Universal Newborn Hearing Screening: A Goal Being Achieved in Hawaii

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Abstract
This article describes the importance of early identification of hearing loss in newborns, the current status of newborn hearing screening in the United States, and the leadership that Hawaii has contributed to that effort. Described are events that may help the nation reach the Year 2010 Health Goals for newborn hearing screening, identification, and intervention.

Introduction
The promise of universal newborn hearing screening got a needed boost in 1989, when the U.S. Surgeon General, C. Everett Koop, established a Year 2000 Health Goal that all children with significant hearing impairment be identified before 12 months of age. While the goal was not achieved by the Year 2000 for all children, significant progress has been made. This article describes the importance of that goal, the current status of newborn hearing screening in the United States, and the leadership that Hawaii continues to demonstrate in helping the nation achieve this goal. The article also describes events that may make this goal a reality over the next decade.

Importance of Early Identification
The devastating effects of a severe-to-profound hearing loss on a child have long been recognized. Lack of hearing, and lack of 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. 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. Inability of the child 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. 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 AT&T HITS delayed.

Technological Advances
Prior to 1990, early identification was hampered not only by physicians’ difficulty in recognizing the presence of a hearing loss, but also by the lack of technological capability of audiologists to diagnose a hearing loss using objective physiological measures. The possibility of universal newborn hearing screening came a step closer to reality with the development of instrumentation to measure oto-acoustic emissions (OAE) and auditory brain stem responses (ABR). This technology was developed in the 1980s. It then became feasible to use an objective, non-invasive screening process that could be administered by hospital staff, paraprofessionals, or volunteers to screen all infants prior to discharge from the newborn nursery. Recent research studies have demonstrated the validity, reliability, and effectiveness of universal newborn hearing screening.

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 Hawaii. 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 Hawaii.

The first state legislation to mandate the hearing screening of all newborns was passed in Hawaii and signed into law in May 1990. 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. The statement concluded that all infants admitted to the neonatal intensive care unit (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 multicenter study to determine the accuracy of three measures of peripheral auditory system status (transient evoked oto-acoustic emis-

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sions, distortion product oto-acoustic emissions, and auditory brain stem responses) applied in the perinatal period for predicting behavioral hearing status at 8 to 12 months corrected age. Seven institutions, with an annual census of 7,179 births, participated in the study.

The results of that study were published in late 2000. The study showed conclusively that all three screening test resulted in low rates of referral and were successful in identifying hearing loss in newborns. Researchers found that, although perfect test performance was never achieved, sensitivity for each measure increased with the magnitude of the hearing loss. This important finding suggests that all three tests can successfully identify newborns with hearing loss, for which interventions 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. 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 that study was establishment of the “gold standard,” using a visual reinforcement audiometry protocol. That 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.

National efforts received a significant boost in 1999 when federal legislation provided 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. Dr. Calvin Sia, as Chair of the Professional Advisory Committee of the American Academy of Pediatrics, established a special Task Force on Universal Newborn Hearing Screening to develop bipartisan consensus to get the legislation passed.

The Walsh Bill included appropriations that provided $3,500,000 in funding for the Maternal and Child Health Bureau in Health Resources and Services Administration. This money was to provide grants to states to develop statewide programs in 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 in the year 2000 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 and Prevention to fund data systems and research activities. The Centers for Disease Control and Prevention 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 provided to both agencies to further expand those activities.

The Centers for Disease Control and Prevention 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.

Hawaii Grants
Hawaii has been fortunate to have projects funded since the Walsh Bill was passed, receiving one grant from the Office of Maternal and Child Health and two grants from the Centers for Disease Control and Prevention. As one of the first group of 22 states funded to enhance statewide systems development, the Hawaii Department of Health, Children with Special Health Needs Branch, received a three-year grant of $136,000 annually to further refine its system to ensure that all young children with hearing loss will achieve developmentally appropriate milestones for language and communication. The grant included two goals: 1) improving the system of hearing screening, assessment, and linkage with early intervention services; and 2) refining family support and early intervention services to meet the needs of young children with hearing loss and their families.

In addition, the Children with Special Health Needs Branch was one of the first agencies in 15 states funded by the Centers for Disease Control and Prevention to enhance the data system, which supports universal newborn hearing screening, and integrate it with related data systems to facilitate completion of the process of assessment, tracking, and research. This grant is for a five-year period, with Hawaii receiving approximately $98,000 each year.

The Center on Disability Studies at the University of Hawaii was the only applicant funded to conduct a research study on the efficacy of the most common screening protocol. Currently underway is a large multi-site national study to validate the two-step screening procedure that first uses OAE, then with follow-up automated ABR for those infants who do not meet the response criteria under OAE. The current protocol requires diagnostic follow-up only for newborns who are referred based on both the screening measures. Concerns arose based on anecdotal data that some newborns, especially those with a mild-to-moderate loss, might fail the OAE screening but pass the ABR screening.

