

Newborn Hearing Screening: Making a Difference

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Newborn hearing screening has become the standard of care in the United States, with over 98% of infants born screened for hearing loss. Appropriate and timely follow-up is critical when an infant does not pass the screening. While there remains room for improvement, newborn hearing screening is making a difference.

Hearing loss is the most frequently occurring congenital condition, with a national prevalence rate of 1.6 per 1000 births [1]. In 2007 the Joint Committee on Infant Hearing (JCIH) issued the *JCIH Position Statement: Principles and Guidelines for Early Hearing Detection and Intervention Programs*, which recommends that all infants should have their hearing screened before 1 month of age [2]. For infants who do not pass the newborn hearing screening, diagnostic audiological evaluation should occur before 3 months of age. Infants with a confirmed hearing loss should be enrolled in an early intervention program before 6 months of age to facilitate age appropriate development of language and social skills. These 1, 3, and 6 months of age recommendations are commonly referred to as 1-3-6.

For the benefits of universal newborn hearing screening to be fully realized, “meticulous attention to many of the processes—including data management, reporting, and communication with families—within the Early Hearing Detection and Intervention [EHDI] system” is required [3]. Experts in all communication options (eg, American Sign Language, spoken language) state that “a child must be exposed to an accessible language on a regular and frequent basis before five years of age to develop full language competence” [4]. Language and learning outcomes are significantly improved when hearing loss is identified and intervention services are begun early [5, 6]. Mild and unilateral hearing loss in children can result in poor language or educational outcomes or lead to a child being incorrectly labeled as having a behavior problem [7]. Recognition of the critical role that appropriate and high quality early intervention plays for children who are deaf or hard of hearing (D/HH) led the JCIH to issue the *Supplement to the JCIH 2007 Position Statement: Principles and Guidelines for Early Intervention After Confirmation That a Child Is Deaf or Hard of Hearing* in 2013. This document “provides comprehensive guidelines for EHDI programs on establishing strong early intervention (EI) systems with

appropriate expertise to meet the needs of children who are D/HH” [8]. These best practice guidelines are designed to lead to well-coordinated EHDI systems to ensure every child who is D/HH is provided the opportunity to achieve optimal language, literacy, and learning outcomes.

In addition, the Centers for Disease Control and Prevention (CDC) has provided extensive guidance on the development, implementation, and use of EHDI information systems (EHDI-IS). The 2015 National EHDI-IS Functional Standards Working Group developed functional standards “intended to identify the operational, programmatic, and technical criteria that all jurisdictional EHDI programs should implement during the process of developing, using, and evaluating an EHDI Information System (EHDI-IS)” [9].

Early Hearing Detection and Intervention in North Carolina

Background

Legislation establishing North Carolina’s Early Hearing Detection and Intervention (NC-EHDI) Program was introduced in the North Carolina General Assembly in 1999 and formally adopted in July 2000 [10]. Administrative rules governing newborn hearing screening and reporting became effective in August of 2000 and were updated in August of 2004 [11]. The administrative rules include requirements for the reporting of all physiologic hearing screenings completed for infants less than 6 months of age and of all diagnostic auditory evaluations and assessments for selection of amplification for infants less than 12 months of age. Results are required to be reported within 5 days of the appointment, including missed appointments. NC EHDI maintains ongoing collaborations with families of children who are D/HH, state agencies, and other community partners. The Early Hearing Detection and Intervention Advisory Committee, established in 2001 by the NC EHDI program, is an active group of stakeholders that meets quarterly and is involved in

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program planning and providing ideas for systems change. The purpose of the EHDI Advisory Committee is to guide the development, coordination, and quality evaluation of community-based EHDI programs.

North Carolina Guidelines and Statistics

NC EHDI aligns with the evidence-based JCIH recommended best practices and guidelines for early hearing detection and intervention. Currently, all of North Carolina's 87 birthing and neonatal facilities offer initial newborn hearing screening services prior to infant discharge from the facility and report screening results using Hearing Link, North Carolina's EHDI-IS. North Carolina had 122,675 live births reported to the EHDI program in 2014, which ranks 9th highest in the United States.

