

# Newborn Hearing Assessment In 2010

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In 1993 the Rhode Island General Assembly mandated universal newborn hearing screening (UHNS) and required third-party payment for screenings (RI Public Law 23-23-13). Rhode Island was the first state in the nation to do so. Today 45 states and territories have statutes related to UNHS. A large body of research supports the effectiveness of newborn hearing screening in detecting hearing loss at an early age and reducing the age at which children with hearing loss are enrolled in Early Intervention (EI) programs. Current challenges include improving post-diagnosis follow-up; ensuring timely enrollment in EI; appropriately and effectively referring for amplification; and monitoring for later-onset hearing loss.

## JUSTIFICATION FOR INFANT HEARING SCREENINGS

Failure to detect hearing loss in children may result in lifelong deficits in speech and language acquisition, poor academic performance, personal-social maladjustments, and emotional difficulties.<sup>1</sup> Neonatal hearing loss is one of the most commonly occurring birth defects with an incidence of 1.1 per 1000 live births, or some 12,000 to 16,000 babies each year in the United States.<sup>2,3</sup> As many as half of these infants have no risk factors for hearing loss; thus hearing loss may not be suspected until the child misses speech milestones. Prior to newborn screening, clinicians in the US did not diagnose hearing loss until an average age of 30 months,<sup>4</sup> by which time the child lagged behind his or her age-peers with respect to speech and language acquisition. In the US in 2007, 66.4% of infants were diagnosed with either normal hearing or hearing loss by age 3 months; in Rhode Island the rate was 93.3%.<sup>5</sup>

## RHODE ISLAND PROGRAMS

Following the passage of legislation mandating UNHS, the Rhode Island Hearing Assessment Program (RIHAP) was founded to oversee and coordinate the screening programs at the eight (now seven) birthing hospitals in the state. RIHAP continues to serve in this role, pro-

viding training, equipment, and technical support for each of these hospitals. As importantly, RIHAP maintains an extensive database of all the outcomes of statewide hearing screenings; coordinates follow-up testing for those who either failed their screening or are otherwise at risk for hearing loss; and facilitates the treatment of infants diagnosed with hearing loss.

RIHAP works with Early Intervention (EI) programs throughout the state, the Family Guidance Program at the RI School for the Deaf, and with audiologists throughout the region. Rather than being the focal point of the program, RIHAP serves as the conduit for children with hearing loss to enter the statewide program,<sup>6</sup> which operates under the oversight of the RI Department of Health and receives funding from state and federal grants as well as support from individual hospitals and treatment facilities.

## NATIONAL PROGRAMS

The Early Hearing Detection and Intervention (EHDI) Program of the Centers for Disease Control and Prevention (CDC) works with federal, national, and state agencies to develop newborn hearing screening programs in each state. As of 2010, 53 of 59 US States, Districts, and Territories have cooperative agreements with EDHI and receive funding and support through the program.<sup>7</sup> EHDI goals include the "1-3-6 plan," which calls for all infants to be screened by age one month, all children who do not pass the screening to receive diagnostic audiological testing by age three months, and all

children with confirmed hearing loss to be enrolled in an appropriate intervention program by the age of six months. Other goals include identification of later-onset hearing loss, coordination of EI with the primary care provider, tracking programs to minimize loss to follow-up, and comprehensive monitoring to assure that each state program is working efficiently to meet these goals.

Currently 45 U.S. states and territories have legislation regarding newborn hearing screening and 26 states have legislation which mandates universal newborn hearing screening. Twenty-nine states require reporting of hearing screening results to the Department of Health.<sup>4</sup> According to 2007 EHDI data, 45 of 47 states and territories reporting statistics for that year screened over 90% of newborns and 37 out of these 47 reporting entities screened over 95% of their infants.<sup>5</sup> Rhode Island consistently screens over 99% of newborns.

