testing by age 3 months for those not passing the screening test, and intervention by age 6 months for those with confirmed hearing loss.

## **Analytic Framework and Key Questions**

Evidence reviews for the USPSTF follow a specific methodology<sup>32</sup> beginning with the development of an analytic framework and key questions in collaboration with members of the USPSTF. The analytic framework represents an outline of the evidence review and includes the patient population, interventions, outcomes, and adverse effects of the screening process (Figure 2). Corresponding key questions examine a chain of evidence about the effectiveness and potential adverse effects of UNHS and subsequent early intervention. This systematic review updates the evidence from the prior 2001 USPSTF review for the following key questions:

1. Among infants identified by UNHS who would not be identified by targeted screening, does initiating treatment prior to age 6 months improve language and communication outcomes?

2. Compared with targeted screening, does UNHS increase the chance that treatment will be initiated by age 6 months for average risk infants? For high risk infants?
3. What are the adverse effects of UNHS and early treatment?

For this review, targeted screening indicates selective screening of newborns based on the presence of risk factors or associated conditions. High-risk newborns are those with risk factors known to be associated with PCHL and/or newborns admitted to the NICU.

Selected key questions addressed in the prior report<sup>3,4</sup> were not updated in this report because they were adequately addressed by existing evidence:

- 1a. Can UNHS accurately diagnose moderate to severe sensorineural hearing impairment?  
What are the sensitivity and false negative rate of screening tests?  
What are the specificity, false positive rate, and predictive value of screening tests?
- 1b. Compared with selective screening, how many more cases are identified?

## **II. METHODS**

### **Literature Search and Strategy**

Literature searches were conducted to systematically identify articles addressing the 3 key questions focusing on evidence that was not included in the 2001 USPSTF evidence review (Appendix B1 – Search Strategies). Databases included the Cochrane Central Register of Controlled Trials, Cochrane Database of Systematic Reviews, Database of Abstracts of Reviews of Effects (through the 4<sup>th</sup> Quarter 2007), and Ovid MEDLINE (2000-November 2007 for key questions 1 and 2; 1996-November 2007 for key question 3). Additional articles were obtained from reference lists of related reviews, studies, editorials, reports, websites, and by consulting experts.

### **Inclusion and Exclusion Criteria**

Investigators reviewed abstracts and selected full-text articles based on inclusion and exclusion criteria specific to each key question (Appendix B2 – Inclusion and Exclusion Criteria). Eligible studies addressed key questions and were English-language, conducted in the U.S. or comparable location, and, for screening studies, included infants screened before age 6 months. Key questions 1 and 2 were addressed by controlled trials and observational studies. Key question 3 on adverse effects was addressed by descriptive as well as comparative studies. Results of surveys were included if response rates were >40%. Appendix B3 catalogues a list of studies excluded from the review.

### **Critical Appraisal**

The quality of studies was rated using design-specific criteria developed by the USPSTF (Appendix B4 – USPSTF Quality Rating Criteria).<sup>32</sup> Each study's overall rating considers internal validity and applicability. Descriptive studies without quality criteria were not rated, but are summarized in the text.

### **Size of Literature Reviewed**

A total of 1316 unique citations were identified by the literature searches and from reference lists, etc. (Appendix B5 – Yields from Searches, Abstract Review, and Article Review). Of these, two studies met inclusion criteria for KQ 1, seven met criteria for KQ 2, and eleven met criteria for KQ3.

## **Data Synthesis**

Data from the full text of the original articles and systematic reviews were abstracted to evidence tables (Appendix C). The data included study, year, setting, patient population, inclusion/exclusion criteria, risk status, methods, and results. An outcomes table estimating the number needed to screen under various assumptions was determined using estimates from the most relevant studies.

## **External Review Process**

The USPSTF liaisons advise the Oregon Evidence-based Practice Center in formulating and reporting this systematic review update. An additional set of outside experts have provided feedback on a draft version of the evidence synthesis (Appendix D).

## III. RESULTS

### **Key Question 1. Among infants identified by UNHS who would not be identified by targeted screening, does initiating treatment prior to age 6 months improve language and communication outcomes?**

#### **Summary**

No randomized controlled trials address this question. In a community-based cohort of both high and average risk children with PCHL, those who had their hearing impairment confirmed by age 9 months or younger had better scores at age 8 years on measures of receptive and expressive language, but not speech, than those confirmed later.<sup>33</sup> Children with PCHL who underwent UNHS had better scores than those who did not on measures of receptive language, but not expressive language and speech. More children undergoing UNHS had confirmation of impairment by age 9 months than those not screened as newborns (67% versus 27%; CI 24-56%;  $p < 0.001$ ).

#### **Evidence**

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

A good-quality retrospective cohort study evaluated the effect of UNHS on speech and language outcomes of children with PCHL (Table 2).<sup>33</sup> This study did not evaluate the effects of universal versus targeted newborn screening. It did 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% to 26%), a possible surrogate for risk at birth, was similar between those who had UNHS versus not, and between those confirmed early versus late. The outcome measures are reported as differences between group means preventing calculation of absolute risk reduction or other estimates of magnitude of effect.

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 to 1997, and underwent speech and language assessment at school age (mean 7.9 years; range 5.4-11.7). Included children were either part of the Wessex Trial,<sup>18</sup> constituting 34% of the birth cohort in this study, or from districts in Greater London providing UNHS or not at the time of birth. Estimates of the completeness of ascertainment of eligible children in the cohort exceeded 95%. Study participants with PCHL represented 71% of eligible children in the cohort and were similar to nonparticipants in age, sex, and severity of

hearing loss. It was not reported if there was differential loss to follow up between screened/unscreened or early/late diagnosed groups.

Protocols for screening and confirmation of hearing impairment were similar at all sites, and all children had bilateral impairment of at least 40 dB hearing level.<sup>18</sup> All children were also screened using the Health Visitor Distraction Test at age 7 to 8 months as usual care in the U.K. Therapy was provided for all 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. The z scores represent the number of standard deviations of the distribution of scores in children with normal hearing by which age-adjusted scores of children with hearing impairment differ from the mean scores of children with normal hearing.

Baseline characteristics were similar between comparison groups (sex, English as first language, nonverbal ability using Raven's Progressive Matrices score, age at assessment, degree of hearing loss [moderate 40-69 dB hearing level, severe 70-94 dB hearing level, profound  $\geq 95$  dB hearing level], presence of other disabilities, mother's education, and occupation of head of household). Outcome measures were adjusted for degree of hearing loss, maternal education, and age-adjusted total Raven's Progressive Matrices scores. 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 history.

Children who had their hearing impairment confirmed by age 9 months or younger had better adjusted mean scores at school age than those confirmed later on the Test for Reception of Grammar (adjusted mean difference 0.90;  $p=0.003$ ), British Picture Vocabulary Scale (adjusted mean difference 0.64;  $p=0.02$ ), and Renfrew Bus Story Test sentence information (adjusted mean difference 0.54;  $p=0.03$ ); but not on the Renfrew Bus Story longest sentences component, or Children's Communication Checklist speech scale. 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 intelligence quotient.

Children who underwent UNHS had better adjusted mean scores than those who did not on the Test for Reception of Grammar (adjusted mean difference 0.59,  $p=0.05$ ) and British Picture Vocabulary Scale (adjusted mean difference 0.47,  $p=0.08$ ), but not the Renfrew Bus Story test or Children's Communication Checklist speech scale. More children undergoing newborn screening had confirmation of impairment by age 9 months than those not screened as newborns (67% versus 27%; CI 24-56%;  $p<0.001$ ). Associations between early confirmation or exposure to UNHS and outcomes were similar in the Wessex and Greater London subgroups.

Limitations of the study include 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 if 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 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.

A fair-quality retrospective cohort study conducted in Australia provides speech and language outcomes for a birth cohort exposed to risk based newborn hearing screening.<sup>35</sup> This study examined the relationship of age at diagnosis of PCHL and severity of impairment on several language, speech, and reading measures in children age 7 to 8 years who were fitted with hearing aids by age 4.5 years (Table 2). All children born in the state of Victoria who were identified with PCHL through risk based screening of infants, universally available behavioral hearing screening at age 8 to 10 months, and other referral mechanisms were included. The government provided services for all eligible children and data obtained at the time of services were used in the study. Children with intellectual disability and non-English speakers were excluded from the study.

Several outcome measures were examined using validated methods including receptive and expressive language (Clinical Evaluation of Language Fundamentals), receptive vocabulary (Peabody Picture Vocabulary Test), cognition (Perceptual Organization Index of the Wechsler Intelligence Scale for Children), articulation (Goldman-Fristoe Test of Articulation), reading comprehension (Reading Progress Test 1), intelligibility (teacher questionnaire), and family functioning (McMaster Family Assessment Device). Evaluators were blinded to the children's history and hearing status. Regression models were constructed that controlled for confounders including nonverbal intelligence quotient, maternal education, paternal occupational prestige, and family functioning.

Few children in the cohort were diagnosed with PCHL younger than age 6 months (n=11) or 12 months (n=28). The mean age of diagnosis was 21.6 months and 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. The severity of impairment contributed significantly to the variance on all measures except reading comprehension. Language outcomes were more than 25 points lower than expected from intelligence quotient scores.

Both the Australian and U.K.<sup>33</sup> studies use similar methodologies utilizing population-based birth cohorts, assessment and intervention using government-supported services, and evaluation of speech and language outcomes at ages 7 to 8 years. However, the small number of children diagnosed by age 6 months in the Australian study may provide inadequate power to evaluate the effect of age of diagnosis on outcomes. Other differences include types of services provided, educational systems, and use of different outcome measures.

Several other observational studies report the effects of early intervention programs.<sup>36-44</sup> These studies were reviewed for the previous USPSTF recommendation and are briefly summarized for this update in Table 2. All of these studies have important methodologic limitations including use of convenience samples, nonblinded assessments, and lack of information on attrition and follow-up, among others.<sup>3,4</sup> All of these studies report better outcomes for children with hearing impairment identified and/or treated early versus late. One study specifically examined the effect of UNHS on expressive, receptive, and total language outcomes.<sup>39</sup> Children who were screened had better



outcomes than those not screened, however, potential selection bias and noncomparability of groups limit the conclusions of this study.

## **Key Question 2. Compared with targeted screening, does UNHS increase the chance that treatment will be initiated by age 6 months for average risk infants? For high risk infants?**

### **Summary**

No trials compare targeted screening with UNHS and report data about initiation of early treatment for average or high risk infants. Data from a large nonrandomized trial and descriptive studies indicate that average and high risk infants with PCHL born in hospitals with UNHS have earlier referral and initiation of treatment than those born in hospitals without UNHS. In the nonrandomized trial, one additional case of PCHL was referred before the age of 6 months for every 1,969 (1,011-12,896) infants in the UNHS population.

