Newborn Hearing Screening: Recommendations and Rationale U.S. Preventive Services Task Force

United States Preventive Services Task Force

Citation


Abstract

This statement summarizes the current U.S. Preventive Services Task Force (USPSTF) recommendations on newborn hearing screening and the supporting scientific evidence, and it updates the 1995 recommendations contained in the Guide to Clinical Preventive Services, second edition. Explanations of the ratings and of the strength of overall evidence are given in Appendix A and Appendix B, respectively. The complete information on which this statement is based, including evidence tables and references, is available in the article, "Universal Newborn Hearing Screening: A Summary of the Evidence" and in the Systematic Evidence Review and Summary of the Evidence on this topic, which can be obtained through the USPSTF Web site (http://www.ahrq.gov/clinic/uspstfix.htm), through the National Guideline Clearinghouse (http://www.guideline.gov), or in print through the AHRQ Publications Clearinghouse (1-800-358-9295).

SUMMARY OF RECOMMENDATION

The USPSTF concludes the evidence is insufficient to recommend for or against routine screening of newborns for hearing loss during the postpartum hospitalization. I recommendation.

The USPSTF found good evidence that newborn hearing screening leads to earlier identification and treatment of infants with hearing loss. However, evidence to determine whether earlier treatment resulting from screening leads to clinically important improvement in speech and language skills at age 3 years or beyond is inconclusive because of the design limitations in existing studies.

Although earlier identification and intervention may improve
the quality of life for the infant and family during the first year of life, and prevent regret by the family over delayed diagnosis of hearing loss, the USPSTF found few data addressing these benefits. The USPSTF could not determine from existing studies whether these potential benefits outweigh the potential harms of false-positive tests that many low-risk infants would experience following universal screening in both high- and low-risk groups.

The USPSTF found good evidence that the prevalence of hearing loss in infants in the newborn intensive care unit (NICU) and those with other specific risk factors (see “Clinical Considerations”) is 10 to 20 times higher than the prevalence of hearing loss in the general population of newborns. Both the yield of screening and the proportion of true positive results will be substantially higher when screening is targeted at these high-risk infants, but selective screening programs typically do not identify all infants with risk factors. Evidence that early identification and intervention for hearing loss improves speech, language, or auditory outcomes in high-risk populations is also limited.

**CLINICAL CONSIDERATIONS**

Currently, universal newborn hearing screening (UNHS) is required by law in more than 30 states and is performed routinely in some health care systems in other states. Selective screening of infants in the NICU and those with other risk factors for hearing loss (see below) is conducted in many settings that do not follow a policy of universal screening. Clinicians should be aware of such screening policies in their practice environments.

Risk factors for sensorineural hearing loss (SNHL) among newborns include NICU admission for 2 days or more; syndromes known to include hearing loss (eg, Usher's syndrome, Waardenburg's syndrome); family history of childhood SNHL; congenital infections (eg, toxoplasmosis, bacterial meningitis, syphilis, rubella, cytomegalovirus, herpes virus); and craniofacial abnormalities (especially morphologic abnormalities of the pinna and ear canal).

If a program for routine hearing screening of newborns is implemented, it should include systematic education to fully inform parents and clinicians about the potential benefits and harms of the testing protocol. Most infants with positive in-hospital screening tests will subsequently be found to have normal hearing, and clinicians should be prepared to provide reassurance and support to parents of infants who need follow-up audiologic evaluation.

If any program for newborn hearing screening is implemented, screening should be conducted using a validated protocol, usually requiring 2 screening tests. Equipment used should be well maintained, staff should be thoroughly trained, and quality control programs to reduce avoidable false-positive tests should be in place. Programs should develop protocols to ensure that infants with positive screening tests receive appropriate audiologic evaluation and follow-up after discharge.

