Universal Newborn Hearing Screening: a Promise Moving Toward Achievement

Jean Johnson, Dr.P.H.
Center on Disability Studies
University of Hawai`i
June Holstrum, Ph.D.
Centers for Disease Control
Yusnita Weirather, M.A.
Newborn Hearing Screening Program
Hawai`i Department of Health
Martha Guinan, M.P.H.
Center on Disability Studies
University of Hawai`i
Roma Johnson, M.A.
MCH LEND Program
University of Hawai`i

Introduction

The promise of universal newborn hearing screening received a needed boost in 1989 when the U.S. Surgeon General C. Everett Koop established as a Year 2000 Health Goal that all children with significant hearing impairment be identified before 12 months of age (Koop, 1993; U.S. Department of Health and Human Services, 1990). While the goal was not achieved by the Year 2000 for all children, significant progress has been made. The purpose of this article is to describe the importance of this goal, the current status of newborn hearing screening in the United States, and events that may make this goal a reality over the next decade.

Importance

The devastating effect of a severe-to-profound hearing loss on a child has long been recognized. Lack of either hearing or recognition by the family that hearing is absent results in delayed acquisition of communication skills that often has a life-long impact on academic achievement, social competence, and vocational opportunities (Mauk & Behrens, 1993; Gallaudet University Center for Assessment and Demographic Study, 1998). While this impact is mitigated if the child is born into a family of deaf parents, more than 87% of all deaf children are born to hearing parents (Holt, Hotto, and Cole, 1994). Lack of ability to communicate during infancy creates a great emotional and functional impact on the family. Often the infant's inability to respond and failure to develop language causes families to suspect the presence of other disabling conditions. Thus, undiagnosed severe-to-profound hearing loss produces uncertainties, stress, and emotional duress during the important early months of parenting.

The negative impact of unilateral and mild bilateral sensorineural hearing loss on the child's education and communication ability has also been documented (Bess, 1985; Blair, et al.,1985; Mauk & Behrens, 1993). Emerging research on the critical importance of auditory competence during the first three years of life underscores the importance of

identifying any hearing loss as early in life as possible to assure that acquisition of communication skills is not delayed (Ruben, 1992; Kuhl, et al., 1992).

Many congenital disabling conditions are obvious to physicians at the time of birth. Not so with hearing loss. Prior to 1993 the average age of identification of a child with a severe-to-profound hearing loss was approximately 2.5 years with significant mild-moderate hearing loss not identified until 5 to 6 years of age. A 1987 study in Hawai`i found that the average age of identification for severe-to-profound hearing loss ranged from 2.8 months to 4.4 years depending on where the child lived and the family's insurance coverage (Johnson, et al., 1997).

Technological Advances

Early identification was hampered not only by physicians' difficulty in recognizing the possibility of a hearing loss, but by the lack of technological capability by audiologists to diagnose a hearing loss using objective physiological measures. Universal screening was attempted using instruments such as the crib-o-gram and risk registers (Downs, 1995), but with poor results. After Kemp (1978) first described oto-acoustic emissions (OAE), the potential for using this method as a non-invasive, objective measurement of cochlear integrity in the newborn became a topic for discussion (Bray & Kemp, 1987; Kemp, 1978).

The possibility of universal newborn hearing screening came a step closer to reality with the development of the OAE and the automated auditory brainstem response (AABR) technology in the 1980s. It now became feasible to use an objective, non-invasive instrument that could be administered by a hospital staff, paraprofessionals, or volunteers to screen all infants prior to discharge from the newborn nursery. A series of research studies have demonstrated the validity, reliability, and effectiveness of universal newborn hearing screening (Finitzo, et al., 1998; Prieve, Stevens, 2000; Spivak, 1998; Vohr, Maxon, 1996).