### **Evidence**

**Wessex Universal Neonatal Hearing Screening Trial.** 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 age 7 to 8 months versus usual care screening alone increases detection and improves early management of infants with PCHL in the U.K.<sup>18</sup> The trial does not measure other benefits to the infants, compare targeted versus universal screening, or compare average versus high risk newborns, but reports characteristics of the cohort.

The trial included all infants born in 4 participating hospitals from 1993 to 1996 including 25,609 born during periods of UNHS and 28,172 not screened as newborns. Two teams of testers (trained nursery nurses) and equipment moved between 2 pairs of hospitals to achieve 4 periods of 4 to 6 months duration with UNHS and 4 without. Most infants were screened within 48 hours of birth. Infants in special-care baby units and neonatal intensive care units (NICU) were screened at the end of their hospital stays. Newborns who screened positive using OAE followed by ABR in those who failed the first test were referred for audiological assessment to determine their hearing level. These are public health services available to all children. All infants were also subjected to screening using the Health Visitor Distraction Test at age 7 to 8 months as usual care.

Results indicate that 87% of all eligible newborns were screened at birth and 8.1% of screened newborns had known risk factors. These include family history of hearing impairment, perinatal infection, birthweight <1.5 kg, anatomical deformity, birth asphyxia, chromosomal abnormality, and exchange transfusion. Sixty-four percent of newborns with PCHL had one or more of these risk

factors (95% CI, 50-77%). Special-care and NICU patients accounted for 36% of infants with PCHL.

The screening protocol had a false positive rate of 1.5% and a false negative rate of 4%. The yield of screening was estimated at 90 cases of PCHL of 40 dB hearing level or more per 100,000 target population (equivalent to 80% of expected prevalence in the population).

Several comparisons between infants undergoing UNHS and those who did not indicated:

- 71 more infants per 100,000 were referred before age 6 months during periods with UNHS versus during periods without. This proportion is equivalent to 19 times higher than nonscreened newborns.
- The odds of confirmation before age 10 months adjusted for severity was 5.0 (1.0-23.0) times greater for screened versus nonscreened newborns.
- The odds of initiating management before age 10 months was 8.0 (1.2-51.0) times higher for screened versus nonscreened newborns.
- Improvement in the ages of confirmation and management were significant specifically for infants with moderate or severe PCHL, but not those with profound impairment.
- The relative risk was 2.3 (1.1-4.7) for detection through newborn screening versus the Health Visitor Distraction Test.

All newborns enrolled in the Wessex Trial were included in an 8-year follow-up study.<sup>17</sup> Long-term follow-up of all screened and nonscreened newborns in the trial allowed more precise calculations of screening performance and effectiveness. Children with abnormal newborn screening tests, abnormal Health Visitor Distraction Tests, or concern for impairment were referred to audiology services. 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, one additional case of PCHL was referred before the age of 6 months for every 1,969 (1,011-12,896) infants in the UNHS population. More children with true PCHL were referred to audiology services prior to age 6 months if they were born during periods with UNHS than during periods without (74% versus 31%; difference 43%; 95% CI 19-60%;  $p=0.001$ ). 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 (2.2-22.0;  $p=0.001$ ). The percentage of all true cases referred was greater at any given age during the first 3 years for children screened as newborns versus not; percentages were similar after age 3 years. The age at referral was lower for children undergoing UNHS versus not (0 months versus 8 months;  $p<0.001$ ). It was noted that 8 children with hearing impairment had screened negative in infancy and 7 had documented progression in severity after detection in infancy. The sum of these 2 figures represents 23% of all cases that might have had progressive losses if the 8 negative screens in infancy had been an accurate reflection of the hearing status of the child at that time.

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, although referrals were early, management was often initiated later than desired (48% after 18 months).

**Descriptive Studies of UNHS Follow-up.** Several descriptive studies report relevant follow-up data from UNHS programs (Table 3).<sup>10, 18-20, 45, 46</sup> The largest and most recent study describes follow-up of hearing impaired infants who were identified during the first phase of a national UNHS program in the U.K.<sup>19</sup> Referred infants with PCHL of 40 dB hearing level or more 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. Infants from the NICU utilized these services at slightly older ages: first follow-up at 9 weeks, diagnosed at 13 weeks, and received hearing aids at 24 weeks. Two studies from the U.S. report partial follow-up information. Hearing impaired infants who underwent UNHS in the Colorado Newborn Hearing Screening Project were referred at a median age of 2.1 months.<sup>10</sup> Infants referred after UNHS in New York State were diagnosed at a median age of 3 months, enrolled in programs at 3 months, and had hearing aids fitted at 7.4 months.<sup>20</sup>

These results contrast with follow-up data for children who did not undergo UNHS. In addition to results of the Wessex Trial described above,<sup>17, 18</sup> descriptive data from parent surveys indicate later ages of diagnosis and initiation of therapy in children not undergoing UNHS. In a national survey of 151 parents with children under age 6 years with hearing impairment in the U.S., children screened as newborns were diagnosed and received hearing aids at younger ages than those not screened.<sup>47</sup> For children with unknown causes for hearing impairment, the median ages of confirmation for screened versus nonscreened children were 4 versus 25 months for mild/moderate impairment, and 2 versus 15 months for severe/profound impairment. The median ages for hearing aid fitting for screened versus nonscreened children were 6 versus 30.5 months for mild/moderate impairment and 4 versus 16 months for severe/profound impairment. A survey of parents of 77 nonscreened children with severe to profound hearing impairment attending specialized educational programs in Illinois indicated the median age of suspicion of hearing loss was 8 months, diagnosis 12 months, and hearing aid fitting 15.5 months.<sup>48</sup>

### **Key Question 3. What are the adverse effects of UNHS and early treatment?**

#### **Adverse Effects of Screening**

Two fair-quality cohort studies,<sup>49, 50</sup> one poor-quality case-control study,<sup>51</sup> and 5 survey studies with >40% response rates<sup>51-55</sup> provided relevant information on adverse effects of newborn hearing screening (Table 4).

In a subset of the Wessex Trial, 100 parents of infants who passed UNHS and 100 parents with infants who did not pass were given questionnaires 2 to 12 months after screening. All infants were

considered low risk for hearing impairment. No differences were found on the Spielberger State-Trait Anxiety Inventory or the Attitudes Toward the Baby Scale between parents with infants who passed and did not pass screening.<sup>49</sup> In another fair-quality cohort study in Austria, using non-validated measures, 85 mothers of infants who did not pass either a one-step or two-step screening test were found to have no significant differences in their levels of concern.<sup>50</sup> In this study, 14% to 21% of parents reported feeling highly concerned, with the mothers whose infants failed the 2nd test showing the highest levels of anxiety.

In a poor-quality case-control study evaluating the impact of false positive results from screening, 2 groups of screen positive infants (failed risk assessment and failed distraction test) and 2 matched control groups were compared.<sup>51</sup> Response rates were 51% or lower in the control groups. The majority of parents whose infants initially screened positive showed relief and improved negative emotions after a normal diagnostic test. However, approximately 20% of parents showed some residual worry 6 months after the normal test.<sup>51</sup>

In a survey of 344 mothers in the U.K. administered 3 weeks and 6 months after newborn hearing screening, there was an increase in worry ( $p < 0.001$ ) and decrease in certainty ( $p < 0.001$ ) as the number of testing recalls increased.<sup>52</sup> Although general understanding of the screening test did not moderate anxiety, mothers who understood that receipt of a positive first test was unlikely to mean that the baby had a hearing loss had lower anxiety ( $p = 0.01$ ) and lower worry ( $p < 0.01$ ) than mothers who did not.

Parents at two university hospitals in Colorado were interviewed following their first newborn hearing screening test that required referral.<sup>53</sup> Although 78% reported not feeling angry about what was happening to them and their child and 81% felt informed, 38% did not feel comforted by hospital staff. Half of parents with a child with a confirmed hearing loss expressed negative emotions including frustration, anger, depression, and confusion.<sup>53</sup>

Mothers of 307 infants participating in the Rhode Island Hearing Assessment Program indicated in a survey that learning about UNHS during hospitalization versus before arriving to the hospital was associated with greater worry.<sup>54</sup> Significantly more mothers with infants who were retested worried about the test results compared to mothers with infants undergoing only one screening test. For mothers of infants requiring retesting, the degree of worry at the time of the retesting was significantly greater than at the first screening test. Factors associated with increased maternal stress about UNHS included mothers who were non-married, bilingual, non-white race, and/or achieving less than a high school education.<sup>54</sup>

In a survey 6 months after UNHS, 87 mothers in Sweden were asked about their feelings about the screening experience.<sup>55</sup> The parents of the 6 of 10 infants that needed to be retested reported anxiety, while the majority were satisfied with the service, thought the information about the procedure was sufficient, and had a positive feeling about the test.<sup>55</sup> A small number of complaints centered around getting information about the test earlier, and the test being too demanding or taking too long.

A survey of 81 Australian mothers with hearing impaired children fitted with hearing aids showed that parents had a generally positive response to ABR screening and mixed responses to the

distraction test.<sup>56</sup> Parents displayed feelings of denial and shock about their child's diagnosis, frustrations in delays in diagnosis, and communication difficulties with providers.<sup>56</sup>

## **Adverse Effects of Early Treatment**

No studies addressed the potential adverse effects of early intervention using hearing aids or other amplification, American Sign Language, English instruction, speech and language therapy, or family education and support.

A growing literature about cochlear implantation in infants and children includes descriptive information about adverse effects. Case series reports of cochlear implantation indicate few surgical complications in children. A series of 300 children receiving cochlear implants in Greece, ranging from age 1 to 17 years, reported an overall rate of 2.3% for major surgical complications and 16% for minor complications.<sup>57</sup> There were no surgical complications among children under age 12 months in small U.S. case series,<sup>58,59</sup> or in those under age 2 years in a small U.K. case series.<sup>60</sup>

The FDA released public health notifications about the increased risk of bacterial meningitis in children with cochlear implants, with highest risks among those using implants with positioners.<sup>61</sup> A positioner is a wedge inserted next to the implanted electrode to facilitate transmission. Implants with the positioner were voluntarily recalled in the U.S. in 2002. Among children who had cochlear implantation from 1997 to 2002 at less than age 6 years, 41 episodes of postimplantation bacterial meningitis occurred among 38 children.<sup>62,63</sup> Of these, 71% had implants with positioners. Although the rate of meningitis decreased as the time after implantation increased, 20% of cases occurred after 24 months or more and were found exclusively in children with positioners.<sup>63</sup> None of the children with meningitis received their implants at less than age 12 months and rates of infection did not indicate age-related risks.

