Universal Screening for Infant Hearing Impairment: Not Simple, Not Risk-Free, Not Necessarily Beneficial, and Not Presently Justified

Fred H. Bess and Jack L. Paradise

Pediatrics 1994;93;330-334

The online version of this article, along with updated information and services, is located on the World Wide Web at:

http://www.pediatrics.org
Universal Screening for Infant Hearing Impairment: Not Simple, Not Risk-Free, Not Necessarily Beneficial, and Not Presently Justified

Screening for hearing impairment of all infants within the first 3 months of life, and preferably before discharge from the hospital newborn nursery, recently has been recommended in a National Institutes of Health Consensus Statement.1 We agree with the desirability of identifying infants with congenital hearing impairment promptly, but we believe that the recommendation of universal screening is ill-considered and at this time ill-advised, and that its implementation might result in more harm than good. We also consider the Consensus Statement’s discussions of the implications of acquired hearing loss during infancy and early childhood to be insufficiently balanced and qualified, and potentially encouraging of overly aggressive management.

The National Institutes of Health Consensus Statement was prepared by an independent 15-member panel representing audiology, otolaryngology, pediatrics, speech and hearing sciences, epidemiology, health care administration, various other child care areas, and the general public. The panel considered presentations and discussions at a 1½ day Consensus Development Conference on the Early Identification of Hearing Impairment in Infants and Young Children convened jointly by the National Institute on Deafness and Other Communication Disorders and the NIH Office of Medical Applications of Research, and cosponsored by the National Institute of Child Health and Human Development and the National Institute of Neurological Disorders and Stroke. The Consensus Statement has not, to our knowledge, been reprinted in any pediatric or general medical journal, but its recommendation of universal screening for hearing impairment by 3 months of age has been reported in pediatric news publications,2,3 and free single copies of the Statement are available on request from NIH (1-800-644-6627).

Four main concerns seemed to underly the panel’s recommendation of early screening. First, the prevalence of hearing impairment in infants and young children: “approximately 1 of every 1000...born deaf, many more...born with less severe degrees of hearing impairment...[still] others develop[ing] hearing impairment during childhood.” Second, the concern that “reduced hearing acuity [during the first three years of life] interferes with the development of speech and verbal language skills,” and that “significantly reduced auditory input...adversely affects the developing auditory nervous system and can have harmful effects on social, emotional, cognitive, and academic development, as well as on...vocational and economic potential.” Third, the concern that, despite persistent efforts to achieve early identification of children with hearing impairment, “the average age of identification in the United States remains close to three years [while] lesser degrees of hearing loss may go undetected even longer, [with] the result...that...for many hearing-impaired infants and young children, much of the crucial period for language and speech learning is lost.” And fourth, the concern that the current practice of limiting hearing screening to infants who meet criteria for inclusion in a high-risk register4 “misses 50 percent of children who are eventually diagnosed with severe to profound hearing impairment.”

The consensus panel recommended a two-stage screening protocol. All infants would be screened with a test that measures evoked otoacoustic emissions (EOAEs), acoustic responses produced in the inner ear by physiologic activity of the outer hair cells of the cochlea, and measured with a sensitive microphone placed in the ear canal.5 Infants who failed the EOAE test would receive an auditory brainstem response (ABR) screen at 40 dB nHL.* Failure at this second stage would call for a comprehensive diagnostic hearing assessment within 6 months.

OBJECTIONS TO THE CONSENSUS STATEMENT

What are our objections to the Consensus Statement? Mainly, we are concerned that the consensus panel’s recommendation of universal infant screening falls short of being justified on grounds of practicability, effectiveness, cost, and harm-benefit ratio. In addition, we are concerned that the Consensus Statement’s dire and sweeping admonitions concerning the long-term developmental effects of early-life “hearing impairment” do not discriminate sufficiently between the acknowledged adverse effects of moderate to severe, persistent hearing loss, and the entirely speculative and perhaps nonexistent effects of mild or moderate temporary hearing loss. The reader is thus encouraged implicitly by the Consensus Statement to take an activist approach in screening for, and in managing, the conductive hearing losses that occur commonly throughout infancy and early childhood in association with episodes of otitis media—an approach that may involve various medical and surgical interventions, yet that remains largely unsubstantiated. Because the latter issue has been discussed in detail previously,6,7 we confine our remaining comments to a consideration of the Consensus Statement’s main recommendation of universal screening of young infants for hearing impairment.