**SCIENTIFIC EVIDENCE**

**EPIDEMIOLOGY AND CLINICAL CONSEQUENCES**

Each year, an estimated 5,000 infants are born in the United States with moderate, severe, or profound bilateral SNHL. The estimated prevalence of bilateral SNHL is 1-2 per 1,000 newborns in the U.S., but may be 10-20 times higher among infants in the NICU than in the healthy nursery population. Prevalence of bilateral SNHL is also increased in infants with other selected risk factors (see “Clinical Considerations”).

The diagnosis of congenital hearing loss is often delayed. In one survey conducted before hearing screening was common, the median age at diagnosis was 13 months for infants with severe to profound bilateral SNHL and 17 months for those with mild to moderate hearing losses. Children with hearing loss experience delayed development in language, learning, and speech. Impairment exists as early as age 3 years and has consequences throughout life, leading to lower reading abilities, poorer school performance, and under- or unemployment.

**ACCURACY, RELIABILITY, AND SHORT-TERM IMPACT OF SCREENING TESTS**

Between 50% and 75% of infants with moderate to profound bilateral SNHL have one or more specific risk factors (see “Clinical Considerations”). Until recently, most newborn hearing screening programs in the United States focused on identifying and screening infants at risk for SNHL. However, these programs typically do not identify infants at risk for hearing loss due to failure to administer screening questionnaires or loss to follow-up, and they will miss affected infants who have no risk factors.

In the late 1990s, the development of rapid, low-cost screening tests made it feasible to implement screening programs for all newborns for congenital hearing loss during the birth hospitalization. Two types of tests are commonly
used: otoacoustic emissions (OAEs) and auditory brainstem response (ABR). Typically, screening programs use a two-stage screening approach (either OAE repeated twice, OAE followed by ABR, or automated ABR repeated twice). Criteria for defining a “pass” or “fail” on the initial screening test vary, and results are sensitive to equipment, the tester's training, and ongoing quality control.

The true sensitivity and specificity of newborn hearing screening are difficult to estimate from most screening programs. One large, good-quality study measured the sensitivity and specificity of OAE and ABR using an independent “gold standard,” visual reinforcement audiometry, performed at 8 to 12 months. One-stage screening with an ABR or OAE test can detect 80% to 95% of affected ears, depending on how an abnormal test result is defined. The two-stage protocol of OAE and ABR missed 11% of affected ears, but was more specific than testing with the ABR or OAE alone. Because the prevalence of SNHL is low, there are many more false positives than true positives, especially in low-risk populations. Overall, 6.7% of infants who failed in-hospital screening tests were eventually diagnosed with bilateral SNHL in the best study of newborn hearing screening; among those without risk factors for hearing loss, only 2% of those failing such screening tests were later found to have SNHL.

Children who fail in-hospital screening tests are usually referred for repeat testing between 2 and 8 weeks after discharge; positive second-stage results are usually validated by a combination of otolaryngologic and audiologic consultation, diagnostic ABR testing, or other electrophysiologic testing that can be performed as early as age 3 months. Visual reinforcement audiometry cannot be performed reliably before age 8 to 9 months.

Universal newborn hearing screening reduces the age at which infants with hearing loss are diagnosed and treated. Studies of statewide universal newborn hearing screening programs in the United States have found that the mean age of identification of hearing impairment has decreased from 12-13 months before screening programs were introduced to 3-6 months since their introduction. The mean age at which infants receive hearing aids has been reduced from 13-16 months before universal newborn hearing screening programs began to 5-7 months following their introduction. In a large controlled study comparing in-hospital UNHS with no screening, UNHS significantly increased the number of infants with hearing loss referred to audiologists by the age of 6 months and increased the probability that infants with moderate and severe hearing loss would be diagnosed by the age of 10 months (57% vs 14%). Compared with selective screening of high-risk newborns, universal screening would result in the early diagnosis (before 10 months) of one additional case for every 1,441 infants screened, and early treatment (before 10 months) of one additional case for every 2,401 newborns screened, by one estimate.