Although several studies of psychological issues relating to cochlear implantation in children, such as parental distress, have been published, only one specifically focused on young children. Parents of 28 children (age 12 to 30 months) undergoing cochlear implantation in Turkey noted on questionnaires that making the decision for cochlear implantation was stressful.<sup>64</sup> 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 outcomes table model (Table 5). Assumptions for the model include proportion of newborns considered high risk,<sup>17</sup> prevalence of PCHL in high risk and average risk populations,<sup>3</sup> proportion not screened in the

hospital,<sup>65</sup> sensitivity of 2-stage screening,<sup>17</sup> compliance with follow up testing (estimated), accuracy of diagnostic tests,<sup>3</sup> and proportion of average risk newborns diagnosed with PCHL by 3 months (estimated). Using these assumptions, if 10,000 newborns underwent UNHS, there would be 11 to 12 diagnosed cases by age 3 months, 86 false positive screening tests, and possibly 1 missed case. The number needed to screen (NNS) to diagnose one case would be 878. If only high-risk newborns underwent screening, there would be 4 or 5 diagnosed cases, 6 false positive screening tests, and 8 or 9 missed cases. The NNS to diagnose one case would be 178.

## IV. DISCUSSION

### Summary of Review Findings

Evidence addressing the 3 key questions in this review is summarized in Table 6.

A good-quality community-based cohort study of both high and average risk children with PCHL indicated that those who had early versus late confirmation and those who had UNHS versus none had better language scores at age 8 years.<sup>33</sup> In this study, 67% of children undergoing UNHS had confirmation of impairment by age 9 months compared to 27% of those not undergoing UNHS. In contrast, a fair-quality community-based cohort study of children with hearing impairment who did not undergo UNHS indicated no relationship between age at diagnosis and language, speech, and reading measures at age 7 to 8 years.<sup>35</sup> Few children were diagnosed by age 6 months in this cohort.

These studies provide stronger evidence for the long-term benefits of UNHS than previous studies.<sup>3,4</sup> Although previous studies were consistent in reporting improved outcomes for children diagnosed early versus late, all had important methodologic limitations.<sup>3,4</sup> The use of large community-based birth cohorts providing prospectively collected data, blinded assessments of validated outcome measures, and adjustment for confounders improves the internal validity and generalizability of the cohort studies.<sup>33,35</sup>

A good-quality nonrandomized trial of a large birth cohort of both high and average risk newborns indicates that infants identified with PCHL through UNHS have significantly earlier referral, diagnosis, and treatment than those identified in other ways.<sup>17,18</sup> These findings are corroborated by multiple descriptive studies of ages of referral, diagnosis, and treatment including reports of UNHS program follow-up measures,<sup>10,20,46</sup> historical comparisons,<sup>16,66,67</sup> and comparisons between screened and non-screened children.<sup>47,48</sup>

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. In the first phase of a national UNHS program in the U.K., well infants had a median age of first follow-up at age 4 weeks, diagnosis at 10 weeks, enrollment in education services at 10 weeks, and hearing aid fitting at 14 weeks.<sup>19</sup> Data from populations not undergoing UNHS indicate age of diagnosis typically between 15 to 24 months with treatment following several months later.<sup>16,47,48,66,67</sup> In the absence of UNHS, children with more severe hearing impairment are generally diagnosed and treated earlier than children with less severe impairment.<sup>47,66,68</sup>

Limited follow-up data exist specifically for high risk infants who could be identified by targeted screening. Approximately half of infants with PCHL identified by UNHS have risk factors for hearing impairment.<sup>10,17-20</sup> Patients of NICUs undergoing UNHS had slightly later ages of

confirmation and treatment compared to well infants in the U.K. study,<sup>19</sup> potentially relating to the complexity of co-existing health problems.

Since the previous review, efforts have been made to study both the short and long-term effects of UNHS on a variety of outcomes including negative emotions, parental worry and anxiety, and attitudes toward infants. Recent cohort studies indicate no significant differences in measures of concern, anxiety, and parental attitudes for families with newborns who pass versus those who do not pass the newborn screening test. No studies addressed the adverse effects of a child with PCHL being screened or diagnosed late.

Other studies indicate that usual parental reactions to an initial non-pass on the screening test include worry, questioning, and distress. Negative emotions resolve for most parents when a diagnostic test is provided with a normal result. Although some parents show residual worry months after a normal test, most concern improves over time. Parents of children with confirmed hearing loss show greater levels of frustration and confusion than those parents of normal hearing children. These parents may need increased support and comfort during the screening process and at the time of diagnosis. Information on the screening test, timely access to appropriate follow-up testing and intervention, and integrated and individualized family services and support within the healthcare system have been linked to a positive UNHS experience for parents.

Hearing screening programs are generally well accepted and tolerated by the parents of newborns and have demonstrated cost-effectiveness.<sup>69</sup> With legislation for UNHS being enacted in most U.S. states over recent years, screening practices and procedures have become routine in the postpartum hospital setting. Technicians and parents have become more comfortable with the routine practice of UNHS and show less anxiety about such procedures than when they were first introduced (Personal communication [written] October 6, 2003).<sup>24</sup>

Less has been published about the adverse effects of early interventions. To fully address this question, studies would compare adverse outcomes between children initiating treatment at age 6 months as current practice standards advise versus those initiating treatment later. Currently available studies comparing early versus late interventions focus on benefits. Now that practice standards exist, it may 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 in the U.S. is that they were conducted outside the U.S. Although the method of screening and the inpatient maternity experience are likely similar, the processes of referral, follow-up, and treatment would be expected to differ. Differences can be attributed to many factors such as dissimilar health care systems, practice patterns, coverage, access, educational systems, and populations. Currently, there is no standard method in the U.S. to track children through these processes to ultimately obtain language outcomes on a birth cohort as done in the U.K. study,<sup>33</sup> although approaches to do so are being piloted. Factors influencing follow-up and treatment in the U.S. would need to be considered as well as exposure to UNHS when determining long-term outcomes.



## Future Research

More studies of long-term functional outcomes related to UNHS are needed to support the findings of the U.K. study.<sup>33</sup> Other functional outcomes, such as school performance, social interactions, and quality of life, may be more relevant to children and their families and future research should include these also. Studies conducted among different populations with dissimilar health systems would help determine the generalizability of the U.K. results. Research from UNHS programs can be utilized to identify best practices and guide process and quality improvement efforts. Standardization of nomenclature, methods, and measures would allow collaborative research nationally and internationally.

## Conclusions

Universal newborn hearing screening is recommended as a practice standard by professional organizations and is mandated in 39 U.S. states.<sup>21</sup> Screening techniques have high performance characteristics and can be performed on a sleeping newborn in less than 5 minutes. False positive rates have been reduced by using 2-step screening and repeat screening for newborns failing to pass the first test. Screening has been implemented as routine newborn care in many U.S. hospitals and rates of refusals are low. The feasibility of screening in the context of community practice has been demonstrated for average risk newborns with short lengths of stay as well as high risk patients in NICUs. Methods of referral, diagnosis, and treatment are more variable than screening, and are dependent on system processes, insurance coverage, community practice, and social and economic barriers, among other influences.

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.

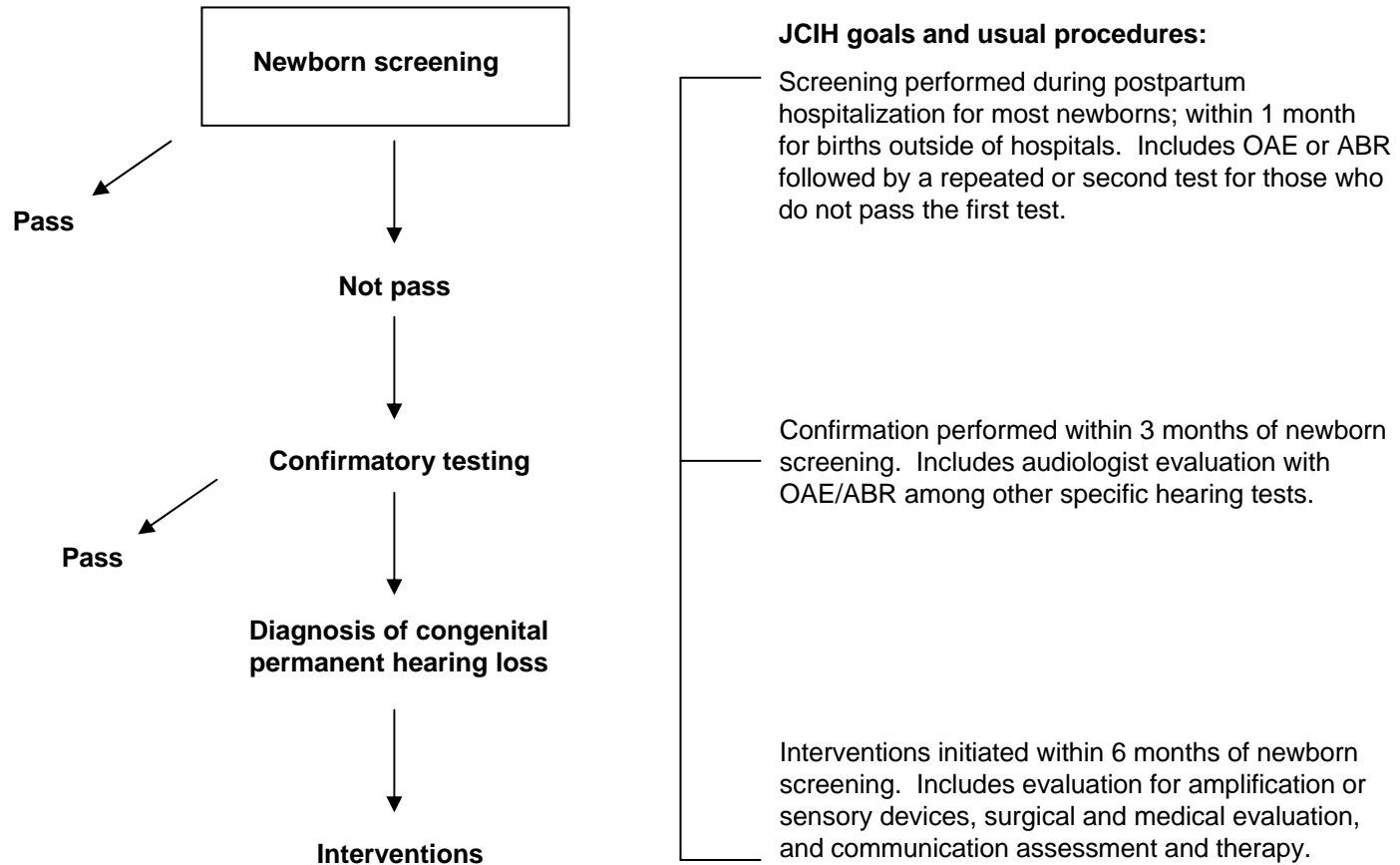
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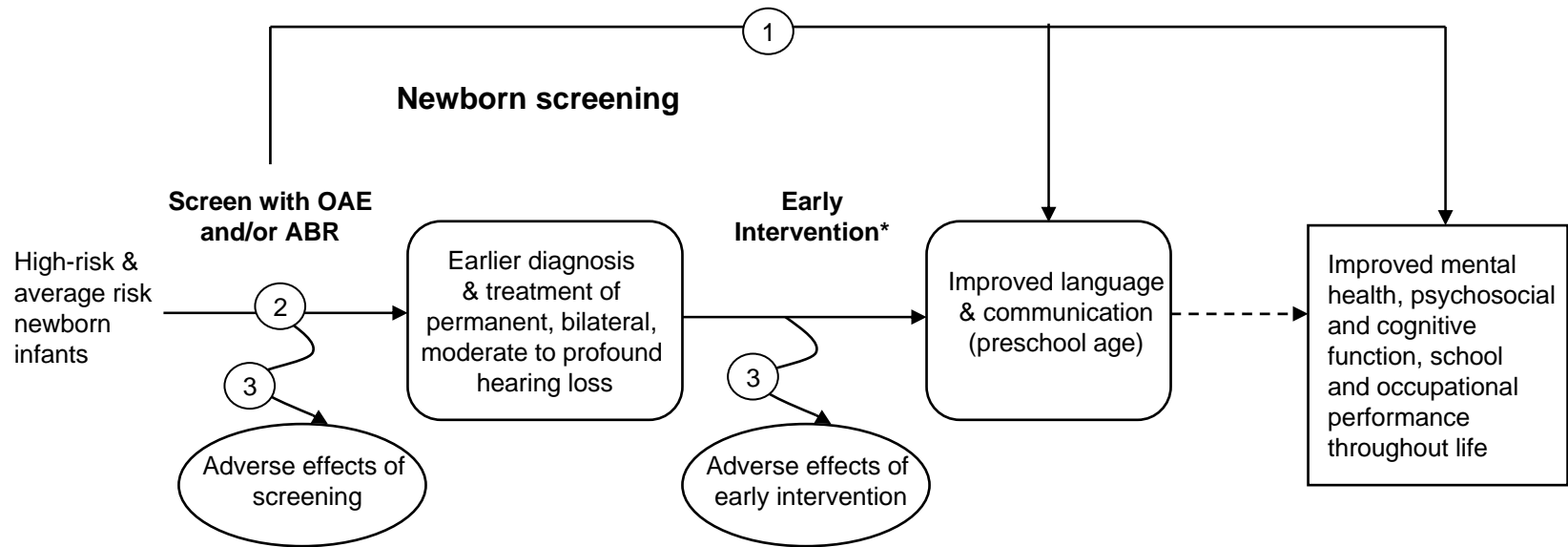
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**Figure 1. Process of Screening and Follow-up**