Hearing Link meets 84% of the CDC's EHDI-IS Functional Standards. The system meets 100% of the minimum data element requirements and 86% of the core data element requirements outlined in the Functional Standards. The 2 remaining core data elements (mother's date of birth and father's date of birth) are collected as maternal and paternal age at the time of the infant's birth in North Carolina's system. Data in the NC EHDI-IS is linked with both the North Carolina State Laboratory of Public Health and Vital Records.

There are 11 regional EHDI consultants who provide training and consultation to pediatric audiologists, physicians, and other child service providers on the newborn screening process, the use of NC EHDI-IS, and recommended practices in diagnosis of hearing loss, amplification fitting, and referral to and enrollment in early intervention services. They also provide technical assistance, training, and consultation

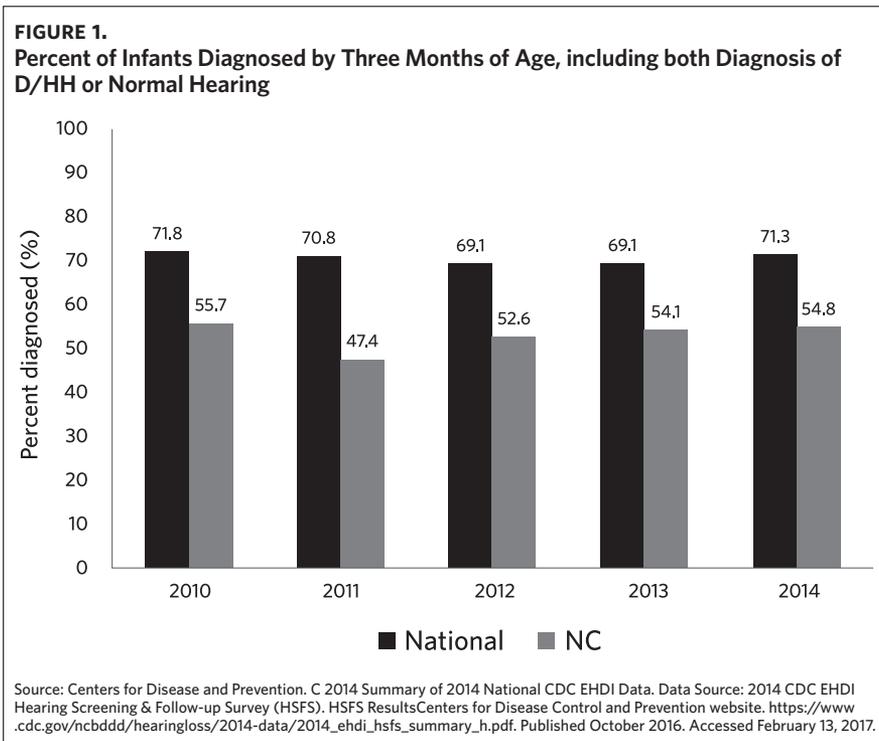
to families and other providers on normal communication development, and conduct educational programs for community groups and agencies regarding newborn hearing screening, language development, and communication-related issues.

NC EHDI consistently exceeds the benchmark for completion of newborn hearing screening by 1 month of age, but still faces significant challenges in reaching the 3-6 benchmarks. Of North Carolina live births reported to the EHDI program in 2014, 97.9% received a newborn hearing screening prior to one month of age. Data show 779 infants did not pass the newborn hearing screening process (0.6%). Of those infants, 247 (31.7%) never received the recommended outpatient rescreen. Of the 70% that did receive the recommended rescreen, 225 infants were diagnosed with permanent hearing loss [12].

Challenges

Even though there have been many successes with newborn hearing screening, several challenges remain to reach the ultimate goal for every D/HH child to reach their full potential.

First, specialized training in pediatric audiology is required to effectively diagnose and treat hearing loss in infants. There are only a few qualified pediatric audiologists in each state, resulting in long-distance and costly travel for families. An infant may be referred first to a local ENT (Ear, Nose and Throat physician) or audiologist who cannot perform the required test battery and then be sent to a pediatric audiologist later. This can increase parental anxiety or confusion. Timeliness of diagnosis is also an important factor,



and though improvement has been made in each of the past 3 years, only 55% of infants referred for diagnostic evaluation in North Carolina were diagnosed by 3 months of age, compared to the national average of 69% (see Figure 1). Due to the rapid growth of the child, frequent visits to the audiologist are also necessary.

