

NE-EHDI Biannual Audiologist Newsletter

The Nebraska Early Hearing Detection and Intervention Program develops, promotes, and supports systems to ensure all newborns in Nebraska receive hearing screenings, family-centered evaluations, and early intervention as appropriate.

COVID-19

Thank you to those of you who have informed NE-EHDI of your revised procedures due to COVID-19!

During the pandemic, NE-EHDI will continue our current follow-up process of contacting families and medical professionals via e-mail, regular mail, secure e-fax, or phone. We will strive to meet the Joint Committee on Infant Hearing (JCIH) 1-3-6 recommendations to complete the hearing screen by 1 month of age, to complete the audiologic diagnostic evaluation by 3 months of age, and complete enrollment into early intervention by 6 months of age. However, we understand that there may be delays in the follow-up process during this uncertain time, and we will work with families and professionals to accommodate any changes due to limited access to care.

It will be important for all of us to work together when the restrictions are lifted to ensure any additional hearing screenings, diagnostic evaluations and referrals to EDN are caught up as quickly as possible so no child who may be deaf or hard of hearing is left behind or overlooked.



Working together to help children identified as Deaf or Hard of Hearing reach their full potential.



Joint Committee on Infant Hearing Position Statement

02

Diagnostic Audiology and Audiological Interventions



Visit the [JCIH website](#) to read the entire Year 2019 Position Statement: Principles and Guidelines for Early Hearing Detection and Intervention Program.

01

Newborn Hearing Screening

- Endorsement for well-born infants only who are screened by AABR and do not pass, rescreening and passing by OAE testing is acceptable, given the very low incidence of auditory neuropathy in this population.

PLEASE NOTE:

- This does not pertain to NICU infants.
- We still encourage medical providers to inform parents verbally and in writing that OAE does not rule out all types of hearing loss, including auditory neuropathy if used for rescreening after an AABR for well-born infants.
- Endorsement of rescreening in the medical home in some circumstances. If the rescreening is performed in the provider’s office, the provider is responsible for reporting results to the state EHDI program.

- A reaffirmation of the importance of fitting hearing aid amplification using objective, evidence-based protocols to ensure maximal audibility.
- Reaffirmation of the need to provide families with individualized support and information specific to language and communication development to support children who are deaf or hard of hearing by providing exposure to language models at the earliest possible age to ensure optimal cognitive, emotional, and educational development.

JCIH 1-3-6 Goals

The Joint Committee on Infant Hearing has established 1-3-6 goals for newborn hearing screening.



All infants should receive a hearing screening by **one** month of age..

All infants who refer should receive a diagnostic evaluation by **three** months of age.

All infants who are identified as deaf or hard of hearing should begin receiving early intervention services by **six** months of age.

03

Early Intervention and Family Support



- Reaffirmation of the need to provide families with individualized support and information specific to language and communication development to support children who are deaf or hard of hearing by providing exposure to language models at the earliest possible age to ensure optimal cognitive, emotional, and educational development.
- Recognition that some families may benefit from infant mental health supports. Infant mental health is a field of research and practice that focuses on optimizing social, emotional, behavioral, and cognitive development of infants in the context of the emerging relationships between parents and infants.

04

Medical Considerations



- Reaffirmation of the need for otologic/medical evaluation and management of the newly-identified infant to be carried out as soon as possible following confirmation, in an effort to address potentially reversible conditions, discover associated medical disorders that can impact the infant's general health, and identify conditions that can impact communication strategy choice.
- Recognition that Congenital Cytomegalovirus has a larger impact than previously recognized.
- Updated risk indicators for congenital hearing conditions, including a new table with specified intervals for audiologic evaluation.*
- Consideration of reduction in the FDA-approved age for cochlear implantation to less than 12 months.