National Activities

When these developments in technology showed that universal newborn hearing screening was now a possibility, political and policy activities accelerated to make it a reality. Two states led the way: Rhode Island and Hawai`i. The U.S. Maternal and Child Health Bureau provided the first federal support to encourage statewide programs. The Rhode Island Hearing Assessment Project was begun at Women and Infants Hospital in Rhode Island in 1989 and expanded in 1991 to include a pilot site in Hawai`i.

The first legislation to mandate the hearing screening of all newborns was passed in Hawai`i and signed into law in May 1990 (Johnson, et al., 1997). Concurrent with these developments, the National Institutes on Health (NIH) issued a Consensus Statement on Early Identification of Hearing Impairment in Infants and Young Children in 1993 (National Institute on Deafness and Other Communication Disorders, 1993). The statement concluded that all infants admitted to the neonatal intensive care units (NICU) should be screened for hearing loss before hospital discharge and that universal screening should be implemented for all infants within the first three months of life.

Following its Consensus Statement, the NIH sponsored a multi-center study to determine the accuracy of three measures of peripheral auditory system status (transient evoked otoacoustic emissions, distortion product otoacoustic emissions, and auditory brain stem responses) applied in the perinatal period for predicting behavioral hearing status at 8 to 12 months corrected age (Norton, et al., 2000). Seven institutions, with an

annual census of 7,179 births, participated in the study. The evaluation of newborn hearing screening programs required a longitudinal study in which newborn screening results were compared with a "gold standard" based on behavioral audiometric assessment when the newborn was mature enough to give reliable, objective responses to a hearing test.

The results of that study were published in late 2000 (Harrison, et al., 2000; Cone-Wesson, et al., 2000; Norton, et al., 2000; Vohr, et al., 2000; Sininger, et al., 2000; Gorga, et al., 2000; Norton, Gorga, et al., 2000; Keefe, et al., 2000; Folsom, et al., 2000; and Widen, et al., 2000). The study showed conclusively that all three screening tests resulted in low rates of referral and all were successful in identifying hearing loss in newborns (Norton, Gorga, et al., 2000). Researchers found that although perfect test performance was never achieved, sensitivity for each measure increased with the magnitude of hearing loss. This important finding suggests that all three tests can successfully identify newborns with hearing loss for which intervention could be immediately recommended.

Another important finding from the study was that only a small percentage of infants with a conventional risk indicator for hearing loss actually had a hearing loss, but a much larger number of infants with hearing loss did not have a risk indicator (Cone-Wesson, 2000). These findings support the importance of universal neonatal hearing screening using reliable, objective measures rather than targeting for follow-up only those newborns with risk indicators.

Another important outcome of the study was establishment of the "gold standard" using a visual reinforcement audiometry protocol. This large-scale study demonstrated that the hearing of more than 95% of infants can be accurately evaluated prior to one year of age using a standardized behavioral protocol (Widen, et al., 2000).

During the mid-1990s the Maternal and Child Health Bureau continued to provide some limited federal funding to provide technical support to states and hospitals seeking to implement universal newborn hearing screening. Between 1994-1997 a grant was awarded to the National Center for Hearing Assessment & Management at Utah State University and from 1997-2000 a similar grant was awarded to the Marion Downs National Center for Infant Hearing at the University of Denver. Over those six years these two institutions provided technical assistance to hospitals and states to support the development of universal newborn hearing screening programs. The Centers for Disease Control and the U.S. Department of Education's Office of Special Education Programs also began providing limited funding, primarily in support of the development of data and surveillance systems.

National efforts received a significant boost in 1999 when federal legislation was passed to provide additional federal support and funding. Representative James Walsh of New York had become an advocate for the early identification of hearing loss introducing the Walsh Bill to encourage states to implement universal newborn hearing screening. The legislation provided $3,500,000 in funding for the Maternal and Child Health Bureau in Health Resources and Services Administration to provide grants to states to develop statewide universal newborn hearing screening, evaluation, and intervention programs. As a result of the legislation, the Maternal and Child Health Bureau funded grants for 22 states to enhance development of statewide systems and a grant to the National Center for

Hearing Assessment and Management to provide technical assistance and support for states and hospitals.