Second, issues related to data reporting include underreporting of rescreening and/or diagnostic evaluations, amplification fitting, risk factors for late-onset or progressive hearing loss, and early intervention referral, eligibility, enrollment, and outcome data. North Carolina follows national trends for referral to early intervention (see Figure 2), but does not meet the national average for enrollment into early intervention (see Figure 3). Current administrative rules in North Carolina do not include requirements to report enrollment in early intervention, date of Individualized Family Service Plan (IFSP), or intervention assessment (eg, speech, language, literacy) results to the EHDI program. There are also limitations to long-term outcome data. For example, the Department of Public Instruction and schools for the deaf can provide data on children who are identified as hearing impaired and have an Individualized Education Plan (IEP). However, they are unable to report outcomes for children who may have a hearing impairment, but do not require an IEP.

Third, false-positive (and false-negative) rates for newborn hearing screening remain a concern. Existing research has mostly been conducted by equipment manufacturers and used for marketing. There are many factors that can influence the accuracy of newborn hearing screening including the skill of the person conducting the screening, calibration of the screening equipment, noise in the room where the screening is being conducted, pass/fail algorithms used

by manufacturers, number of screening attempts made for each infant, and more. Another concern is the parental anxiety a false-positive screening may cause. There is evidence that this anxiety is not long-lasting for parents, especially when clear communication occurs between the screener(s) and the family [13]. False-negative newborn hearing screening results can result in parental complacency and/or late diagnosis, as described by a parent below:

We are Deaf parents of 3 Deaf children. In our personal experience the newborn screenings were not without failure. All 3 children passed the screen at birth as hearing. The first child we didn't find out she was actually Deaf till she was over 1 year old (lots of suspecting, but we had no reason to question the initial screenings)... After that life event we realized we needed to have each child tested and not just as newborns. Both of the others passed in the hospital but ABRs at 2 months showed they were actually Deaf.

— Erin O'Donnell, parent of 3 Deaf children

Benefits

The American Speech-Language-Hearing Association (ASHA) has reported the costs for screening newborns for hearing loss are estimated to be \$25-\$60 per infant, depending on screening protocol and technology used, staffing, and supplies. Vohr, et al [14] reported the costs per identified child were \$14,000-\$17,000, including diagnosis and intervention. Phenylketonuria (PKU), hypothyroidism, and sickle cell anemia are universally included in newborn screening, and hearing loss can be identified in a newborn for less than one-tenth of the cost for any of those 3 conditions. Lifetime economic savings have been estimated at \$429,900 per person for early identified hearing loss or