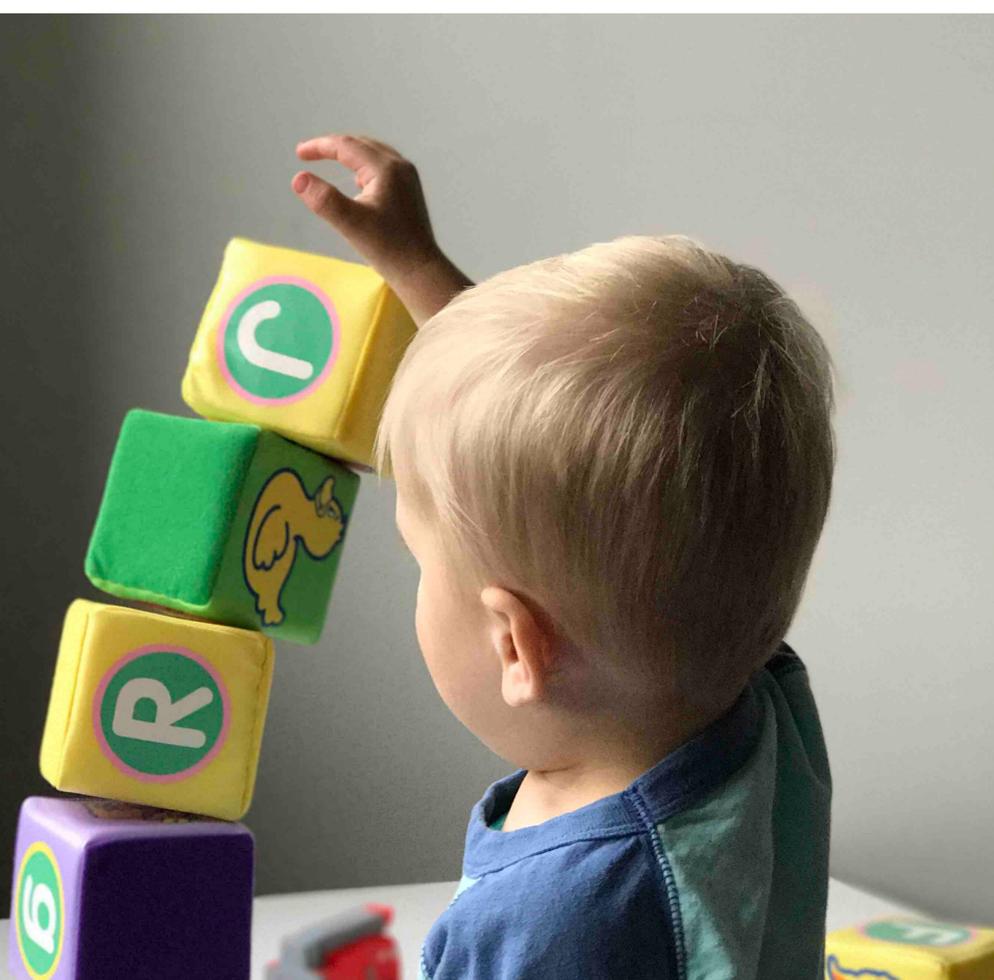
*See Table 1 - Risk Factors for Early Childhood Hearing Loss: Guidelines for Infants who Pass the Newborn Hearing Screen on page 6.

Medical Considerations cont.

Per JCIH, the goals of early hearing detection and intervention (EHDI) are to maximize language and communication competence, literacy development, and psychosocial well-being for children who are deaf or hard of hearing. Without appropriate language exposure and access, these children will fall behind their hearing peers in communication, language, speech, cognition, reading, and social-emotional development, and delays may continue to affect the child's life into adulthood. With early detection and appropriate, targeted intervention, developmental milestones for an infant who is deaf or hard of hearing can be expected to be achieved, more accurately reflecting the child's true potential (Tomblin, Oleson, Ambrose, Walker, & Moeller, 2014; Yoshinaga-Itano, Baca, & Sedey, 2010). Focusing on the importance of prompt diagnosis and timely, high-quality early intervention for such infants, EHDI systems should facilitate seamless transitions for infants and their families through the processes of screening, audiologic and medical diagnosis, and intervention.

Regardless of previous hearing-screening outcomes, all infants with or without risk factors should receive ongoing surveillance of communicative development beginning at 2 months of age during well-child visits in the medical home (AAP Committee, 2017). This recommendation provides an alternative, more inclusive strategy of surveillance of all children within the medical home based on the pediatric periodicity schedule (AAP Committee, 2017). All infants who do not pass the speech-language portion of developmental screening in the medical home or for whom *The Journal of Early Hearing Detection and Intervention* 2019; 4(2) 5 there is a concern regarding hearing or language should be referred for speech-language evaluation and audiology assessment. This protocol permits the detection of children with either missed neonatal or delayed-onset hearing loss, irrespective of the presence or absence of a high-risk indicator.

Both the family and the primary care provider should be advised that passing a hearing screening performed either by OAE or by AABR testing does not imply that hearing thresholds are within normal limits (WNL), only that thresholds are not greater than approximately 35-40 dB HL.