## BENEFITS OF NEWBORN HEARING SCREENING

Research into the efficacy of UNHS confirms that newborn hearing screening significantly speeds the diagnosis of hearing loss and initiation of intervention. In 2008 the US Preventive Services Task Force recommended routine screening of newborns for hearing loss based on strong evidence of the positive long-term benefits of early diagnosis.<sup>8</sup> Sininger et al. concluded that infants screened for hearing loss are diagnosed 24.6 months earlier, fit with amplification 23.5 months earlier,

Table 1: Hearing Screening:  
Comparison of RI to Totals for the 2007 EDHI Data

State / Territory	Rhode Island	Totals (n = 47)
Total Births	13,304	3,449,300
Died/Parents Declined Screening	136	15,523
Infants Eligible for Screening	13,168	3,433,777
Number of Infants Screened	13,123	3,345,629
Percent Screened	99.7 %	97.4 %
Number of Infants Not Screened	45	88,148
Percent Not Screened	0.3%	2.7%

[http://www.cdc.gov/ncbddd/ehdi/2007-data/2007\\_Screen\\_Web\\_Rev.pdf](http://www.cdc.gov/ncbddd/ehdi/2007-data/2007_Screen_Web_Rev.pdf)

**Table 2: Diagnosis of Hearing Loss:  
Comparison of RI to Totals for the 2007 EDHI Data**

State / Territory	Rhode Island	Totals (n = 44)
Total Screened	13,123	2,881,738
Total Not Passed	203	60,950
Total Normal Hearing	165	22,266
Total Normal Hearing <3 Mos Age	158	15,295
Total Hearing Loss	14	3,340
Total Hearing Loss < 3 MosAge	9	1,757
Total Diagnosed	179	25,696
Total Diagnosed < 3 MosAge	167	17,052
Percent Diagnosed < 3 Mos Age	93.3%	66.4%

[http://www.cdc.gov/ncbddd/ehdi/2007-data/Diag\\_2007\\_Age\\_Web\\_Rev.pdf](http://www.cdc.gov/ncbddd/ehdi/2007-data/Diag_2007_Age_Web_Rev.pdf)

and enrolled in EI 20.2 months earlier than infants who are not screened.<sup>9</sup> Vohr et al. reported in a RI cohort that participation in EI services by three months of age was associated with improved early language skills in children with hearing loss. Though children with moderate to profound hearing loss continued to have expressive and receptive language delays, the benefits of EI for all children with hearing loss were demonstrated.<sup>10</sup> Yoshinaga-Itano concluded that those children with hearing loss and no other disabilities, when identified early and appropriately treated, have the ability to develop normal language skills.<sup>11</sup>

Advances in cochlear implant technology and surgical techniques have coincided with the proliferation of UNHS programs. The net result of this confluence has been the implantation of ever-younger patients identified as profoundly hearing impaired shortly after birth. These procedures have been found to be both safe and efficacious in multiple studies.<sup>12,13</sup> Sharma et al. and Dorman et al. have shown that neural plasticity within the auditory system begins to decline after approximately 3.5

years of age and that earlier implantation tends to result in normal or near-normal central auditory function.<sup>14,15</sup> Similarly, Philips et al. determined that earlier screenings led to more successful outcomes among children who were diagnosed with profound hearing loss and promptly fitted with cochlear implants. They concluded that this earlier intervention resulted in improved auditory receptive skills and speech intelligibility.<sup>12</sup>

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### UPDATES TO HEARING SCREENING PROGRAMS

The **Joint Committee on Infant Hearing (JCIH)**, a multi-agency task force concerned with the issues of childhood hearing loss, issued a position statement in 2007 with several revisions to their prior statement from 2000.<sup>3</sup> One significant change was the expansion of the defini-

tion of hearing loss to include hearing loss specific to the auditory nerve and to revise recommended screening protocols for infants at risk for these conditions. There is strong evidence that infants in the **neonatal intensive care unit (NICU)**, and specifically those with a history of hyperbilirubinemia, sepsis, and exposure to gentamicin, are at risk for damage to the eighth cranial nerve and/or auditory brainstem.<sup>16-18</sup> **Otoacoustic Emissions (OAE)** testing, which has traditionally been the initial screening tool of choice, measures the response of cochlear outer hair cells and thus may be normal even in the presence of retrocochlear dysfunction. To address these concerns, JCIH made new recommendations for the use of **automated auditory brainstem response (AABR)** screening in the NICU rather than OAE screening. Though generally more time consuming than OAE and somewhat less sensitive to mild hearing loss, the AABR is sensitive to dysfunction of the auditory nerve and thus will allow the hearing screening team to detect neural as well as cochlear hearing losses.