EFFECTIVENESS OF EARLY INTERVENTION TO IMPROVE LANGUAGE OUTCOMES

There are no prospective, controlled studies that directly examine whether newborn hearing screening and earlier intervention result in improved speech, language, or educational development.

Although several retrospective studies have variously concluded that infants entering treatment programs at younger ages, or infants identified in hospitals with universal screening programs, have better long-term language outcomes, all of these studies have significant methodological flaws.

All of the available retrospective studies began with a convenience sample of children enrolled in early intervention programs, rather than with an inception cohort of children at the point of identification of hearing loss. None described loss to follow-up between enrollment in the intervention program and the age of assessment, and criteria for inclusion and exclusion were not clearly described. In most studies, early identification was not necessarily the result of screening. Therefore, underlying differences between children identified or enrolled early and those identified or enrolled late may have contributed to the observed language differences. Although some studies attempted to adjust for appropriate confounding factors, the USPSTF judged that statistical adjustment cannot compensate for the potential biases arising from unbalanced cohort selection, concluding that the studies do not establish the effectiveness of early identification and treatment.

OTHER POTENTIAL BENEFITS OR HARMS OF SCREENING AND TREATMENT

Because UNHS reduces the average age for intervention by 6 to 9 months, improved hearing or increased prelanguage stimulation over that period might, in themselves, be considered important benefits of newborn hearing screening. In addition, there might be a psychological benefit to parents or to hearing-impaired children of avoiding regret in the
future due to the delayed diagnosis and treatment of hearing impairment. However, the USPSTF was unable to identify any evidence that would allow it to assess the magnitude of these potential benefits or determine whether they alone were sufficient to offset the potential harms of screening.

Because most positive screening tests are false positives, the most likely potential adverse effects of screening are parental anxiety and misunderstanding, and labeling of normal infants as hearing-impaired until the definitive diagnosis can be made months later. Even a small increased risk of these effects could have a large impact on the net benefit of a screening program. In low-risk populations, there are 25 to 50 false positives for each true case of hearing impairment. In existing newborn hearing screening programs, 13% to 31% do not follow up for definitive testing, which might allay concerns about the baby’s health.

Findings from studies that evaluated parental anxiety are mixed. In the largest controlled trial of screening, parents whose infants were screened had similar anxiety and attitudes as parents whose infants were not screened. In another survey, 98% of parents said they would give permission for screening, 95% said they would prefer screening even if the baby failed, and 85% said that anxiety caused by failing a screening test would be outweighed by the potential benefit of early detection. In other studies, false-positive results produced significant or lasting anxiety in 3% to 14% of parents, even after follow-up testing. No studies have evaluated whether parental anxiety has any long-term effect on parent-child interaction.

Because definitive diagnoses may take months to confirm, false-positive diagnosis of SNHL may occasionally lead to unnecessary intervention in an infant who hears normally. In one large screening trial, the initial audiologic diagnosis was incorrect in 2 of 27 infants diagnosed with SNHL (7%), and the infants proved to have normal hearing when re-examined at age 4 months or 10 months. The yield of newborn hearing screening is comparable to or better than those for newborn screening programs for other disorders, including hemoglobinopathy and phenylketonuria. Thus, if the effects of screening and subsequent treatment on longer-term language outcomes could be confirmed, the cost-effectiveness of newborn hearing screening might be equal or superior to that of many other newborn screening services.

RECOMMENDATIONS OF OTHERS

The Joint Committee on Infant Hearing 2000 Position Statement, developed and approved by the American Academy of Audiology, the American Academy of Pediatrics (AAP), the American Speech-Language-Hearing Association (ASHA), the Council on Education of the Deaf, and Directors of Speech and Hearing Programs in State Health and Welfare Agencies, endorses early detection of and intervention for infants with hearing loss (early hearing detection and intervention, [EHDI]) through integrated, interdisciplinary state and national systems of UNHS, evaluation, and family-centered intervention. Audiologic evaluation and medical evaluations should be in progress before 6 months of age. Infants with confirmed hearing loss should receive intervention before 6 months of age from health care and education professionals with expertise in hearing loss and deafness in infants and young children.