WHEN IS SCREENING APPROPRIATE?

For a disorder to warrant being screened for, several criteria must be met concerning, respectively, the

* nHL (normal Hearing Level)—click stimulus is referred to in terms of dB above the behavioral threshold of a group of normal listeners.
disorder, the screening procedure, and the circumstances of eventual treatment. The disorder must be important, i.e., it must cause substantial mortality, morbidity, or suffering. The screening procedure must be safe, acceptable, simple, reliable, valid, reasonably low in cost, and practicable. Finally, when the disorder is found to be present, the treatment that follows must be efficacious, available, accessible, and readily complied with, and, crucially, early treatment must be more effective than later treatment.

**IMPOR TANCE OF EARLY SENSORINEURAL HEARING LOSS**

No dispute exists as to whether early childhood hearing loss can impose burdens on the affected child, on the child’s family, and on society. Early onset of persistent congenital or acquired hearing loss in the moderate to profound range (41 to 100 dB) affects speech perception adversely. This, in turn, may result in impairment of both receptive and expressive speech and language development, reduction in academic achievement, and disturbances in social and emotional development. Further, difficulties in communication and frequently associated poor academic performance may lead to lowered self-esteem and to social isolation. Even children with unilateral sensorineural hearing loss or mild (26 to 40 dB) bilateral sensorineural hearing loss may experience difficulties in speech or language development, speech recognition under adverse listening conditions (e.g., classroom noise), educational achievement, and psychosocial behavior.

Hearing impairment also imposes a significant economic burden; relatively few deaf persons are employed in professional, technical, and managerial positions. The lifetime economic cost of congenital deafness is estimated currently to exceed 1 million dollars. Other, less tangible costs borne by affected individuals and by their families derive from emotional stress, breakdowns in family communication, and isolation from peers and educational systems.

**SCREENING TESTS FOR HEARING LOSS IN YOUNG INFANTS**

Because the screening procedure recommended in the Consensus Statement consists of two stages, the EOAE test first, followed by the ABR for those who fail the EOAE, we will consider the attributes of both tests as screening instruments. Because the ABR is the more familiar of the two tests and is widely accepted for use in both screening and diagnosis, it may serve to some extent as a standard against which to compare the EOAE.

**Safety**

Safety is not an issue with either test.

**Acceptability and Simplicity**

Acceptability to patients is not an issue with either test because the patients are newborns or young infants and neither test is invasive or traumatic. However, acceptability to the clinicians performing the tests, an often overlooked issue, is another matter. Using the diagnostic ABR equipment available in hospitals often is difficult and time-consuming, and test results are often difficult to interpret, even by professionals. In contrast, using EOAE equipment and automated ABR equipment appears to be easier and less time-consuming, and probably can be accomplished satisfactorily by trained nonprofessionals.

**Reliability**

The reliability of a test refers to the extent to which the test gives consistent results on repeated trials, by either the same or different observers, in the same individual. One might expect the reliability of both the ABR and the EOAE as screening tests to be high, but the issue warrants additional study.

**Validity and Predictive Value**

Validity consists of two components: sensitivity, the proportion of individuals with a disorder who have a positive test for the disorder, and specificity, the proportion of individuals without the disorder who have a negative test. Additional test characteristics of key importance in screening for a disorder are positive predictive value, the probability of an individual having the disorder when the test is positive, and negative predictive value, the probability of not having the disorder when the test is negative. The sensitivity and specificity of a test for a disorder tend to remain relatively stable, but the predictive value of the test for the disorder is highly dependent on the prevalence of the disorder in the population being tested. For a disorder of very low prevalence, even a highly specific test is likely to have a low positive predictive value. For example, a test that has a sensitivity of 90% and a specificity of 90% in detecting a disorder will have a positive predictive value of 79.4%, and thus a false-positive rate of 20.6%, if the prevalence of the disorder is 30%. However, the positive predictive value decreases to 50% if the prevalence is only 10%, to 8.3% if the prevalence is only 1%, and to 0.9%, with a false-positive rate of 99.1%, if the prevalence is only 0.1%.