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 infants with a significant hearing loss are being routinely missed by this procedure. The University of Hawaii is leading this study in collaboration with the National Center for Hearing Assessment and Management and twelve birthing centers across the nation. This is a two-year grant funded at more than $500,000. The findings of this study will determine whether this most common screening protocol should be modified to assure that no infant with a significant hearing loss is missed in the screening process.

Year 2000 Position Statement
Another important boost for universal newborn hearing screening occurred in June 2000, when the Joint Committee on Infant Hearing officially endorsed universal newborn hearing screening with the publication of the Year 2000 Position Statement: Principles and Guidelines for Early Hearing Detection and Intervention Programs. 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 all the relevant organizations serving deaf children and adults.

The Joint Committee on Infant Hearing 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 early intervention activities with the goal of maximizing linguistic and communicative competence and literacy development for all children, especially those who are deaf or hard of hearing.

Current National Status

With the opening of the millennium, although much had happened across the nation in the decade following publication of Surgeon General Koop’s goal that by the year 2000 all infants with a significant hearing loss would be identified by 12 months of age, efforts fell short of that goal. Since Hawaii 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.

As of June 2000, the following nine states had universal screening programs in place, screening 95% or more of all births: Arizona, Colorado, Connecticut, Hawaii, Massachusetts, Mississippi, Rhode Island, Utah, and Wyoming. Unfortunately, efforts to assure the necessary diagnostic follow-up and effective intervention services were spotty in all states, included those nine states. Linkages to facilitate a smooth, seamless statewide system of services is essential for the program to be successful in achieving the goal of improving communication skills of deaf or hard-of-hearing children. 20 Obtaining resources for assuring follow-up and providing the 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. 21,22

The findings of Yoshinaga-Itano have been supported by the more recent work of Moeller. 23 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 suggest 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 in early identification of hearing impairment in infants and young children. The NIDCD has awarded research grants related to optimizing amplification for infants and young children, for improving speech, for evaluation of cochlear implants, and for investigating 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 federal partner. OSERS has just funded the University of North Carolina to develop a new training center in early intervention that will 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 and Prevention 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 newborn hearing screening, diagnosis, tracking, and follow-up for infants identified in Utah. This study is also funded by the Centers for Disease Control and Prevention.

Current Status in Hawaii

Hawaii’s program has made great strides over the past decade in implementing universal newborn hearing screening. The number of hospitals providing screening has increased from one hospital in 1991-1992 to 15 hospitals in 1999-2000. When the last hospital in the state, Molokai Hospital, began screening babies in January 2000, newborn hearing screening became the standard of care for all babies born in birthing facilities. In the first six months of 1999, 98% of all newborns had their hearing screened prior to hospital discharge. The number of children identified with a confirmed hearing loss has increased each year, from only 20 in 1991-1992, to 121 in 1998-1999. The average age of identification has decreased from 16 months in 1991 to only one month in 2000.

As a result of newborn hearing screening, incidence rates have been established. The rates for the period of January 1998 through June 1999 were an incidence of permanent hearing loss of 4.3 per 1,000 births. Of those births, 3.4% had a sensorineural hearing loss and 0.9% had a permanent conductive hearing loss.

Medical Home. The success of Hawaii’s program is attributed to its strong linkage to the medical home of each newborn. Hearing screening results and recommendations are provided to each child’s Medical Home. Physicians discuss those results with the family during their first well-baby check-up. The encouragement by physicians for families to complete any recommended follow-up has facilitated successful follow-up for a high percentage of the newborns. Once the diagnosis is made, families return to their Medical Home for assurance and guidance. It is routine practice for pediatricians to contact the early intervention system for consultation to discuss the infant’s needs.

Because hearing loss can occur after birth, physicians’ enhanced surveillance of children’s hearing status has resulted in earlier detection of late onset hearing loss. Three children who passed
newborn hearing screening, but had risk factors for late onset of hearing loss, were promptly referred by their physicians for audiological evaluation and were found to have a significant hearing loss.

**Legislation.** Legislation was passed in the 2001 Session of the Hawaii Legislature to amend the state's mandate. This bill strengthened Hawaii's legislation by requiring rules and regulations to standardize newborn hearing screening and to outline the responsibilities of the different entities.

**Cochlear Implants.** As a result of the very early identification of deafness in newborns, the use of cochlear implants is becoming a common option among families for these young children. The cochlear implant is an electronic device that is surgically implanted in the ear (temporal bone), with the external part worn like a hearing aid. However, a cochlear implant is not a hearing aid; it does not make sounds louder or clearer. It is a medical device that bypasses damaged parts of the inner ear and electronically stimulates the auditory nerve.