The federal legislation awarded another $3,500,000 to the Centers for Disease Control to fund data systems and research activities. The Centers for Disease Control funded grants to 15 states to develop tracking and surveillance systems to support screening, identification, intervention, and follow-up activities. In the federal budget for the 2001 FY a significant increase in funding was given to both agencies to further expand these activities.

The Centers for Disease Control identified four research priorities. These priorities included issues related to the effectiveness and cost of early detection of hearing impairment programs, causes and associated factors for hearing loss, benefits of early identification and intervention for children with hearing loss, and psychological and family issues.

Currently underway is a large multi-site national study to validate the two-step screening procedure that first uses OAE, with follow-up ABR for those infants who do not meet the response criteria under OAE. The current protocol requires diagnostic follow-up only for newborns who do not pass either of the screening measures. Concerns arose that some newborns, especially those with a mild-to-moderate loss, might fail the OAE screening, but pass the ABR. A longitudinal national study is currently in the implementation phase to follow at least 1,000 newborns who fail the OAE, but pass the ABR to determine by nine months of age whether any infants with a significant hearing loss are being routinely missed by this procedure. The University of Hawai`i is leading this study in collaboration with the National Center for Hearing Assessment and Management.

Year 2000 Position Statement

All related professional organizations officially endorsed universal newborn hearing screening in mid-2000 with the publication of the Year 2000 Position Statement: Principles and Guidelines for Early Hearing Detection and Intervention Programs (Joint Committee on Infant Hearing, 2000). Membership on the Joint Committee on Infant Hearing includes representatives from the American Academy of Audiology, the American Academy of Otolaryngology, the American Academy of Pediatrics, the American Speech-Language Hearing Association, the Council on Education of the Deaf, the Conference of Educational Administrators of Schools and Programs for the Deaf, the Convention of American Instructors of the Deaf, the National Association of the Deaf, and the Association of College Educators of the Deaf and Hard of Hearing. These organizations include the relevant organizations serving deaf children and adults. The Committee spoke with a single voice to endorse early detection and intervention for infants with hearing loss. Thus, newborn hearing screening now represents the standard of care in newborn nurseries.

That statement acknowledges the importance of not only the screening-identification process, but the critical importance of early intervention activities with the goal of maximizing linguistic and communicative competence and literacy development for all children, especially those who are deaf and hard of hearing. The position statement incorporates the following eight principles (Joint Committee on Infant Hearing, 2000):

1. All infants have access to hearing screening using a physiologic measure. Newborns who receive routine care have access to hearing screening during their hospital birth admission. Newborns in alternative birthing facilities, including home births, have access to and are referred for screening before one month of age. All newborns or infants who require neonatal intensive care receive hearing screening prior to discharge from the hospital.

2. All infants who do not pass the birth admission screen and any subsequent rescreening begin appropriate audiologic and medical evaluations to confirm the presence of hearing loss before three months of age.

3. All infants with confirmed permanent hearing loss receive services before six months of age in interdisciplinary intervention programs that recognize and build on strengths, informed choice, traditions, and cultural beliefs of the family.

4. All infants who pass newborn hearing screening but who have risk indicators for other auditory disorders and/or speech and language delay receive ongoing audiologic and medical surveillance and monitoring for communication development.

5. Infant and family rights are guaranteed through informed choice, decision-making, and consent.

6. Infant hearing screening and evaluation results are afforded the same protection as all other health care and educational information.

7. Information systems are used to measure and report the effectiveness of services. Although state registries measure and track screening, evaluation, and intervention outcomes for infants and their families, efforts should be made to honor a family's privacy by removing identifying information whenever possible.