In general, the ABR is highly sensitive in screening for moderate to severe hearing impairment in newborns and young infants. However, because the ABR uses a click stimulus it is primarily able to detect high-frequency hearing loss; the rare infant with low-frequency loss may be missed. The specificity and positive predictive value of the ABR, when used as a screening test in well-baby nurseries, appear problematic. Thus, in keeping with the values we have cited above, the Consensus Statement notes the problem of over-referral as a consequence of ABR screening: “In the well-baby nursery...for every child with significant hearing impairment, more than 100 babies are referred.” The same concerns necessarily apply to a recently devised, automated ABR instrument that is simpler to use and less expensive than conventional ABR equipment.
most studies of EOAE testing in newborns have been conducted under “laboratory” conditions, ie, by skilled professionals in sound-treated rooms. Use of the test by nonprofessionals in newborn nurseries, as would be required in the interest of practicability for mass screening, has not been evaluated but obviously risks being more problematic. Finally, additional problems reside in the facts that protocols using commercially available instrumentation for measuring EOAEs are based on normative data derived from adults rather than infants, that no consensus exists regarding pass/fail criteria, and that follow-up behavioral data on newborns who have passed the EOAE screen remain scant.

As for the two-stage, EOAE-ABR screening sequence recommended in the Consensus Statement, no data on actual use are available, to our knowledge. However, it is possible to gain some notion of the risks of the results to be anticipated, at best, from full implementation of this two-stage screen by using, in part, estimates cited in the Consensus Statement and by generously assuming that the EOAE and the ABR tests each have a sensitivity of 100%. Under these circumstances, of the approximately 4 million infants born in the United States each year and receiving the EOAE screen, 403 600 would fail it. Of these, 399 600 would be false positives and 4000 would be true positives. All 403 600 would then receive the ABR screen, of whom 359 640 would pass and 43 960 would fail. The 359 640 who passed would be discharged, but flagged for rescreening at age 3 to 6 months. The 43 960 who failed would be referred for comprehensive evaluation to differentiate the 4000 true positives from the 39 960 false positives.

Cost
Calculation of the cost of the proposed screening program must take into account not only the direct cost of the screening procedure itself but also the cost of the assessment, monitoring, and intervention that would be undertaken as a consequence of the screening. Especially important are the costs deriving from false-positive tests: both the monetary costs of parents’ lost time from work, transportation to care facilities, otherwise unnecessary tests, and unnecessary treatments; and the human and probably more consequential costs of attendant parental anxiety, distraction, and potential misunderstanding, of disturbance of family function, and of unnecessary or harmful procedures or treatments carried out on children. We estimate that the direct monetary costs alone of the proposed program, assuming ideal conditions, would approximate $200 000 000 annually.

Practicability
Currently, newborns in the United States are often discharged from hospital nurseries within 24 hours and usually within 48 hours. As noted previously, it is within that period that the specificity of EOAE testing is at its lowest level. Therefore, to undertake EOAE testing on newborns before hospital discharge is to invite even larger numbers than otherwise of false-positive tests, or to delay hospital discharge, or both, as well as to add to the burden of change, heightened activity, and emotion often borne by parents at that time. Further, an estimated 25% of births in this country occur in rural or remote areas, many of which lack qualified audiological professionals and sophisticated audiometric equipment; the second-stage screening and follow-up evaluation of infants from such areas would pose formidable problems of logistics and cost.

TREATMENT OF HEARING LOSS IN YOUNG INFANTS

Efficacy
Once hearing impairment has been accurately diagnosed, is treatment efficacious? Much theoretical understanding, intuitive belief, and clinical experience argue in favor of efficacy. Nonetheless, no direct evidence demonstrates conclusively that intervention appropriate by current standards results in more good than harm to the child and the family. Several case-control and cohort studies have suggested that intervention improves children’s use of residual hearing and their speech-language skills, social and emotional status, and academic performance, but other studies have failed to show such benefits.

Critical review of all of the studies reveals important limitations in design or methodology that render interpretation of their findings difficult. These limitations include inadequate description of the population studied; selection bias; weighting of the study population with children who have severe or profound, as opposed to mild or moderate, sensorineural hearing loss; small sample size; questionable matching of cases and controls; failure to adjust for potentially confounding variables; failure to document the degree of compliance with recommended treatment; questionable validity and preciseness of outcome measures; failure to use blind examiners; and failure to differentiate maturation effects from treatment effects.