**Abbreviations:** ABR, auditory brainstem response; JCIH, Joint Committee on Infant Hearing; OAE, otoacoustic emissions.

**Figure 2. Universal Newborn Hearing Screening Analytic Framework and Key Questions**



**Key Questions**

- (1) Among infants identified by UNHS who would not be identified by targeted screening, does initiating treatment prior to 6 months of age improve language and communication outcomes?
- (2) Compared with targeted screening, does UNHS increase the chance that treatment will be initiated by 6 months for average risk infants? For high-risk infants?
- (3) What are the adverse effects of UNHS and early treatment?

\*Hearing aids or other amplification, cochlear implants, American Sign Language and/or English instruction, speech & language therapy, family education & support.

**Abbreviations:** ABR, auditory brainstem response; OAE, otoacoustic emissions; UNHS, universal newborn hearing screening.

**Table 1. Recommendations of Other Groups**

Group, Year	Recommendation
Joint Committee on Infant Hearing (JCIH), 2000 <sup>5, 6</sup>	Endorses early detection and intervention for infants with hearing loss through integrated, interdisciplinary state and national systems of UNHS, evaluation, and family-centered intervention. Statement includes the following guidelines: <i>Newborn screening:</i> Screening during hospital stay for those receiving routine or NICU care; screening before age 1 month for those born in alternative birthing facilities. <i>Diagnosis:</i> Appropriate audiologic and medical evaluations to confirm the presence of hearing loss before age 3 months for those not passing screening tests. <i>Interventions:</i> All infants with confirmed permanent hearing loss receive services before age 6 months in interdisciplinary intervention programs that recognize and build on strengths, informed choice, traditions, and cultural beliefs of the family.
CDC's Early Hearing Detection and Intervention (EHDI) Program, 2006 <sup>70</sup>	Supports the NIH, JCIH, and AAP positions in recommending UNHS before hospital discharge, diagnostic evaluation before age 3 months, and initiation of appropriate intervention services before age 6 months. The age of a child when a hearing loss is diagnosed is important to the development of the child's speech, language, cognitive, and psychosocial abilities. Without universal screening by age 1 month, the average age at which hearing loss is identified in children is 2 to 3 years.
American Academy of Pediatrics (AAP) Task Force on Newborn and Infant Hearing, 2000 <sup>5, 6</sup>	Supports the JCIH recommendations.
NIH (NIDCD - National Institute on Deafness and Other Communication Disorders), 2006 <sup>71</sup>	Supports the JCIH recommendations.
American Speech-Language-Hearing Association (ASHA), 2006 <sup>21</sup>	The following states have passed legislation for UNHS: Arkansas, California, Colorado, Connecticut, Delaware, Florida, Georgia, Illinois, Indiana, Iowa, Kansas, Kentucky, Louisiana, Maine, Maryland, Massachusetts, Mississippi, Missouri, Montana, Nebraska, Nevada, New Hampshire, New Jersey, New Mexico, New York, North Carolina, Ohio, Oklahoma, Oregon, Pennsylvania, Rhode Island, South Carolina, Texas, Utah, Virginia, Washington D.C., West Virginia, Wisconsin, and Wyoming.
European Consensus Development Conference on Neonatal Hearing, 1999 <sup>72</sup>	Identification by UNHS at or shortly after birth has the potential to improve quality of life and opportunities for those affected. Targeted newborn testing on only the 6-8% of infants at increased risk (e.g., NICU and family history of hearing impairment) reduces costs but cannot identify more than 40-50% of cases. Targeted newborn hearing screening in parallel with 7-9 month behavioral testing is more expensive and less effective than UNHS. Risks associated with UNHS include anxiety from false-positive results and possible delayed diagnosis from false-negative results, but these risks are acceptable in view of expected benefits.
National Deaf Children's Society, UK, 2006 <sup>73</sup>	Deaf children need to develop fluent language skills in order to understand and influence the world around them, by whichever communication approach is most appropriate for them. Deaf children, young people and their families should have access to high quality services that offer diversity and choice and meet the needs of the individual child.

**Table 2. Cohort Studies Reporting Language Outcomes**

<b>Study, Year (Quality)</b>	<b>Selection of Subjects</b>	<b>Comparability and Maintenance of Early vs. Late Groups</b>	<b>Adjustment for Confounders</b>	<b>Results</b>
Kennedy et al, 2006 <sup>33</sup> (Good)	120 children with bilateral permanent hearing impairment identified from a large birth cohort of 157,000 children in southern England assessed at a mean of 7.9 years of age (range 5.4 to 11.7). Children were either part of the Wessex Trial or from districts in Greater London providing UNHS or not at the time of birth. 63 age-matched children with normal hearing were used to derive z scores for outcome measures. Children with known postnatal causes were excluded.	Baseline characteristics were similar between comparison groups (sex, English as first language, nonverbal ability, age at assessment, degree of hearing loss, other disabilities, mother's education, and occupation of head of household). Follow-up and attrition likely similar between groups because all were provided assessment, therapy, and follow-up as a public health service.	Outcome measures were adjusted for confounders using multiple linear regression for degree of hearing loss, maternal education, and age-adjusted nonverbal ability.	Children with hearing impairment confirmed by age 9 months or younger 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 a speech scale. All aggregate scores for receptive and expressive language were significantly better for the early confirmation group. 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.
Wake et al, 2005 <sup>35</sup> (Fair)	88 children age 7 to 8 years born in Victoria, Australia who were fitted with hearing aids by age 4.5 years for congenital hearing impairment. Services were provided by the government for all eligible children and data were collected. Hearing screening included audiology referral for infants with risk factors and a universally available behavioral hearing screen at age 8 to 10 months. Children with intellectual disability and non-English speakers were excluded from the study.	Few children were diagnosed younger than age 6 months (n=11) or 12 months (n=28); comparisons between early vs. late diagnosed children were not reported.	Outcome measures were adjusted for confounders using multiple regression for non-verbal IQ, maternal education, paternal occupational prestige, and family functioning.	Age at diagnosis did not contribute significantly to variance on language, speech, or reading measures except receptive vocabulary; severity of impairment contributed to variance on all measures except reading comprehension. Language outcomes were more than 25 points lower than expected from IQ scores and were related to the severity of impairment, but not age at diagnosis. Age at diagnosis was negatively correlated with severity at diagnosis.



**Table 2. Cohort Studies Reporting Language Outcomes**

<b>Study, Year (Quality)</b>	<b>Selection of Subjects</b>	<b>Comparability and Maintenance of Early vs. Late Groups</b>	<b>Adjustment for Confounders</b>	<b>Results</b>
<i>Reviewed for prior report:</i>				
Apuzzo and Yoshinaga-Itano, 1995 <sup>44</sup> (Fair)	Convenience sample of 69 high-risk infants diagnosed between ages 2 and 25 months. Children with severe cognitive delay were excluded.	Late-identified group was more likely to have severe to profound hearing loss (65% vs. 50%). No report of attrition or follow-up rates.	One-way ANOVA did not adjust for SES, family involvement, or other potential confounders.	At age 40 months, infants identified before age 2 months had higher mean Minnesota Child Development Inventory (MCDI) scores for expressive language (p<0.01).
Calderon and Naidu, 2000 <sup>37</sup> (Fair)	Cohort of 80 children with profound hearing loss enrolled in Early Child Hearing Intervention (ECHI) in Seattle Washington. Children with developmental delay were excluded. Cohort grouped by 3 levels by age of entry into program: <1 year (n=9), 12-24 months (n=39), >24 months (n=32). The method of sampling is not described, but the design excluded patients who entered the program but did not graduate.	Not reported. Late diagnosed group had less severe to profound loss (36% vs. 66%). Overall loss to follow-up not reported. Because the early-diagnosed group was in the program longer, they had more opportunity to drop out, so a differential loss to follow-up is likely.	Controlled for degree of hearing loss, degree of outcome impairment that was present upon entry into program (baseline test levels).	At age 3 years, age at entry to program explained 43.5% of the variance in receptive language and 49% of the variance in expressive language. Children treated before age 2 years had better outcomes than those treated after age 2 years. Only 3 children entered the program prior to age 6 months.
Mayne et al, 2000 <sup>43</sup> (Poor)	Convenience sample of 113 children ages 24 to 73 months, divided into those diagnosed before and after age 6 months. The number of low-risk infants and the role of UNHS in identifying subjects are not described. Overlap of sample with previous CHIP studies was not reported.	Demographic comparisons of the groups were not reported. No report of attrition of follow-up rates.	Regression analysis adjusted for degree of hearing loss, mode of communication, other disabilities, parents' hearing, CQ, mother's education, ethnicity, SES.	At ages 24 to 36 months, age at diagnosis explained 23% of the variance in expressive language scores.