05**Referrals**

If the referral for the pediatric diagnostic audiology evaluation did not originate with the infant's primary care provider (PCP), a copy of the diagnostic audiology report should be sent to the PCP with recommendations for medical and otologic evaluations (AAP Committee, 2017; AAP, 2014a, 2014b) and the state EHDI program CDC, 2016a). In addition, a referral to the state Part C early intervention program must be made upon confirmation of a child being deaf or hard of hearing. Although the Part C revised guidelines state the referral must be made within seven days, immediate referral with a goal of 48 hours is recommended by JCIH in the early intervention best practices document (JCIH, 2013). Based on the 1-3-6 guidelines, referral to Part C should always be completed as soon as a child is diagnosed as deaf or hard of hearing, and always prior to six months of age. Diagnosis does not imply that thresholds are determined for all test frequencies, but rather, based on key frequencies (e.g., 500 Hz and 2000 Hz), it can be shown through air- and bone-conduction testing that probable permanent threshold elevation exists in one or both ears.

06

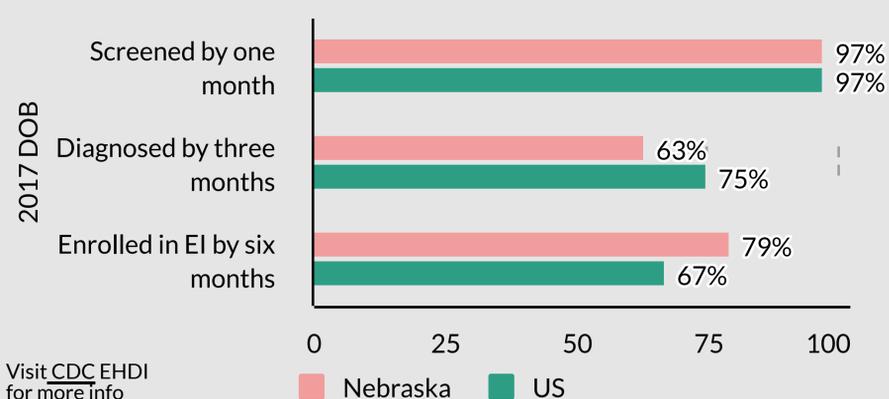
Role of the Audiologist Following Diagnosis



It is often the audiologist who provides the first information to the family for an identified child following diagnosis. The audiologist has the responsibility for communicating the information to the family in an empathetic, non-biased, open-ended fashion, and in a language (spoken or signed) that is accessible to the parents, using interpreters/ translators as necessary (Fitzpatrick, Durieux-Smith, Eriks-Brophy, Olds, & Gaines, 2007; Pizur-Barnekow, Darragh, & Johnston, 2011). Families retain information at different rates and require different styles of information delivery. Because of the complexity and volume of information, redundancy in the message and delivery of the message through multiple avenues (e.g., written, verbal, website, video presentation, etc.) is important. Most often, the audiologist is responsible for referral to the early intervention system. In some systems, the audiologist is the primary professional who conveys information about early intervention opportunities; in other sites it is an early intervention provider with specialized knowledge and skills in working with children who are deaf or hard of hearing.

Early, careful, and comprehensive education of families and caregivers regarding the nature of language acquisition is invaluable in encouraging families to seek appropriate early intervention services for their child. When counseling families, information regarding communication modes, methodologies, and technologies should be provided in a comprehensive and non-biased fashion. Families should be offered written materials in an accessible format and language. Information about listening and spoken language, signed language, and combined approaches should be provided. Additionally, information about amplification options (hearing aids, cochlear implants, visual and auditory assistive technologies) should be provided as appropriate for the infant's audiologic diagnosis, recognizing the possibility of progression of hearing thresholds to a more severe degree (ASHA, 2008a). Families benefit from contact with other parents who are trained to provide parent-to-parent or family-to-family support, and also benefit from contact with a trained professional who is deaf or hard of hearing (Moeller et al., 2013).

NE vs. US 1-3-6 Goals



Nebraska Hands & Voices Guide By Your Side

Family support plays a critical role in positive outcomes for children who are deaf or hard of hearing. The Nebraska Hands & Voices/Guide by Your Side program offers unbiased support to families of children who are deaf or hard of hearing.

Table 1
Risk Factors for Early Childhood Hearing Loss: Guidelines for Infants who Pass the Newborn Hearing Screen