8. Programs should provide data to monitor quality, demonstrate compliance with legislation and regulations, determine fiscal accountability and cost effectiveness, support reimbursement for services, and mobilize and maintain community support.

Current Status

With the opening of the millennium, although much had happened across the nation over the past decade, efforts fell short of Surgeon General Koop's goal that by 2000 all infants with a significant hearing loss would be identified by 12 months of age. Since Hawai`i passed the first legislation in 1990, 31 other states have passed legislation to mandate universal newborn hearing screening. Legislation is in progress in eight other states (National Center for Hearing Assessment and Management Web site, January 18, 2001). Figure 1 shows the states with legislation, states with legislation pending, and states without legislation. Some states are achieving success in instituting voluntary programs without the need for mandating legislation.

As of June 2000 the following nine states had universal screening programs in place screening 95% or more of all births: Arizona, Colorado, Connecticut, Hawai`i, Massachusetts, Mississippi, Rhode Island, Utah, and Wyoming. Unfortunately, efforts to assure the necessary diagnostic follow-up effective intervention services were spotty in all states including those nine states. Linkages to facilitate a smooth seamless statewide system of services relative to the goals of universal newborn hearing screening is essential for the program to be successful in achieving its goals (Vohr, Simon, and

Letourneau, 2000). Obtaining resources for assuring follow-up and providing necessary intervention services remains a challenge for many programs.

These failures to provide appropriate diagnostic and intervention services are particularly disturbing given the results of several recent studies. Research sponsored by the NIH has demonstrated that deaf and hard-of-hearing infants who receive intervention before six months of age maintain language development commensurate with their cognitive abilities through the age of five years (Yoshinaga-Itano, 1995; Yoshinaga-Itano, et al., 1998).

The findings of Yoshinaga-Itano have been supported by the more recent work of Moeller (2000). She studied a group of 112 children with hearing loss who were enrolled at various ages in a comprehensive intervention program. She found that significantly better language scores were associated with early enrollment in intervention. Moeller's results suggested that success is achieved when early identification is paired with early interventions that actively involve families.

The National Institute on Deafness and Other Communication Disorders (NIDCD) at the NIH is currently supporting research in hearing, speech and language, and early identification of hearing impairment in infants and young children. NIDCD has awarded research grants related to optimizing amplification for infants and young children, for improving speech, for evaluation of cochlear implants, and for information development to investigate the delays between the identification of hearing loss in infants and referrals for intervention.

The Office of Special Education and Rehabilitative Services (OSERS) in the U.S. Department of Education is another important federal partner. OSERS has just funded the University of North Carolina to develop a new training center in early intervention to develop modules for training personnel who will be working with infants who are deaf or hard of hearing. The center will also address issues involving early identification and referral in assessment, evaluation, and intervention.

The Centers for Disease Control has awarded funds to Utah State University to conduct research on the etiology of pre-lingual hearing loss. This study will be based on children identified through the Utah State Newborn Hearing Screening Program. The researchers will attempt to determine the causes of hearing loss for all identified infants. Infants for whom no known cause is identified will be tested for mutations of the connexin 26 gene.

Utah State University will also conduct a cost analysis study to identify the resources and costs of hearing screening, diagnosis, tracking, and follow-up for infants identified through the Utah newborn hearing screening program. This study is also funded by the Centers for Disease Control.

The Healthy People 2010 Goals call for even more ambitious achievements than did the Year 2000 Goals. The intent is that infants will be screened for hearing loss by one month, have audiologic evaluations by three months, and be enrolled in appropriate intervention services by age six months.

The recently published research documenting the efficacy of neonatal hearing screening, the recognition that early identification and intervention can obliterate the communication delays resulting from late identification, the surge in state legislation, and the major new federal funding efforts to strengthen state efforts will hopefully result in

eradication of the delayed identification of hearing loss in young children. Such an outcome will have positive results for children, their families, and society.

[Figure 1 here]

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