**Table 2. Cohort Studies Reporting Language Outcomes**

Study, Year (Quality)	Selection of Subjects	Comparability and Maintenance of Early vs. Late Groups	Adjustment for Confounders	Results
<i>Reviewed for prior report:</i>				
Moeller, 2000 <sup>36</sup> (Fair)	Convenience sample of 112 5-year-olds who completed the Diagnostic Early Intervention Program in Lincoln, Nebraska. Children with non-verbal IQ <70 and those who did not participate in program through age 5 were excluded. The number of low-risk infants and the role of UNHS in identifying subjects are not described. Outcome assessments were made pre- and post-intervention.	Not reported. No report of attrition or follow-up rates. Early identified children may have more opportunity to drop out, although differential drop out may be less of a problem at 5 years than in studies assessing closer to enrollment.	Multiple regression analysis adjusted for family involvement, degree of hearing loss and non-verbal IQ.	At age 5 years, family involvement accounted for 57% of variance in vocabulary and age of enrollment accounted for 11.5%. Adjusted mean vocabulary and reasoning scores were within normal range among children enrolled prior to age 11 months but were lower for later-identified children (11 to 23 months 0.69 SD lower, 24 to 35 months 0.99 SD lower).
Yoshinaga-Itano and Apuzzo, 1998 <sup>40</sup> (Poor)	Convenience sample of 40 high-risk infants, divided into those identified and treated before age 6 months (n=15) and those treated after age 18 months (n=25). Children with severe cognitive delay were excluded (DQ<60).	Late-identified group was more likely to have severe to profound hearing loss (52% vs. 47%). No report of attrition or follow-up rates.	Sex, severity of hearing loss, cognitive function, and other disabilities were examined in 2-way ANCOVAs, not multiple regression (no simultaneous adjustment for multiple confounders).	At age 40 months, infants identified before age 6 months had better adjusted mean MCDI scores for expressive language (81.1 vs. 64.3, p<0.05 ) and receptive language (84.4 vs. 70.1, p<0.05).
Yoshinaga-Itano and Apuzzo, 1998 <sup>41</sup> (Poor)	Convenience sample of 82 infants, ages 19 to 36 months, with mild to profound hearing loss, divided into those identified before age 6 months (n=34) and between ages 7 and 18 months of age (n=48). Early group identified by high-risk registry; late group by usual care. Children with severe cognitive delay were excluded (DQ<60).	Late-identified group was more likely to have severe to profound hearing loss (77% vs. 42%). No report of attrition or follow-up rates.	Sex, severity of hearing loss, cognitive function, and other disabilities were examined in 2-way ANCOVAs, not multiple regression (no simultaneous adjustment for multiple confounders).	At age 26 months, infants identified before age 6 months had better adjusted mean MCDI scores for expressive language (76.2 vs. 56.6, p=0.001), receptive language (82.1 vs. 58.3, p=0.002), MacArthur CDI adjusted mean receptive vocabulary (200 vs. 86.4, p<0.001), and expressive vocabulary (117 vs. 54, p<0.03).

**Table 2. Cohort Studies Reporting Language Outcomes**

Study, Year (Quality)	Selection of Subjects	Comparability and Maintenance of Early vs. Late Groups	Adjustment for Confounders	Results
<i>Reviewed for prior report:</i>				
Yoshinaga-Itano et al, 1998 <sup>42</sup> (Poor)	Convenience sample of 150 children ages 13 to 36 months with mild to profound hearing loss, divided into those identified before (n=72) or after (n=78) age 6 months. The number of low-risk infants and the role of UNHS in identifying subjects are not described. Selection bias is likely because the design probably excluded infants who were diagnosed to have hearing loss but did not enter the program, or who entered, but were lost to follow-up.	At baseline, compared groups differed in some demographic characteristics and in the proportion of subjects with cognitive impairment and severe to profound hearing loss (CQ <80, 29% early group vs. 56% late group; severe to profound hearing loss 34% early group vs. 46% late group). No report of attrition or follow-up rates.	There was stratification by CQ (<80 vs. >80). Other covariates (sex, minority status, maternal education level, Medicaid status, severity, mode of communication, other disabilities) were examined singly in 2-way ANCOVAs.	At ages 13 to 36 months, adjusted mean MCDI receptive language LQ was higher for those identified before age 6 months (79.6 vs. 64.6, p<0.001). Mean MCDI expressive LQ was higher (78.3 vs. 63.1, p<0.001) and total language (79 vs. 64, p<0.001) was higher in early-identified group. No differences in LQ among 4 age of identification levels in late-identified group.
Yoshinaga-Itano et al, 2000 <sup>39</sup> and 2001 <sup>38</sup> (Poor)	Children born in a hospital with a UNHS program in effect at time of birth (n=25) were compared to children born in a hospital without a UNHS program (n=25). All subjects had been enrolled in CHIP program. Eligibility for the screened group was determined by the availability of an assessment of language outcomes. The creation of the study groups and description of the patients limited the conclusions that could be drawn.	The exposure was birth at a hospital with a UNHS program, not age of identification. Because the groups were drawn from different hospitals and time periods, factors other than exposure to UNHS might have influenced outcomes. Selection of subjects and assessment of outcome were unblinded, and neither the number of excluded subjects, nor the reasons for exclusion, are reported.	Pairs matched on age of testing (9-61 months), degree of hearing loss (mild, moderate, moderately severe, profound), and CQ.	Mean scores (SE) for expressive, receptive, and total language were within normal range for the screened group and 18 to 21 points higher (p<0.001) than the unscreened group (expressive language 82.9 [3.7] vs. 62.1 [4.3]; receptive language 81.5 [3.7] vs. 66.8 [4.0]; total language 82.2 [3.3] vs. 64.4 [3.9]). Language development was within normal range for 56% of the screened group compared to 24% of the unscreened group.

**Abbreviations:** ANCOVA, analysis of covariance; ANOVA, analysis of variance; CHIP, Children's Health Insurance Program; CQ, cognitive quotient; DQ, developmental quotient; ECHI, Early Child Hearing Intervention; LQ, language quotient; MCDI, Minnesota Child Development Inventory; SE, standard error; SES, socioeconomic status; UNHS, universal newborn hearing screening.

**Table 3. Descriptive Studies of Universal Newborn Hearing Screening Follow-up**

<b>Author, Year, Location</b>	<b>Program Description</b>	<b>Age Screened</b>	<b>Number of Cases</b>	<b>Proportion High Risk</b>	<b>Age Referred</b>	<b>Age at First Follow-up</b>	<b>Age Diagnosed</b>	<b>Age Enrolled in Programs</b>
Uus and Bamford, 2006 <sup>19</sup> UK	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 $\geq 40$ dB hearing level.	Before hospital discharge	169	54% with risk factors*	At screening	Well infants: median age 4 weeks; NICU: 9 weeks	Well infants: median age 10 weeks; NICU: 13 weeks	Median age 10 weeks for education services
Joseph, 2003 <sup>45</sup> Singapore	UNHS of 4,387 newborns in 1999-2001 at 1 hospital using OAE. Positives were rescreened at ~2 weeks, and again at 6 weeks if needed. Referred if specific criteria not met.	Most within 24 hours	8	38% high risk	Those that tested positive at 6 weeks were referred for formal evaluation	NR	7 of 8 by 7 months	Interventions in place by age 9 months for 4
Bailey, 2002 <sup>46</sup> Australia	UNHS of 12,708 newborns in 5 hospitals in 2000-2001 using OAE and ABR if needed. Referred $\geq 35$ dB hearing level.	Before hospital discharge	9	5 NICU; 8 with risk factors	NR	NR	NR	NR
Mehl, 2002 <sup>10</sup> 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 $>35$ dB hearing level in 1 or both ears.	Before hospital discharge	291 (71% bilateral)	47% with risk factors	Median age 2.1 months	NR	NR	NR

**Table 3. Descriptive Studies of Universal Newborn Hearing Screening Follow-up**

<b>Author, Year, Location</b>	<b>Age of Hearing Aid Fitting</b>
Uus and Bamford, 2006 <sup>19</sup> UK	Well infants: median age 14 weeks; NICU: 24 weeks
Joseph, 2003 <sup>45</sup> Singapore	NR
Bailey, 2002 <sup>46</sup> Australia	6 had hearing aids by 6 months; 1 at 19 months
Mehl, 2002 <sup>10</sup> Colorado	NR

**Table 3. Descriptive Studies of Universal Newborn Hearing Screening Follow-up**

Author, Year, Location	Program Description	Age Screened	Number of Cases	Proportion High Risk	Age Referred	Age at First Follow-up	Age Diagnosed	Age Enrolled in Programs
Dalzell, 2000 <sup>20</sup> 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 age 3 months: younger for well babies and for severe or profound impairment	Median age 3 months
Wessex UNHS Trial Group, 1998 <sup>18</sup> UK	UNHS arm of the Wessex Trial including 25,609 newborns screened in 1993-1996 with OAE and ABR if needed. Referred ≥40 dB hearing level.	3	23	41% special care units; 74% had risk factors	All by age 6 months	NR	67% by age 10 months	63% by age 10 months

\*Includes NICU for > 48 hours, family history of hearing impairment, craniofacial anomaly.

**Abbreviations:** ABR, auditory brainstem response; dB, decibels; NICU, neonatal intensive care unit; OAE, otoacoustic emissions test; UNHS, universal newborn hearing screening.

**Table 3. Descriptive Studies of Universal Newborn Hearing Screening Follow-up**

<b>Author, Year, Location</b>	<b>Age of Hearing Aid Fitting</b>
Dalzell, 2000 <sup>20</sup> New York	Median age 7.5 months

Wessex  
UNHS Trial  
Group,  
1998<sup>18</sup>  
UK

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\*Includes NICL

**Abbreviations**

**Table 4. Studies of Potential Adverse Effects of Screening**

<b>Study, Year (Quality)</b>	<b>Study Design</b>	<b>Subjects</b>	<b>Screening</b>	<b>Setting</b>	<b>Results</b>
Kennedy, 1999 <sup>49</sup> (Fair)	Retrospective cohort	Parents of average-risk newborns: 100 passed UNHS and 100 did not pass; subset of Wessex Trial.	1. OAE 2. ABR	UK; 2-12 months after UNHS	No differences in scores on the Spielberger State-Trait Anxiety Inventory & Attitudes Toward the Baby Scale between parents of newborns who passed UNHS versus not passed.
Weichbold, 2001 <sup>50</sup> (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 2nd screening, 42% were not concerned, 37% were slightly concerned, and 21% were highly concerned. Differences in proportions between groups were not statistically significant.
Poulakis, 2003 <sup>51</sup> (Poor)	Case control	Parents of infants: 108 at risk for hearing impairment; 2. 64 controls for Group 1; 3. 103 failed distraction test; 4. 53 controls for Group 3.	1. Distraction test	Australia	Parent concerns about language development, general development, and perceived vulnerability to ill health did not differ among the 4 groups. Approximately 18% of parents continued to feel worried 6 months after the definitive hearing testing. 6% rated the test procedures as somewhat difficult/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 ( $p<0.05$ ).
Crockett, 2006 <sup>52</sup>	Survey (questionnaire)	Parents of 722 screened newborns (53% response rate) 1. 103 with 1 or 2 negative tests; 2. 81 with 3rd negative test; 3. 105 with 3rd test positive in 1 ear; 4. 55 with 3rd test positive in both ears.	OAE; ABR final test	UK; 3 weeks & 6 months post screening	Significant trends for increased anxiety ( $p<0.05$ ), increased worry ( $p<0.001$ ), and decreased certainty ( $p<0.001$ ) as number of tests increased. Parents in Group 4 who understood test implications had lower anxiety ( $p=0.01$ ) and lower worry ( $p<0.01$ ) versus those who did not.
de Uzategui, 1997 <sup>53</sup>	Survey (questionnaire)	Parents of 201 screened newborns who were referred for further testing (51% response rate).	Not reported	Colorado, USA; 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.