Availability and Accessibility
It is improper to screen for a disorder without certainty that facilities for suitable follow-up care of individuals who fail the screen are both available and accessible. Previously we expressed concern about the availability of qualified professionals in rural or remote areas to undertake the volume of follow-up diagnostic testing that a universal screening program would generate; the concern applies equally to the availability of such professionals for treatment. Moreover, families in such areas often lack health insurance or are underinsured, so that even if suitable professional services were available, they might not be affordable for such families until such time as a national health insurance program had come into being.

Compliance
The issue of compliance was not addressed in the Consensus Statement, but noncompliance has been one of the principal limitations of existing hearing screening programs for newborns. In some studies, proportions ranging from 25% to 80% of infants who failed newborn screening have been lost to follow-up despite aggressive recruiting efforts and the offering...
of cost-saving incentives to parents. In other studies, after early identification of hearing loss, lag times of 8 to 9 months have transpired before infants returned for interventive services. To our knowledge, no data have been reported on compliance in hearing screening programs for infants beyond the newborn period. It seems reasonable to anticipate that noncompliance would constitute a problem of substantial magnitude in any universal screening program, and that substantial effort and resources would be required to minimize its effects.

Early versus Later Treatment

Screening for hearing impairment can be justified only if treatment initiated before the impairment has become apparent is more effective than treatment initiated afterward. Although supported by theory and belief, no empirical evidence, to our knowledge, supports the proposition that outcomes in children with congenital hearing loss are more favorable if treatment is begun early in infancy rather than later in childhood (eg, 6 months vs 18 months).

CONCLUSIONS

In our judgment, before any societal decision is made as to whether to institute a universal screening program for hearing impairment in young infants, many questions for which answers are not now available must be answered. To answer those questions will require extensive research. Clearly, the authors of the Consensus Statement were not unmindful of the scope of the problem, for they included in the Statement a listing of proposed future studies they considered important. Among these were: "controlled trials of screening by audiologists versus trained nonprofessionals or volunteers; controlled trials of the influence of different settings...on the effectiveness of screening procedures; comparison of early intervention with later intervention...evaluate the validity and reliability of screening instruments...test the feasibility of screening methods...in infant populations...in remote satellite clinics..." determine whether a two-tier screening system...works better than [a single screen]...study the cost-effectiveness of universal screening for infant hearing impairment." We heartily agree that these studies, and other studies, are important—indeed, crucial. So crucial, in fact, that, until they have been conducted and their results tabulated, no rational decision on undertaking universal screening for infant hearing loss is possible. We, too, believe that early identification is important; however, the precipitate launching of mass screening could work to deter the eventual development of an effective early identification program. In the meantime, to identify infants at risk for hearing impairment, continued reliance on the high-risk register as recommended by the Joint Committee on Infant Hearing, but in combination with an automated rather than conventional ABR screen, seems to be a more practical, cost-effective approach.

Fred H. Bess, PhD
Division of Hearing and Speech Sciences
Vanderbilt University School of Medicine

Bill Wilkerson Center
Nashville, TN

Jack L. Paradise, MD
Dept of Pediatrics
University of Pittsburgh School of Medicine
Children's Hospital of Pittsburgh
Pittsburgh, PA

REFERENCES

42. Weisel A. Early intervention programs for hearing impaired children—evaluation of outcomes. Early Child Dev Care. 1990;41:77-87

ETHICS COMMITTEE ROAST

In a corner the ethics committee was still debating. It had been in session for three weeks after its latest reconstitution. Everybody had at least agreed that proper representation from the community should include a vegetarian priest, a fundamentalist muezzin, a presocratic engineer, a medicine man from Gambia, two officials from the shepherds' union, and a third year Hispanic schoolgirl. . .

Quietly the nurse tiptoed into the room to borrow a screwdriver from the engineer to disconnect the cardiogrump. "It's OK," she said, "the machine has given its informed consent." Everybody looked relieved.


Submitted by Student
Necessarily Beneficial, and Not Presently Justified

Universal Screening for Infant Hearing Impairment: Not Simple, Not Risk-Free, Not
Fred H. Bess and Jack L. Paradise
Pediatrics 1994;93;330-334

Updated Information
including high-resolution figures, can be found at:
http://www.pediatrics.org

Citations
This article has been cited by 22 HighWire-hosted articles:
http://www.pediatrics.org#otherarticles

Permissions & Licensing
Information about reproducing this article in parts (figures, tables) or in
its entirety can be found online at:
http://www.pediatrics.org/misc/Permissions.shtml

Reprints
Information about ordering reprints can be found online:
http://www.pediatrics.org/misc/reprints.shtml