Other changes in JCIH recommendations include refinement of audiological and medical follow-up of infants with suspected hearing loss, specifications for EI programs, recommendations for data collection and management at the state level, and communication of hearing screening results to parents as soon as possible. Many states, including Rhode Island, have traditionally discussed only the results of failed screenings with parents, while passed screenings were recorded but not universally discussed with parents. Often even the fail results were communicated to the parents through the mail only after the newborn had been discharged from the hospital. These newest guidelines call for universal “results-to-parents” before discharge from the hospital.

**Table 3: Intervention/Referral to EI:  
Comparison of RI to Totals for the 2007 EDHI Data**

State / Territory	Rhode Island	Totals (n = 44)
Total Hearing Loss	14	3,364
Eligible for EI	10	2,539
Total Enrolled EI	10	1,792
Percent Enrolled in EI - <i>of those Eligible</i>	100%	70.5%
Percent Enrolled in EI - <i>of those w. Hearing Loss</i>	71.4%	53.3%
Enrolled EI - Before 6 Months	7	1,058
Percent Enrolled EI - Before 6 Months	70.0%	59.0%

[http://www.cdc.gov/ncbddd/ehdi/2007-data/2007\\_Enrolled\\_Part\\_C\\_EI\\_Web\\_Rev.pdf](http://www.cdc.gov/ncbddd/ehdi/2007-data/2007_Enrolled_Part_C_EI_Web_Rev.pdf)

### RECENT DEVELOPMENTS IN HEARING ASSESSMENT IN RHODE ISLAND

Hearing screening and diagnostic data for Rhode Island for 2007 are shown in Tables 1 and 2. Though RI has excellent rates of screening and low fail rates, the UNHS and EI programs in RI continue to adjust to meet national recommendations. In late 2009/early 2010 all of the birthing hospitals in Rhode Island started provid-

ing the results of all newborn hearing screenings to parents prior to discharge. Also in 2010 a new hearing screening protocol will be implemented in the NICU at Women & Infants Hospital. To address evidence-based concerns about neural hearing loss, initial screenings will be performed using AABR rather than OAE, following the recommendations of the JCIH.

Early Intervention data for Rhode Island are summarized in Table 3. EI programs remain a vital link in the care of children with hearing loss and other developmental challenges, providing services for children up to the age of three years. Though EI has suffered recent budget cuts, the program continues to meet federal requirements for the **Individuals with Disabilities Education Act (IDEA)** for infants, toddlers, and their families. Efforts are under way to ensure the uniformity of services across EI agencies and to coordinate these intervention services with primary care providers. Through focused “mentor groups,” EI ensures that at least one staff member in each facility receives specialized training to deal with children with autism, hearing loss and deafness, and other challenging conditions.

The Rhode Island Auditory-Oral Program opened in 2005 as a collaborative effort of the RI Department of Education, the Rhode Island School for the Deaf, participating school districts, and other organizations. This program offers specialized instruction for children who have hearing loss or are deaf to learn to listen and speak with the benefit of hearing aids or cochlear implants. The program caters to both preschool and school-aged children with classrooms including children with normal hearing and speech as well as those children with hearing loss or who are deaf. Professionals and family members work together at home and in the classroom to foster the skills necessary for receptive and expressive spoken language.

Finally, efforts are underway to improve the reporting of audiology data to the RI Department of Health. State law mandates the reporting of hearing loss identified before the age of five years; the JCIH recommends the reporting; and it is hugely important for coordination of care between audiologists, physicians, educators, and others charged with the stewardship of children with hearing loss.

The KIDSNET program has expanded to include hearing assessment data, and pediatric audiologists statewide are being trained to use this resource. At the same time, the program is being modified based on the observations and recommendations of these audiologists, with the goal of implementing a quick and comprehensive reporting mechanism and reference tool that can then help facilitate interventions.

## CONCLUSIONS

Rhode Island led the way toward universal newborn hearing screening 16 years ago and our state continues to be the vanguard of advancements. Research into long-term outcomes of infants diagnosed with hearing loss is ongoing, as is investigation of methods for reducing the number of children lost to follow-up. Over the next few years, individuals who were among the first generation of infants in the UNHS era will reach the age of majority, attend higher education, and enter the workforce. Only as these events unfold will we be able to gauge the true efficacy of our newborn hearing screening programs and their impact in Rhode Island.

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## Disclosure of Financial Interest

The authors and spouses/significant others have no financial interest to disclose.

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