Currently, four infants in Hawaii received cochlear implants when they were between the ages of 18-20 months. The surgery is performed in Hawaii at Queen's Hospital. Three of these children are now enrolled in a communication play-group using an auditory-oral approach. One child entered a Department of Education preschool in fall 2001. The initial responses of these four infants encourages cautious optimism that this device will help these children achieve age-appropriate oral communication skills.

**Auditory Dys-synchrony.** Another by-product of earlier identification is enhanced understanding and earlier diagnosis of auditory dys-synchrony, previously referred to as auditory neuropathy. This condition is where the cochlea is intact and functioning but auditory brainstem responses are absent or abnormal. This condition results in abnormal behavioral responses to sound and has previously been mistaken for profound hearing loss. With the availability of OAE testing, differential diagnosis of this condition is now possible.

Five children have been diagnosed with this condition in Hawaii, following newborn hearing screening. A team-approach is required to provide appropriate habilitative management for these children.

**Data Integration.** At the state and national level, attention is being directed toward developing an integrated data system for data systems on newborns. The purpose is to streamline data entry by avoiding redundancy, to improve the integrity of data, and to provide a basis for epidemiological and outcomes research. Hawaii is currently working toward integrating newborn screening data (metabolic and hearing) at the hospital level. Further efforts will be directed toward linking these data with those of the Title V Children with Special Health Needs Program and with the Part C Early Intervention Program.

**Genetics Issues.** Another major challenge has been providing information to families regarding genetics and hearing loss of unknown etiology. Although no uniform protocol is in place, national groups are addressing this issue, and recommendations are expected soon. Some families may request a genetics study. In the fall of 2001, the Hawaii Department of Health submitted a grant to develop a protocol that could be customized to this population.

**Case Studies**

But despite the tremendous successes the program has achieved, some newborns unfortunately continue to elude the early identification process. The following two case studies illustrate Hawaii newborns who did not benefit from the universal newborn hearing screening program in recent years.

**Kekulani.** Kekulani was referred by his pediatrician for an audiological evaluation at the age of two because she had not yet uttered her first word. Her birthing hospital record indicated that she had failed the newborn hearing screening prior to discharge but had not returned for the rescreening. Several attempts were made to contact the family, but the mother refused the rescreening because she felt certain her daughter's hearing was normal. Results of the audiological evaluation found a bilateral, profound, sensorineural hearing loss.

**Jimmy.** Jimmy was 40 months old when he was first seen for an audiological evaluation to rule out hearing loss. He had no intelligible expressive language at that time. The hospital record did not indicate that any hearing screening had been done, but a written notation in his medical record stated, "hearing is okay." The basis for this statement is unknown. Test results indicated a profound sensorineural loss in the left ear with a severe mixed loss in the right ear. Further ENT examination revealed the presence of a cholesteatoma in the right ear, which was likely congenital. Monitoring of the unilateral hearing loss in the left ear would have produced a much earlier identification of the cholesteatoma and, likely, less of a permanent hearing loss in the right ear. Because of the late identification, this child did not benefit from early intervention services.

**Summary**

The Healthy People 2010 Goals call for even more ambitious achievements than did the Year 2000 Goals. The goal is that infants will be screened for hearing loss by one month, have audiologic evaluations by three months, and be enrolled in appropriate early intervention services by age six months. Hawaii has already achieved these goals! But as the preceding case studies illustrate, further improvements are needed to assure that not a single newborn is missed.

The recently published research documenting the efficacy of newborn hearing screening, the recognition that early identification and intervention can mitigate 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 the eradication of the delayed identification of hearing loss in young children. Such an outcome will have positive results for children, their families, and society.

Unlike many other areas (mental health services, funding of public education, etc.) where Hawaii is ranked at the bottom of the nation, universal newborn hearing screening is an area in which the state has demonstrated national leadership, leading the nation on most key indicators.

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**References**


**HAWAII POISON CENTER**

**OAHU: 941-4411**
**NEIGHBOR ISLANDS TOLL-FREE:**
**1-800-362-3585**
Free Hotline 24 Hours a Day.

**POISON CENTER TIPS**

- Keep the number of the Hawaii Poison Center on or near your telephone.
- If you suspect a poisoning, do not wait for signs and symptoms to develop. Call the Hawaii Poison Center immediately.
- Always keep Ipecac Syrup in your home. (This is used to make a person vomit in certain types of poisoning.) Do not use Ipecac Syrup unless advised by the Hawaii Poison Center.
- Store all medicines, chemicals, and household products out of reach and out of sight, preferably locked up.
- A good rule to teach children is to “always ask first” before eating or drinking anything—don’t touch, don’t smell, don’t taste.

**Donate to help us save lives.**
Mail checks, payable to:
Hawaii Poison Center
1319 Punahou Street, Honolulu, HI 96826