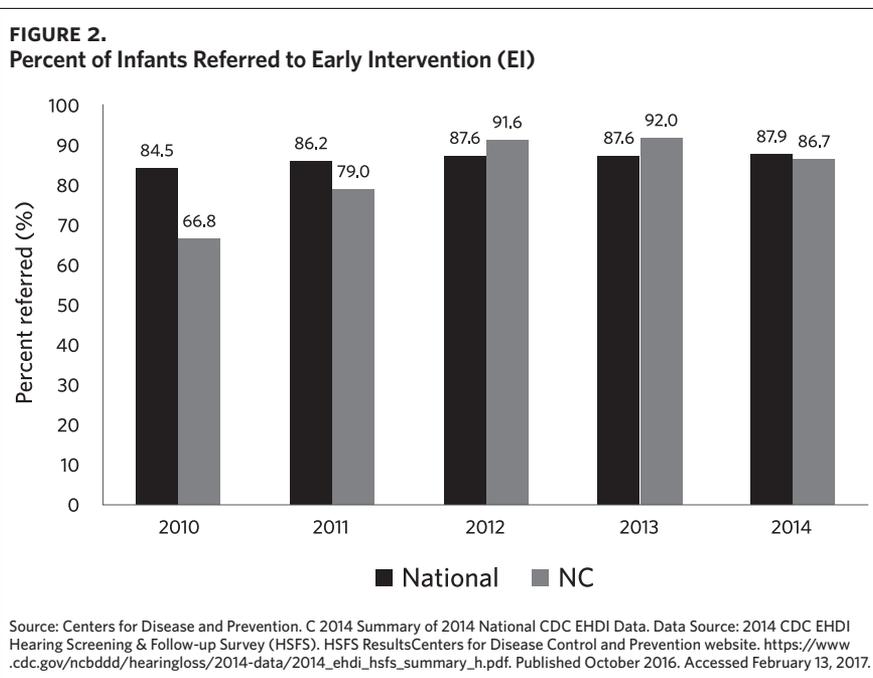
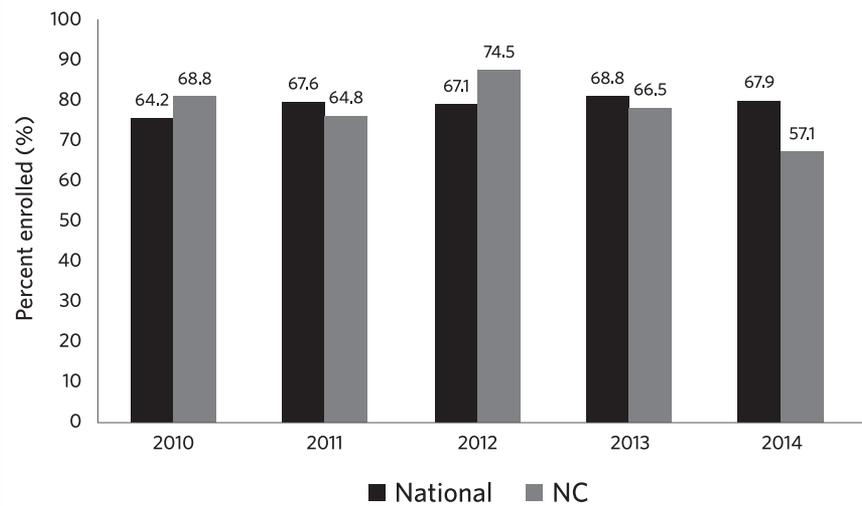


FIGURE 3.
Percent of Infants Enrolled in Early Intervention (EI) by Six Months of Age



Source: Centers for Disease and Prevention. C 2014 Summary of 2014 National CDC EHDl Data. Data Source: 2014 CDC EHDl Hearing Screening & Follow-up Survey (HSFS). HSFS ResultsCenters for Disease Control and Prevention website. https://www.cdc.gov/ncbddd/hearingloss/2014-data/2014_ehdi_hsfs_summary_h.pdf. Published October 2016. Accessed February 13, 2017.

deafness [15]. From 2004-2014, 1,353,957 infants received a newborn hearing screening and 2,985 infants were diagnosed with permanent hearing loss in North Carolina. Using the costs and economic savings cited above, the savings for our state have been \$1,164,808,650 over 10 years.

As mentioned previously, early detection and intervention can facilitate age appropriate development of language and social skills, as described by these two parents:

I am a strong supporter of newborn hearing screening. My oldest son wasn't identified until he was a year old, which caused a delay in his speech and language. My youngest son was tested as a newborn so we were able to get his services started really early, which allowed us to close the speech and language gap fairly quickly. There is a noticeable difference when it comes to their speech and language. I believe that had my oldest been properly diagnosed at birth he wouldn't have had to work as hard to catch up to his hearing peers.

— Alicia Spencer, parent of 2 D/HH children.
Executive Director/Parent Lead. NC Hands & Voices

Newborn hearing screening is a good investment. Getting intervention and quality services early prevents years of trying to make up for it (academically, socially, financially) later.

— Michael Shoun, father of a child with hearing loss

Conclusion

Is newborn hearing screening making a difference? Yes. Can newborn hearing screening be made better? Yes. Through newborn hearing screening and early intervention, we are developing confident young D/HH individuals who might just change the world! NCMJ

I am the perfect girl in my family. I'm a little too perfect from the others because they are not Deaf. So, too bad for them!

— Kaitlyn, age 8

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