**Table 4. Studies of Potential Adverse Effects of Screening**

Study, Year (Quality)	Study Design	Subjects	Screening	Setting	Results
Hergils, 2000 <sup>55</sup>	Survey (questionnaire)	Parents of 83 screened newborns (95% response rate).	OAE	Linkoping, Sweden; well baby visit, 5-6 months old	76 were satisfied with screening, 3 neutral, 3 dissatisfied, 1 did not know. Screening raised questions for 28 and no questions for well baby visit, 5-6 months old. 44. 79 were positive about the test and 4 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 newborn. Complaints included getting information about the test earlier and test methodology. Parents of 6 of the 10 infants needing retesting reported anxiety.
Russ, 2004 <sup>56</sup>	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 a generally positive response 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 were also reported.
Vohr, 2001 <sup>54</sup>	Survey (interview)	Mothers of 307 screened newborns (85% response rate); Mothers of 40 newborns needing rescreening (90% response rate).	OAE	Rhode Island, USA	Significantly more mothers with infants who were rescreened worried about the test results compared to mothers with infants undergoing only one screening (p<0.001). 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<0.001). Greater worry at the initial screening was seen in mothers with less than high school education (p=0.003) and who were bilingual (p=0.006), non-married (p=0.02), and non-white race (p=0.005). Learning about screening during hospitalization versus before arriving was also associated with greater worry (p=0.012).

**Abbreviations:** ABR, auditory brainstem response OAE, otoacoustic emissions; UNHS, universal newborn hearing screening.

**Table 5. Yield of Screening in a Hypothetical Cohort of 10,000 Newborns for Moderate to Profound PCHL**

Relevant Factors	Probability or effect size	Hypothetical Model	
		UNHS	High-risk screening*
<b>Assumptions</b>			
Proportion high risk <sup>17</sup>	0.08		
Target group for screening		10000	800
Prevalence <sup>3</sup>			
High risk group	0.008		
Average risk group	0.0008		
Miss rate for UNHS (proportion not screened in hospital; estimate)			
In high risk	0.05		
In average risk	0.05		
Follow-up rate for misses	0.9		
Miss rate for high-risk screening			
In high risk <sup>65</sup>	0.23		
Follow-up rate for misses	0		
Sensitivity of 2-stage screening <sup>17</sup>	0.92		
Specificity of 2-stage screening <sup>17</sup>	0.99		
Compliance with follow-up (estimate)	0.9		
Accuracy of diagnostic ABR <sup>3</sup>			
Sensitivity	1		
Specificity	0.995		
Proportion of low-risk diagnosed by 3 months without screening (estimate)	0.1		
<b>Results</b>			
Number 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 months		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 number of cases		13	13
False positive screening tests		86	6
Normal infants incorrectly diagnosed to have PCHL at first post-hospital audiologic examination		<1	<1
NNS to diagnose 1 case		878	178
NNS to diagnose 1 additional case by 3 months		1333	NA

\*High risk defined by risk factors (family history of hearing impairment, perinatal infection, low birthweight, anatomical deformity, birth asphyxia, chromosomal abnormality, exchange transfusion).

**Abbreviations:** ABR, automated brainstem response; NA, not applicable; NNS, number needed to screen; PCHL, permanent congenital hearing loss; UNHS, universal newborn hearing screening.

**Table 6. Summary of Evidence**

Review Year	Conclusions	Study Designs	Quality of Evidence
<b>Key Question 1. Compared with targeted screening, does universal screening increase the chance that treatment will be initiated by 6 months for average risk infants? For high risk infants?</b>			
<i>Initial 2000 review:</i>	<i>UNHS leads to earlier identification and treatment of infants with hearing loss.</i>		
Updated review:	Confirms that children identified with hearing loss through UNHS have earlier referral, diagnosis, and treatment than those identified in other ways. Direct evidence comparing targeted vs. universal screening and average vs. high risk infants is not available.	Controlled trial, cohort study, descriptive data	A good-quality nonrandomized trial of a large birth cohort of both high and average risk newborns compared UNHS to usual care. Those undergoing UNHS had earlier referral, diagnosis, and initiation of treatment than those who did not. Other descriptive studies of UNHS support these results.
<b>Key Question 2. Among infants identified by universal screening who would not be identified by targeted screening, does initiating treatment prior to 6 months of age improve language and communication outcomes?</b>			
<i>Initial 2000 review:</i>	<i>Inconclusive</i>	<i>Cohort study, descriptive data</i>	<i>Several poor and fair-quality studies suffer from selection bias and baseline differences between compared groups. These studies did not specifically describe outcomes in the subgroup of children who would be identified by UNHS but not by selective screening.</i>
Updated review:	Children identified with hearing impairment through UNHS and provided therapy have better language outcomes at school age than those identified in other ways.	Cohort study	In a fair/good-quality community-based cohort of both high and average risk children with permanent bilateral hearing impairment, those who had early vs. late confirmation and those who had UNHS vs. none had better language scores at age 8 years.
<b>Key Question 3. What are the adverse effects of universal screening and early treatment?</b>			
<i>Initial 2000 review:</i>	<i>Inconclusive</i>	<i>Opinions</i>	<i>Most postulated adverse effects have not been evaluated in studies.</i>
Updated review:	<p>Screening: Limited studies indicate no major adverse psychosocial impact with screening.</p> <p>Treatment: Limited studies indicate few surgical complications in infants receiving cochlear implants, although they have an increased risk for meningitis.</p>	<p>Cohort study, descriptive data, opinions</p> <p>Descriptive data</p>	<p>Screening A fair-quality retrospective cohort study showed no differences on anxiety and attitude toward infant scores of mothers of infants who passed and did not pass screening tests. Survey design studies show mixed results and have low completion rates.</p> <p>Treatment Most adverse effects of treatment have not been evaluated in infants. Surgical series of cochlear implants indicate few complications, although risk for meningitis is increased and may persist for several years after implantation.</p>

## Appendix A. Abbreviations

AAP	American Academy of Pediatrics
ABR	Auditory brainstem response
AHRQ	Agency for Healthcare Research and Quality
ANCOVA	Analysis of covariance
ANOVA	Analysis of variance
ASHA	American Speech-Language-Hearing Association
CDC	Center for Disease Control
CHIP	Children's Health Insurance Program
CI	Confidence interval
CQ	Cognitive quotient
dB	Decibels
DQ	Developmental quotient
ECHI	Early child hearing intervention
EHDI	Early hearing detection and intervention
EPC	Oregon Evidence-based Practice Center
FDA	Food and Drug Administration
HRSA	U.S. Health Resources and Services
Hz	Hertz
IDEA	Individuals with Disabilities Education Act
JCIH	Joint Committee on Infant Hearing
LQ	Language quotient
MCDI	Minnesota Child Development Inventory
NICU	Neonatal intensive care unit
NIDCD	National Institute on Deafness and Other Communication Disorders
NIH	National Institute of Health
NNS	Number needed to screen
OAE	Otoacoustic emissions
PCHL	Permanent congenital hearing loss
RCT	Randomized controlled trial
SE	Standard error
SES	Socioeconomic status
UNHS	Universal newborn hearing screening
USPSTF	U.S. Preventive Services Task Force

## Appendix B1. Search Strategies

### Overall Searches:

#### Database: EBM Reviews - Cochrane Central Register of Controlled Trials

- 1 (universal\$ and (newborn\$ or infant\$) and hearing and screen\$).mp.
- 2 ((deaf\$ or hearing) adj5 (infant\$ or infancy or neonat\$ or newborn\$)).mp.
- 3 1 or 2

#### Database: EBM Reviews - Cochrane Database of Systematic Reviews

- 1 (universal\$ and (newborn\$ or infant\$) and hearing and screen\$).mp.
- 2 ((deaf\$ or hearing) adj5 (infant\$ or infancy or neonat\$ or newborn\$)).mp.
- 3 1 or 2

#### Database: EBM Reviews - Database of Abstracts of Reviews of Effects

- 1 (universal\$ and (newborn\$ or infant\$) and hearing and screen\$).mp.
- 2 ((deaf\$ or hearing) adj5 (infant\$ or infancy or neonat\$ or newborn\$)).mp.
- 3 1 or 2

### Key Question 1 & 2 Specific Search:

#### Database: Ovid MEDLINE

- 1 exp hearing disorders/
- 2 exp hearing impaired persons/
- 3 1 or 2
- 4 infant/ or infant, newborn/
- 5 (universal\$ and (newborn\$ or infant\$) and hearing and screen\$).mp.
- 6 3 and 4
- 7 5 or 6
- 8 limit 7 to humans
- 9 limit 8 to english language
- 10 8 not 9
- 11 limit 10 to abstracts
- 12 9 or 11
- 13 exp Mass Screening/
- 14 screen\$.mp.
- 15 exp Hearing Tests/
- 16 exp Otoacoustic Emissions, Spontaneous/
- 17 (otoacoustic\$ adj2 emission\$).mp. [mp=title, original title, abstract, name of substance word, subject heading word]
- 18 (teoae or dpoae).mp. [mp=title, original title, abstract, name of substance word, subject heading word]
- 19 exp Evoked Potentials, Auditory, Brain Stem/
- 20 (auditory\$ adj2 brainstem\$).mp. [mp=title, original title, abstract, name of substance word, subject heading word]
- 21 13 or 14 or 15 or 16 or 17 or 18 or 19 or 20
- 22 12 and 21
- 23 exp Cochlear Implants/
- 24 exp Hearing Aids/
- 25 exp manual communication/
- 26 exp rehabilitation of hearing impaired/
- 27 exp hearing disorders/dt, rh, su, th
- 28 23 or 24 or 25 or 26 or 27
- 29 12 and 28
- 30 22 or 29
- 31 exp epidemiologic studies/
- 32 meta analysis/
- 33 exp clinical trials/
- 34 (longitudinal\$ or prospective\$ or retrospective\$ or follow up or cross sectional or cohort).mp.