| | Risk Factor Classification | Recommended Diagnostic Follow-up | Monitoring Frequency |
|----|---|---|---|
| | Perinatal | | |
| 1 | Family history* of early, progressive, or delayed onset permanent childhood hearing loss | by 9 months | Based on etiology of family hearing loss and caregiver concern |
| 2 | Neonatal intensive care of more than 5 days | by 9 months | As per concerns of on-going surveillance of hearing skills and speech milestones |
| 3 | Hyperbilirubinemia with exchange transfusion regardless of length of stay | by 9 months | |
| 4 | Aminoglycoside administration for more than 5 days** | by 9 months | |
| 5 | Asphyxia or Hypoxic Ischemic Encephalopathy | by 9 months | |
| 6 | Extracorporeal membrane oxygenation (ECMO)* | No later than 3 months after occurrence | Every 12 months to school age or at shorter intervals based on concerns of parent or provider |
| 7 | In utero infections, such as herpes, rubella, syphilis, and toxoplasmosis | by 9 months | As per concerns of on-going surveillance |
| | In utero infection with cytomegalovirus (CMV)* | No later than 3 months after occurrence | Every 12 months to age 3 or at shorter intervals based on parent/provider concerns |
| | Mother + Zika and infant with <u>no</u> laboratory evidence & no clinical findings | standard | As per AAP (2017) Periodicity schedule |
| | Mother + Zika and infant with laboratory evidence of Zika + clinical findings | AABR by 1 month | ABR by 4-6 months or VRA by 9 months |
| | Mother + Zika and infant with laboratory evidence of Zika - clinical findings | AABR by 1 month | ABR by 4-6 months |
| | | | Monitor as per AAP (2017) Periodicity schedule (Adebanjo et al., 2017) |
| 8 | Certain birth conditions or findings: • Craniofacial malformations including microtia/atresia, ear dysplasia, oral facial clefting, white forelock, and microphthalmia • Congenital microcephaly, congenital or acquired hydrocephalus • Temporal bone abnormalities | by 9 months | As per concerns of on-going surveillance of hearing skills and speech milestones |
| 9 | Over 400 syndromes have been identified with atypical hearing thresholds***. For more information, visit the Hereditary Hearing Loss website (Van Camp & Smith, 2016) | by 9 months | According to natural history of syndrome or concerns |
| | Perinatal or Postnatal | | |
| 10 | Culture-positive infections associated with sensorineural hearing loss***, including confirmed bacterial and viral (especially herpes viruses and varicella) meningitis or encephalitis | No later than 3 months after occurrence | Every 12 months to school age or at shorter intervals based on concerns of parent or provider |
| 11 | Events associated with hearing loss: • Significant head trauma especially basal skull/temporal bone fractures • Chemotherapy | No later than 3 months after occurrence | According to findings and or continued concerns |
| 12 | Caregiver concern**** regarding hearing, speech, language, developmental delay and or developmental regression | Immediate referral | According to findings and or continued concerns |

Note. AAP = American Academy of Pediatrics; ABR = auditory brainstem response; AABR = automated auditory brainstem response.

* Infants at increased risk of delayed onset or progressive hearing loss

** Infants with toxic levels or with a known genetic susceptibility remain at risk

*** Syndromes (Van Camp & Smith, 2016)

**** Parental/caregiver concern should always prompt further evaluation.



Please e-mail any improvement ideas you have for the EHDI Program and/or how we can work together to better serve families who have a child who is deaf or hard of hearing.

DHHS.NEEHDI@nebraska.gov

THANK YOU



for all the work you do to provide quality services to families in Nebraska!

May is...

Better Hearing & Speech Month



Communication at Work

For 2020, the American Speech-Language-Hearing Association's (ASHA) theme is "Communication at Work." They have many resources to help you celebrate all month long.

Please check for the latest materials and information on BHSM activities on the ASHA website.

New resources will be added throughout May 2020!



Please remember to make the referral to the Early Development Network (EDN) which provides early intervention for families with children birth to age three with developmental delays and/or health care needs and connects families to needed services.

This will help children identified as Deaf or Hard of Hearing (D/HH) stay on track with their hearing peers for developing speech, language, and social skills. Or if the child has not been identified as D/HH, but you suspect a significant level of developmental delay, then also please refer to EDN. Early intervention gives children who are D/HH the opportunity to reach their full potential.



June is...



Please take the time to educate your community about congenital cytomegalovirus (CMV). Congenital (CMV) is the leading viral cause of birth defects and disabilities, the leading cause of non-genetic childhood hearing loss, and 91% of women DON'T know about it. The awareness rate of congenital CMV as compared to other congenital diseases and infections is alarmingly low, and yet approximately 0.7% or 30,000 newborns are born with congenital CMV in the United States each year.

If a baby presents as symptomatic, congenital CMV (cCMV) can be diagnosed by testing a newborn baby's saliva, urine, or blood using polymerase-chain-reaction (PCR) testing. Ideally these specimens are collected for testing before twenty-one days of life in order to confirm a diagnosis of congenital CMV infection because, after three weeks, it is hard to determine if the baby could have contracted the infection through nursing or by exposure to siblings or others who may be shedding, or passing the virus.

A definitive diagnosis of CMV may provide the family an opportunity for antiviral therapy and focused surveillance hearing testing since these children are typically at-risk for late-onset or progressive hearing loss.

More information can be found at the National CMV Foundation website