The Centers for Disease Control and Prevention supports universal newborn hearing screening through its Early Hearing Detection and Intervention (EHDI) Program, which assists states in implementing screening and intervention programs and supports research and data collection on EHDI programs. The 1993 National Institutes of Health Consensus Development Panel also recommended universal screening for hearing impairment prior to 3 months of age in order to identify and initiate treatment for all hearing-impaired infants by 6 months of age. A publication promoting the early identification of hearing loss has been published by the Maternal and Child Health Bureau of the Health Resources and Services Administration (HRSA). HRSA supports universal screening and has provided funding to assist states in developing such programs.

The American Academy of Family Physicians (AAFP) and the Canadian Task Force on Preventive Health Care are currently reviewing their positions on universal newborn hearing screening.

The American College of Obstetricians and Gynecologists recommends screening for hearing loss in neonates with any of the following risk factors: family history of hereditary childhood SNHL, in utero infection, craniofacial anomalies, or maternal diabetes.
birth weight less than 1,500 grams, hyperbilirubinemia requiring exchange transfusion, ototoxic medications, bacterial meningitis, Apgar score of 0-4 at 1 minute or 0-6 at 5 minutes after birth, mechanical ventilation lasting 5 days or longer, or stigmata or other findings associated with a syndrome known to include a sensorineural or conductive hearing loss.\textsuperscript{19}

The British National Coordinating Centre for Health Technology Assessment supports universal neonatal hearing screening, supplemented by a targeted infant distraction test at about 7 months of age, primarily for those children not screened neonatally.\textsuperscript{20}

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**APPENDIX A**

**U.S. PREVENTIVE SERVICES TASK FORCE RECOMMENDATIONS AND RATINGS**

The Task Force grades its recommendations according to one of 5 classifications (A, B, C, D, I) reflecting the strength of evidence and magnitude of net benefit (benefits minus harms):

A. The USPSTF strongly recommends that clinicians routinely provide [the service] to eligible patients. The USPSTF found good evidence that [the service] improves important health outcomes and concludes that benefits substantially outweigh harms.

B. The USPSTF recommends that clinicians routinely provide [the service] to eligible patients. The USPSTF found at least fair evidence that [the service] improves important health outcomes and concludes that benefits outweigh harms.

C. The USPSTF makes no recommendation for or against routine provision of [the service]. The USPSTF found at least fair evidence that [the service] can improve health outcomes but concludes that the balance of benefits and harms is too close to justify a general recommendation.

D. The USPSTF recommends against routinely providing [the service] to asymptomatic patients. The USPSTF found
at least fair evidence that [the service] is ineffective or that harms outweigh benefits.

I. The USPSTF concludes that the evidence is insufficient to recommend for or against routinely providing [the service]. Evidence that [the service] is effective is lacking, of poor quality, or conflicting and the balance of benefits and harms cannot be determined.

APPENDIX B

U.S. PREVENTIVE SERVICES TASK FORCE

STRENGTH OF OVERALL EVIDENCE

The USPSTF grades the quality of the overall evidence for a service on a 3-point scale (good, fair, poor):

Good: Evidence includes consistent results from well-designed, well-conducted studies in representative populations that directly assess effects on health outcomes.

Fair: Evidence is sufficient to determine effects on health outcomes, but the strength of the evidence is limited by the number, quality, or consistency of the individual studies, generalizability to routine practice, or indirect nature of the evidence on health outcomes.

Poor: Evidence is insufficient to assess the effects on health outcomes because of limited number or power of studies, important flaws in their design or conduct, gaps in the chain of evidence, or lack of information on important health outcomes.

References

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