## Appendix B1. Search Strategies

35 exp comparative study/  
36 31 or 32 or 33 or 34 or 35  
37 exp "Outcome and Process Assessment (Health Care)"/  
38 "Sensitivity and Specificity"/  
39 exp diagnostic errors/  
40 exp time factors/  
41 exp age factors/  
42 40 or 41  
43 30 and 36  
44 limit 43 to yr="2000 - 2007"  
45 30 and 37  
46 limit 45 to yr="2000 - 2007"  
47 30 and 38  
48 limit 47 to yr="2000 - 2007"  
49 30 and 39  
50 limit 49 to yr="2000 - 2007"  
51 30 and 42  
52 limit 51 to yr="2000 - 2007"  
53 44 or 46 or 48 or 50 or 52  
54 limit 30 to (controlled clinical trial or guideline or meta analysis or randomized controlled trial)  
55 limit 54 to yr="2000 - 2007"  
56 53 or 55  
57 3 and 21  
58 3 and 28  
59 57 or 58  
60 limit 59 to humans  
61 limit 60 to english language  
62 limit 60 to abstracts  
63 61 or 62  
64 limit 63 to yr="2000 - 2007"  
65 (infant\$ or infancy or newborn\$ or neonat\$).mp. [mp=title, original title, abstract, name of substance word, subject heading word]  
66 64 and 65  
67 36 and 66  
68 38 and 66  
69 37 and 66  
70 67 or 68 or 69  
71 limit 66 to (controlled clinical trial or guideline or meta analysis or randomized controlled trial)  
72 70 or 71  
73 56 or 72

### Key Question 3 Specific Search:

#### Database: Ovid MEDLINE

1 exp hearing disorders/  
2 infant/ or infant, newborn/  
3 (universal\$ and (newborn\$ or infant\$) and hearing and screen\$).mp.  
4 1 and 2  
5 3 or 4  
6 limit 5 to humans  
7 limit 6 to english language  
8 6 not 7  
9 limit 8 to abstracts  
10 7 or 9  
11 (adverse effect\$ or harm\$ or stigma\$).mp. or exp stress, psychological/et or exp life change events/ or exp prejudice/ or exp stereotyping/ or exp self concept/  
12 10 and 11  
13 exp Diagnostic Errors/  
14 10 and 13  
15 12 or 14

## Appendix B2. Inclusion and Exclusion Criteria

### Reasons for Inclusion in Results

- Provides data that address key questions
- Study designs: systematic review, randomized controlled trial, controlled trial without randomization, observational cohort or case-control study, descriptive study that provides unique data (e.g. survey of parental anxiety about screening)
- Hearing impairment identified prior to 6 months of age
- Primary care feasible or referable (see definitions below)
- Applicable years: 2000 to present

### Reasons for Inclusion in Other Sections

- Provides context or background, or addresses methodology, epidemiology, or cost

### Reasons for Exclusion

- Not relevant to key questions
- Study designs: Editorials, letters, non-systematic reviews, non-comparative studies, case studies, chapter, comment/opinion, etc
- Study not conducted in a country generalizable to the US population
- Non-English
- Non-humans
- No data provided

### Criteria for judging if an intervention is primary care *feasible*:

- Whom Targeted: Somehow involves individual-level identification of being a patient in need of an intervention.
- Who Delivered: Usually involves primary care clinicians (physicians in family practice, internal medicine, obstetrics-gynecology, pediatrics, general practice), other physicians, nurses, nurse practitioners, physician assistants, or related clinical staff (dietitians, health educators, other counselors) in some direct or indirect way—or, at least, the intervention would be seen as connected to the health care system by the participant.
- How Delivered: To individuals or small groups (15 or less). Does not involve only or primarily group-level interventions outside the primary care setting to achieve behavioral change. Generally involves no more than a total of 8 group sessions and an intervention time period no longer than 12 months.
- Where Delivered: Could be delivered anywhere (including via the web, interactive technologies, in the home) if linked to primary care as above.

### Definition of primary care *referable*:

In order for an intervention to be feasible for primary care *referral*, it would need to be conducted in a healthcare setting or else be widely available in the community at a national level (such as a car seat fitting station within a hospital).

## Appendix B3. Studies Excluded from Results of Review

### Outside Scope of Report

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### Study Design Limitations

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## Appendix B4. U.S. Preventive Services Task Force Quality Rating Criteria\*

### RANDOMIZED CONTROLLED TRIALS (RCTs) AND COHORT STUDIES

#### *Criteria:*

- Initial assembly of comparable groups: RCTs—adequate randomization, including concealment and whether potential confounders were distributed equally among groups; cohort studies—consideration of potential confounders with either restriction or measurement for adjustment in the analysis; consideration of inception cohorts.
- Maintenance of comparable groups (includes attrition, cross-overs, adherence, and contamination).
- Important differential loss to follow-up or overall high loss to follow-up.
- Measurements: equal, reliable, and valid (includes masking of outcome assessment)
- Clear definition of interventions.
- Important outcomes considered.
- Analysis: adjustment for potential confounders for cohort studies, or intention-to-treat analysis for RCTs.

#### *Definition of ratings based on above criteria:*

- Good:** Meets all criteria: Comparable groups are assembled initially and maintained throughout the study (follow-up at least 80 percent); reliable and valid measurement instruments are used and applied equally to the groups; interventions are spelled out clearly; important outcomes are considered; and appropriate attention to confounders in analysis.
- Fair:** Studies will be graded “fair” if any or all of the following problems occur, without the important limitations noted in the “poor” category below: Generally comparable groups are assembled initially but some question remains whether some (although not major) differences occurred in follow-up; measurement instruments are acceptable (although not the best) and generally applied equally; some but not all important outcomes are considered; and some but not all potential confounders are accounted for.
- Poor:** Studies will be graded “poor” if any of the following major limitations exists: Groups assembled initially are not close to being comparable or maintained throughout the study; unreliable or invalid measurement instruments are used or not applied at all equally among groups (including not masking outcome assessment); and key confounders are given little or no attention.

### CASE CONTROL STUDIES

#### *Criteria:*

- Accurate ascertainment of cases.
- Nonbiased selection of cases/controls with exclusion criteria applied equally to both.
- Response rate.
- Diagnostic testing procedures applied equally to each group.
- Measurement of exposure accurate and applied equally to each group.
- Appropriate attention to potential confounding variable.

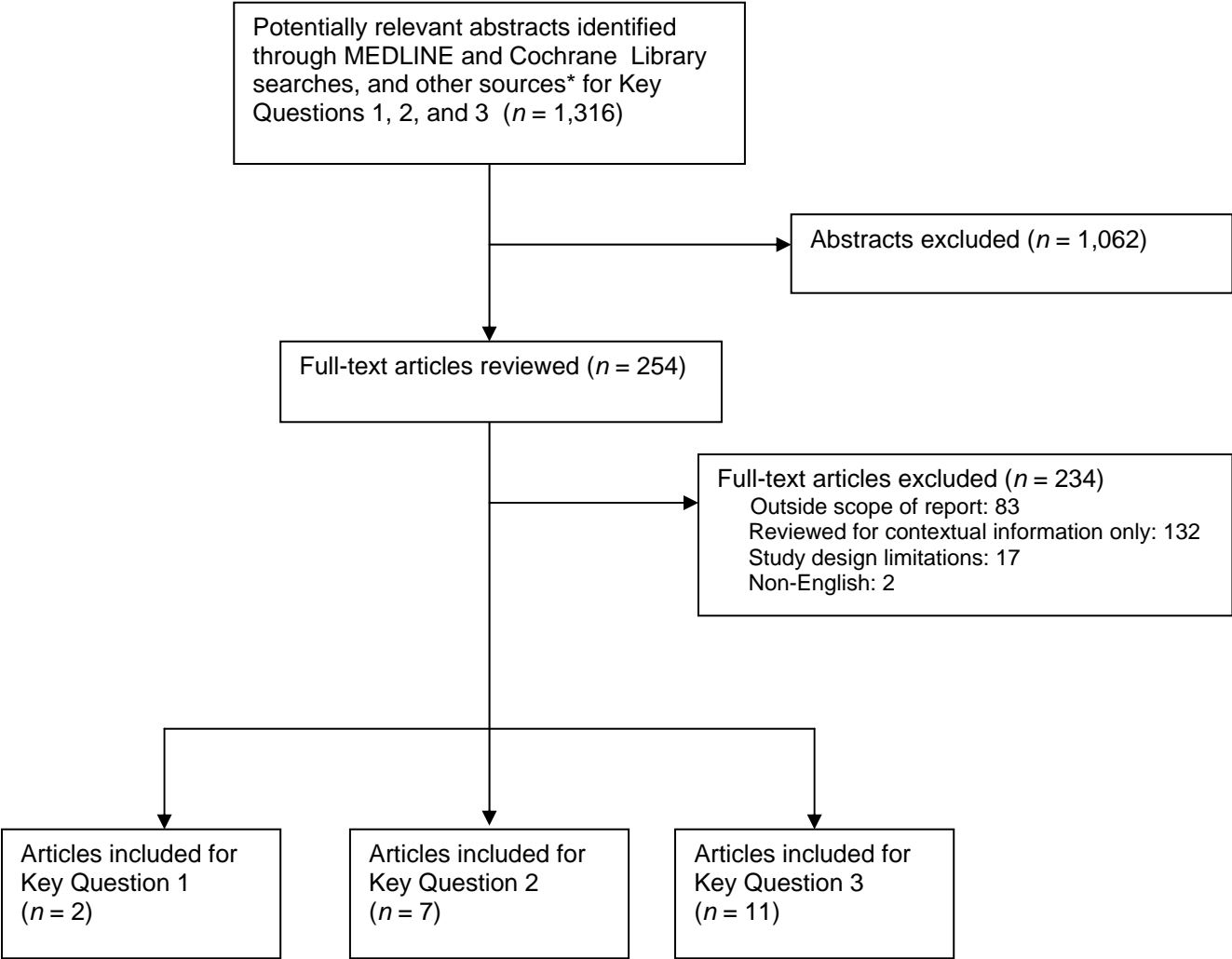
## Appendix B4. U.S. Preventive Services Task Force Quality Rating Criteria\*

### *Definition of ratings based on criteria above:*

- Good:** Appropriate ascertainment of cases and nonbiased selection of case and control participants; exclusion criteria applied equally to cases and controls; response rate equal to or greater than 80 percent; diagnostic procedures and measurements accurate and applied equally to cases and controls; and appropriate attention to confounding variables.
- Fair:** Recent, relevant, without major apparent selection or diagnostic work-up bias but with response rate less than 80 percent or attention to some but not all important confounding variables.
- Poor:** Major selection or diagnostic work-up biases, response rates less than 50 percent, or inattention to confounding variables.

\*Created using information from Harris RP, Helfand M, Woolf SH, et al. Current Methods of the USPSTF: A Review of the Process. *Am J Prev Med* 2001;20(3S):21-35.

**Appendix B5. Yields from Searches, Abstract Review, and Article Review**



\*Identified from reference lists, experts, etc.

## Appendix C. Evidence Table of Key Screening Studies

Study, Year, Location	Study Design and Quality Score	Population	Inclusion & Exclusion Criteria	Referral criteria	Confounders
Kennedy, 2005 <sup>17</sup> UK	Nonrandomized controlled trial, Good	8-year follow-up of all infants born in 4 participating hospitals in southern England from 1993-1996 in the Wessex Trial including 25,609 who had UNHS and 28,172 not screened.	All children enrolled in the Wessex Trial were included and those with abnormal tests at birth or subsequently were followed by audiology services. Information about their diagnoses and management was obtained from multiple sources (records, therapists, etc.). Children with postnatal causes of hearing impairment were excluded.	See Wessex, 1998. All children were also subjected to screening using the Health Visitor Distraction Test at age 7-8 months as usual care in the UK. Some children were referred due to parent or clinician concern.	Severity of hearing impairment.
Kennedy, 2006 <sup>33</sup> UK	Retrospective cohort, Good	120 children with bilateral permanent hearing impairment identified from a large birth cohort in southern England assessed at a mean of 7.9 years of age (range 5.4 to 11.7). Children were either part of the Wessex Trial or from districts in Greater London providing UNHS or not at the time of birth. 63 age-matched children with normal hearing were used to derive z scores for outcome measures.	All children with bilateral permanent childhood hearing impairment of at least 40 dB hearing level identified from a cohort of 157,000 children born in 8 districts of southern England between 1992-1997. Children with known postnatal causes were excluded.	UNHS was performed in some district hospitals. Protocols for identification and confirmation of hearing impairment were similar at all sites (see Wessex, 1998). All children were also subjected to screening using the Health Visitor Distraction Test at age 7-8 months as usual care in the UK. Therapy was provided to all children as a public health service and included audiology services.	Baseline characteristics were similar between comparison groups (gender, English as first language, nonverbal ability using Raven's Progressive Matrices score, age at assessment, degree of hearing loss (moderate [40-69 dB HL], severe [70-94 dB HL], profound [more than 95 dB HL]), other disabilities, mother's education, and occupation of head of household). Outcome measures were adjusted for severity, maternal education, and age-adjusted total Raven's Progressive Matrices scores.

## Appendix C. Evidence Table of Key Screening Studies

Study, Year, Location	Outcomes measured	Results
Kennedy, 2005 <sup>17</sup> UK	Proportion of hearing impaired children referred before age 6 months, proportion referred before age 3 years, age at referral, sensitivity, specificity, positive likelihood ratio, and negative likelihood ratio of screening, magnitude of effect.	<p><b>Proportion referred before age 6 months:</b> 11/35 (31%) with true hearing impairment born during periods without UNHS, 23/31 (74%) born during periods with UNHS (difference 43%, CI 19-60%, P=0.001).</p> <p><b>Adjustment for effect of severity of hearing impairment on age of referral:</b> increased the odds ratio between UNHS and early referral from 6.3 to 6.9 (CI 2.2-22.0, P=0.001).</p> <p><b>Referral before age 3 years:</b> the percentage of all true cases referred was greater at any given age during the first 3 years for children screened vs. not; percentages were similar after age 3.</p> <p><b>Age at referral:</b> lower with UNHS than not (0 mo vs. 8 mo, P&lt;0.001).</p> <p><b>Test sensitivity and specificity:</b> 22/24 (0.92) and 20,960/21,279 (0.98).</p> <p><b>Positive and negative likelihood ratios:</b> 61 and 0.08.</p> <p><b>Magnitude of effect:</b> one additional case of bilateral hearing impairment was referred before age 6 months for every 1969 (CI 1011-12,896) infants in the UNHS population.</p>
Kennedy, 2006 <sup>33</sup> UK	Receptive language: Test for Reception of Grammar, British Picture Vocabulary Scale, aggregate scores. Expressive language: Renfrew Bus Story Test (sentence information and 5 longest sentences), aggregate scores. Speech: Children's Communication Checklist (speech scale).	<p><b>Age at confirmation of hearing impairment:</b> children confirmed early (age 9 months or younger) had better scores than those confirmed later on the Test for Reception of Grammar (adjusted mean difference 0.90, P=0.003), British Picture Vocabulary Scale (0.64, P=0.02), Renfrew Bus Story Test, sentence information (0.54, P=0.03); but not on the Renfrew Bus Story, longest sentences component, or Children's Communication Checklist, speech scale. All aggregate scores for receptive and expressive language were significantly better for the early confirmation group.</p> <p><b>Universal newborn hearing screening:</b> children who underwent UNHS had better scores than those who did not on the Test for Reception of Grammar (0.27, P=0.05), British Picture Vocabulary Scale (0.47, P=0.08), but not the Renfrew Bus Story test or Children's Communication Checklist, speech scale. Aggregate scores for receptive language were better for the UNHS group.</p>

## Appendix C. Evidence Table of Key Screening Studies

Study, Year, Location	Comments
Kennedy, 2005 <sup>17</sup> UK	<p>The numbers of cases offered screening but refused (n=7) will likely decrease as screening becomes an established clinical service.</p> <p>Although referral was early, management was often initiated later (48% after 18 mos), reflecting the evolving management system of the 1990s. 23% of all cases (screened and not) had progressive impairment that could be missed at birth.</p>
Kennedy, 2006 <sup>33</sup> UK	<p>Difference in higher scores for early vs. late confirmation are equivalent to an increase of 10 to 12 points in the verbal compared with nonverbal intelligence quotient.</p> <p>Estimated size of benefit is likely underestimated because the system of screening and follow-up has improved since the first cohort underwent this process.</p> <p>Not clear if children not screened as newborns had onset of hearing impairment after birth or not, a sensitivity analysis indicated that benefit for the screened group would have been higher if all were truly congenital. Speech was assessed on the basis of parental or professional report, rather than by direct measurement, and may lack sensitivity as an outcome measure.</p>

## Appendix C. Evidence Table of Key Screening Studies

Study, Year, Location	Study Design and Quality Score	Population	Inclusion & Exclusion Criteria	Referral criteria	Confounders
Wake, 2005 <sup>35</sup> Australia	Retro-spective cohort, Fair	88 children age 7 to 8 years born in Victoria, Australia who were fitted with hearing aids by age 4.5 years for congenital hearing impairment. Children were identified from a large birth cohort and services were provided by the government for all eligible children.	All hearing impaired children from the birth cohort. Children with intellectual disability and non-English speakers were excluded from the study.	Hearing screening included audiology referral for infants with risk factors and a universally available behavioral hearing screen at age 8 to 10 months.	Outcome measures were adjusted for confounders using multiple regression for non-verbal IQ, maternal education, paternal occupational prestige, and family functioning. Evaluators were blinded to the children's history and hearing status.
Wessex Universal Neonatal Hearing Screening Trial Group, 1998 <sup>18</sup> UK	Nonrandomized controlled trial, Good	All infants born in 4 participating hospitals in southern England from 1993-1996 including 25,609 who had UNHS and 28,172 not screened.	All children born in the participating hospitals. Two teams of testers (trained nursery nurses) and equipment moved between 2 pairs of hospitals to achieve 4 periods of 4-6 months duration with UNHS and 4 without. Infants in special care units and NICUs were screened at the end of their hospital stays.	Newborns with abnormal tests (40 dB hearing level or more using OAE and ABR in those who failed the first test) were referred to local audiological services for assessment and management. These are public health services available to all children. All children were also subjected to screening using the Health Visitor Distraction Test at age 7-8 months as usual care in the UK.	Severity of hearing impairment.

## Appendix C. Evidence Table of Key Screening Studies

Study, Year, Location	Outcomes measured	Results
Wake, 2005 <sup>35</sup> Australia	Receptive and expressive language (Clinical Evaluation of Language Fundamentals), receptive vocabulary (Peabody Picture Vocabulary Test), cognition (Perceptual Organization Index of the Wechsler Intelligence Scale for Children), articulation (Goldman-Fristoe Test of Articulation), reading comprehension (Reading Progress Test 1), intelligibility (teacher questionnaire), and family functioning (McMaster Family Assessment Device).	Age at diagnosis did not contribute significantly to variance on language, speech, or reading measures except receptive vocabulary; severity of impairment contributed to variance on all measures except reading comprehension. Language outcomes were more than 25 points lower than expected from IQ scores and were related to the severity of impairment, but not age at diagnosis. Age at diagnosis was negatively correlated with severity at diagnosis.
Wessex Universal Neonatal Hearing Screening Trial Group, 1998 <sup>18</sup> UK	Proportion screened, proportion with risk factors, test performance, yield of screening vs. usual care, proportion with early referral, odds of early confirmation of impairment, odds of early management.	<p><b>Proportion of newborns screened at birth:</b> 87%.</p> <p><b>Risk factors:</b> proportion screened with known risk factors=8.1%; proportion with impairment who had risk factors=64% (CI 50-77%).</p> <p><b>Test performance:</b> false positive rate=1.5%, false negative rate=4% (distraction test=27%).</p> <p><b>Yield of screening:</b> 90 cases per 100,000 target population confirmed after referral from UNHS (equivalent to 80% of expected prevalence).</p> <p><b>Screening vs. distraction test (usual care):</b> relative risk=2.3 (CI 1.1-4.7) for detection through UNHS vs. distraction test.</p> <p><b>Proportion with early referral:</b> 71 more infants per 100,000 were referred before age 6 months during periods with UNHS vs. during periods without; proportion is equivalent to 19 times higher than non screened infants.</p> <p><b>Early confirmation of hearing impairment:</b> odds of confirmation before age 10 months adjusted for severity=5.0 (1.0-23.0) times greater for screened vs. non screened infants.</p> <p><b>Early management:</b> odds ratio of management before age 10 months=8.0 (1.2-51.0) times higher for screened vs. non screened infants.</p>



## Appendix C. Evidence Table of Key Screening Studies

Study, Year, Location	Comments
Wake, 2005 <sup>35</sup> Australia	Few children were diagnosed younger than age 6 months (n=11) or 12 months (n=28); comparisons between early vs. late diagnosed children were not reported.
Wessex Universal Neonatal Hearing Screening	87% of all births could be covered by screeners in medium to large maternity units with high rates of discharge within 2 days of birth. Higher coverage was attained after the addition of recall clinics for infants discharged from the hospital without screening and increasing personnel on peak services.
Trial Group, 1998 <sup>18</sup> UK	Average non-pass rate was 1.6%, equivalent to 2 infants requiring follow-up assessment every 3 weeks in an annual birth cohort of 5,000. Proportion with risk factors consistent with other studies (50%) supporting need for universal vs. risk factor based screening. Special-care and NICU infants accounted for 36% of hearing impaired infants. Benefit of screening was underestimated due to use of a run-in period when coverage was lower (67%), difficulty obtaining timely parental consent, cases of profound impairment were more prevalent in the non screened group, and short duration of trial follow-up. Families undergoing UNHS were less anxious than those not screened.

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